Management approach and surgical strategies for retrorectal tumours: a systematic review

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Abstract

Aim The management strategy for retrorectal tumours is complex. Due to their rarity, few surgeons have expertise in management.

Method A systematic literature review was conducted using the PubMed database. English language publications in the years 2011–2015 that assessed preoperative management, surgical strategies and chemoradiotherapy for presacral tumours were included. Two hundred and fifty-one abstracts were screened of which 88 met the inclusion criteria. After review of the full text, this resulted in a final list of 42 studies eligible for review.

Results In all, 932 patients (63.2% female, 36.8% male; \( P < 0.01 \)) with a retrorectal tumour were identified. Most were benign (65.9% vs 33.7%, \( P < 0.01 \)). Imaging distinguished benign from malignant lesions in 88.1% of cases; preoperative biopsy was superior to imaging in providing an accurate definitive diagnosis (91.3% vs 61.4%, \( P < 0.05 \)) with negligible seeding risk. Biopsy should be performed in solid tumours. It is useful in guiding neoadjuvant therapy for gastrointestinal stromal tumours, sarcomas and desmoid type fibromatosis and may alter the management strategy in cases of diffuse large B-cell lymphoma and metastases. Biopsies for cystic lesions are not recommended. The gold standard in imaging is MRI. The posterior Kraske procedure is the most common surgical approach. Overall, the reported recurrence rate was 19.7%.

Conclusion This review evaluated the management strategies for retrorectal tumours. A preoperative biopsy should be performed for solid tumours. MRI is the most useful imaging modality. Surgery is the mainstay of treatment. There is limited information on robotic surgery, single-port surgery, transanal endoscopic microsurgery, chemoradiotherapy and reconstruction.

Keywords Retrorectal tumour, presacral tumour

Introduction

Retrorectal tumours are uncommon. It is estimated in major metropolitan centres that there are one to two patients admitted with a retrorectal tumour per year corresponding to a rate of one in 40 000–60 000 admissions per year [1]. The approach, surgical strategy and operations for retrorectal tumours are markedly different from rectal tumours and, due to their rarity, few surgeons can claim to have great experience in this field.

There is a wide range of types of retrorectal tumour. They are classified as congenital, inflammatory, neurogenic, osseous and miscellaneous [2–4]. Most series in the literature report the majority to be benign [5]. Congenital tumours are the most common [6,7], with chordomas being the most frequent [8].

Retrorectal tumours may be incidental; for example, benign cystic lesions may be asymptomatic in up to 50% of cases unless infection or malignant transformation develops [9]. They have been seen incidentally during gynaecological surgery [10,11] and staging in breast malignancy [12]. When symptomatic they may present with pain in the pelvis, back or abdomen, constipation [13–15], large bowel obstruction [16], palpable rectal mass [17], recurrent pilonidal sinus [18], pelvic [19] and perianal [20] abscess formation, irritable bowel syndrome [21] and abnormal gait [22]. The clinical presentation is heterogeneous and retrorectal tumours often pose diagnostic difficulty [23] as non-specific chronic pain and suppuration are the commonest symptoms [24]. There is usually a significant delay in diagnosis [21,25]. The median age for presentation is
45 years, but congenital and developmental tumours occur in younger patients [7]. The majority of patients are middle-aged women. In infants, presacral teratomas may be part of a syndrome called Currarino syndrome caused by HLXB9 mutation of chromosome 7q36 [26,27] or insertion in MNX1 gene [5].

Once a retrorectal tumour is diagnosed, most patients require surgery [28–31]. For benign tumours, this is because of the risk of misdiagnosis, malignant degeneration and risk of disease and symptom progression. Benign tumours such as tail gut cysts may progress to become neuroendocrine tumours or adenocarcinoma, teratomas have a high risk of malignant transformation, and myelipomas have been described in the literature to harbour malignancy despite a benign appearance. Benign appearing tumours may contain malignancy on pathological evaluation [8,32]. There are many approaches to the management of presacral tumours, and this review assesses the available evidence in the current literature that deals with its preoperative investigations, surgical strategies and neoadjuvant and adjuvant therapies. We also assess whether there is a role for non-operative management.

Method

Search strategy and selection criteria

Searches of PubMed included the search criteria ‘retrorectal tumours’ and ‘presacral tumours’. The last search was conducted on 4 June 2015 resulting in 59 and 192 references for articles published in English from 2011 to 2015. These were reviewed and articles were screened on title, abstract, limited to human studies. Duplicate studies were excluded. Eighty-eight full texts were assessed and studies that did not deal with investigation, management or surgical strategy of retrorectal tumours were also excluded. Case reports and retrospective reviews made up the majority of the articles. The reference list of all eligible studies was checked to avoid missing studies. This resulted in a final reference list of 42 studies.

Results

Preoperative biopsy

Most studies suggest that biopsy of presacral lesions does not add to the surgical strategy but may risk seeding of tumour through the biopsy track and cause a chronic fistula [33]. Biopsies may be performed via the rectal lumen using transrectal ultrasound or through a transneuroforaminal [34], transgluteal, transperineal or transsacral approach. Although there are concerns of biopsy site recurrence, there have been several recent studies which have shown that preoperative biopsy is safe, including that of Messick et al. [23] who demonstrated no recurrence in 24 patients who underwent a diagnostic biopsy for a retrorectal tumour.

The accuracy of preoperative biopsy of presacral tumours is variably reported, particularly for cystic lesions. Mitsuyama et al. [32] reported three of seven benign appearing cysts misdiagnosed by preoperative biopsy. There have been cases of what was deemed initially to be a benign presacral cyst found on the final pathology to contain adenocarcinoma [8,32,33] and carcinoid [35,36]. There have been around 16 cases of carcinoid within a tail gut cyst described in the literature [36]. Mathis et al. [37] reported four (adenocarcinoma three, carcinoid one) of 31 presacral tailgut cysts to contain malignant degeneration. It is suggested that 2–13% of tailgut cysts are associated with malignancy [35]. In the literature, there are approximately 155 cases of presacral tailgut cysts described in 94 articles. Of these, 47 were presacral tailgut cysts with malignant transformation [38] (Table 1).

For solid tumours, recent studies in the literature have shown good concordance between preoperative biopsy and final pathology. In a series of 26 patients with sacrococcygeal teratomas of whom 13 had a preoperative biopsy, all were in concordance with the final pathology [17]. Merchea et al. performed 76 biopsies on solid presacral tumours and demonstrated a sensitivity of 96% and a specificity of 100% for preoperative biopsy [39]; they concluded that preoperative percutaneous biopsy is essential in the investigation for solid presacral tumours to guide management. Preoperative biopsy may also be useful to exclude other possible lesions such as lymphoma, gastrointestinal stromal tumour (GIST) and metastatic deposits. An alternative to preoperative biopsy is to perform an intra-operative frozen section [40].

Onco-fetoprotein

A high CA19-9 level may be present with a presacral lesion, but this does not distinguish a benign cyst from a malignant tumour [41]. There is no clear evidence for the use of onco-fetoproteins in the diagnosis and management of presacral tumours.

Best imaging modality

CT and MRI have an essential role in the assessment of retrorectal tumours [42]. MRI is the more useful [24,43,44] and axial images are the most useful in determining involvement of the pelvic side wall (Fig. 1).
Table 1 Reports on presacral tumours 2011–2015.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Patients</th>
<th>Histopathology</th>
<th>Gender</th>
<th>Benign, malignant</th>
<th>Biopsy</th>
<th>Accuracy of biopsy</th>
<th>Recurrence at biopsy tract</th>
<th>Accuracy of imaging</th>
<th>Recurrence</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hopper et al. [71]</td>
<td>69</td>
<td>Variable</td>
<td>F 42, M 27</td>
<td>B 49, M 20</td>
<td>16</td>
<td>15/16</td>
<td>0/16</td>
<td>MRI 17/18, CT 9/14, other 1/1</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Mitsuyama et al. [32]</td>
<td>1</td>
<td>Neuroendocrine tumour</td>
<td>F 0, M 1</td>
<td>B 0, M 1</td>
<td>1</td>
<td>0/1</td>
<td>0/1</td>
<td>n/a</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Simpson et al. [17]</td>
<td>26</td>
<td>Sacrococcygeal teratomas</td>
<td>F 19, M 7</td>
<td>B 21, M 5</td>
<td>26</td>
<td>26/26</td>
<td>0/1</td>
<td>n/a</td>
<td>3/26</td>
<td>3 recurrences in malignant group, all died</td>
</tr>
<tr>
<td>Sagar et al. [33]</td>
<td>15</td>
<td>Recurrent retrorectal tumours</td>
<td>F 11, M 4</td>
<td>B 13, M 2</td>
<td>Prior histology</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>0/15</td>
<td>0 recurrences</td>
</tr>
<tr>
<td>Sagar et al. [8]</td>
<td>76</td>
<td>Variable</td>
<td>F 50, M 26</td>
<td>B 60, M 16</td>
<td>22</td>
<td>19/22</td>
<td>0/22; 1 had chronic fistula</td>
<td>72/76 (64/76 for definitive diagnosis)*</td>
<td>n/a</td>
<td>74 MRI, 2 CT</td>
</tr>
<tr>
<td>Oh et al. [66]</td>
<td>9</td>
<td>7 Schwannomas, 1 meningioma, 1 neurofibroma</td>
<td>F 4, M 5</td>
<td>B 8, M 1</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>5 robotic procedures</td>
</tr>
<tr>
<td>Mengual-Ballester et al. [18]</td>
<td>1</td>
<td>Cystic teratoma</td>
<td>F 0, M 1</td>
<td>B 1, M 0</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Kim et al. [35]</td>
<td>1</td>
<td>Tailgut carcinoid</td>
<td>F 0, M 1</td>
<td>B 0, M 1</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>0/1</td>
<td>n/a</td>
<td>Carcinoid in tailgut cyst</td>
</tr>
<tr>
<td>Imboden et al. [10]</td>
<td>1</td>
<td>Epidermoid cyst</td>
<td>F 0, M 1</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Gascon Hove et al. [65]</td>
<td>1</td>
<td>Schwannoma</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td>Single-port surgery</td>
</tr>
<tr>
<td>Fong et al. [60]</td>
<td>10</td>
<td>5 Schwannomas, 2 ganglioneuromas, 1 epidermoid, 6 capillary haemangioma</td>
<td>F 7, M 3</td>
<td>B 10, M 0</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
<td>10/10</td>
<td>n/a</td>
<td>All laparoscopic surgery</td>
</tr>
<tr>
<td>Duck et al. [13]</td>
<td>6</td>
<td>5 Tailgut cysts, 1 with SCC, duplication cyst</td>
<td>F 6, M 0</td>
<td>B 5, M 1</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
<td>5/6</td>
<td>n/a</td>
<td>All TEMS, 1 unexpected malignancy SCC</td>
</tr>
<tr>
<td>Duck et al. [63]</td>
<td>12</td>
<td>4 Tailgut cysts, 2 schwannomas, 2 teratomas, 2 sarcomas, 1 epidermoid, 1 lipoma</td>
<td>F 12, M 0</td>
<td>B 10, M 2</td>
<td>3</td>
<td>2/3</td>
<td>n/a</td>
<td>10/12</td>
<td>0/12</td>
<td>All laparoscopic surgery, 2 unexpected malignancies</td>
</tr>
<tr>
<td>Charalampakis et al. [56]</td>
<td>1</td>
<td>Tailgut carcinoid</td>
<td>F 0, M 1</td>
<td>B 0, M 1</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
<td>0/1</td>
<td>n/a</td>
<td>MRI characteristics benign, carcinoid on final histopathology</td>
</tr>
<tr>
<td>Authors</td>
<td>Patients</td>
<td>Histopathology</td>
<td>Gender</td>
<td>Benign, malignant</td>
<td>Biopsy</td>
<td>Accuracy of biopsy</td>
<td>Recurrence at biopsy tract</td>
<td>Accuracy of imaging</td>
<td>Recurrence</td>
<td>Notes</td>
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<td>------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Nedecu et al. [64]</td>
<td>9</td>
<td>Schwannomas, ganglioneuroma, tailgut cysts, meningocoele, paraganglioma</td>
<td>F 7, M 2</td>
<td>B 9, M 0</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
<td>9/9</td>
<td>0/9</td>
<td>All laparoscopic surgery, one conversion for tumour &gt; 7 cm</td>
</tr>
<tr>
<td>Misawa et al. [20]</td>
<td>1</td>
<td>Neuroendocrine tumour</td>
<td>F 1, M 0</td>
<td>B 0, M 1</td>
<td>1</td>
<td>0/1</td>
<td>0/1</td>
<td>n/a</td>
<td>n/a</td>
<td>Aspiration suggestive of carcinoma, final histopathology neuroendocrine tumour</td>
</tr>
<tr>
<td>Messick et al. [23]</td>
<td>87</td>
<td>Variable</td>
<td>F 67, M 20</td>
<td>B 64, M 23</td>
<td>24</td>
<td>24/24</td>
<td>0/24</td>
<td>n/a</td>
<td>7/23</td>
<td>MRI 39/39, CT 22/31, EUS 7/12</td>
</tr>
<tr>
<td>Merchena et al. [39]</td>
<td>73</td>
<td>Variable</td>
<td>F 37, M 36</td>
<td>B 39, M 30, I 4</td>
<td>76</td>
<td>63/76</td>
<td>0/76</td>
<td>63/69 (25/69 for definitive diagnosis)*</td>
<td>n/a</td>
<td>56 percutaneous, 14 open, 3 both biopsies</td>
</tr>
<tr>
<td>Kesici et al. [82]</td>
<td>1</td>
<td>Epidermoid cyst</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>Fluorine-18 FDG avid ganglioneuroma</td>
</tr>
<tr>
<td>Johnson et al. [19]</td>
<td>1</td>
<td>Dermoid cyst</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>Fluorine-18 FDG avid ganglioneuroma</td>
</tr>
<tr>
<td>Holz et al. [84]</td>
<td>1</td>
<td>Tailgut cyst</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>Fluorine-18 FDG avid ganglioneuroma</td>
</tr>
<tr>
<td>Hebert-Blouin et al. [53]</td>
<td>17</td>
<td>12 Schwannomas, 5 neurofibromas</td>
<td>F 14, M 3</td>
<td>B 17, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>Fluorine-18 FDG avid ganglioneuroma</td>
</tr>
<tr>
<td>Damato et al. [50]</td>
<td>1</td>
<td>Tailgut carcinoid</td>
<td>F 1, M 0</td>
<td>B 0, M 1</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td>Fluorine-18 FDG avid ganglioneuroma</td>
</tr>
<tr>
<td>Chereau et al. [24]</td>
<td>47</td>
<td>Variable</td>
<td>F 34, M 13</td>
<td>B 38, M 9</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>MRI 39/39, CT 22/31, EUS 7/12</td>
<td>n/a</td>
<td>MRI for retrorectal tumours with 100% accuracy</td>
</tr>
<tr>
<td>Angelini and Ruggieri [58]</td>
<td>13</td>
<td>Variable</td>
<td>F 4, M 9</td>
<td>B 0, M 13</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>4/13</td>
<td>Modified Osaka technique (posterior only approach)</td>
</tr>
<tr>
<td>Akbulut [38]</td>
<td>1</td>
<td>Tailgut cyst</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>Literature review 47/332 tailgut cyst with malignant transformation</td>
</tr>
<tr>
<td>Authors</td>
<td>Patients</td>
<td>Histopathology</td>
<td>Gender</td>
<td>Biopsy</td>
<td>Accuracy of biopsy</td>
<td>Recurrence at biopsy tract</td>
<td>Accuracy of imaging</td>
<td>Recurrence</td>
<td>Notes</td>
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<tr>
<td>Syed et al. [85]</td>
<td>91; 71 diagnostic cases</td>
<td>Variable, 37 metastatic</td>
<td>F 45, M 46</td>
<td>B 23, M 48</td>
<td>71</td>
<td>69/71 n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
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<tr>
<td>Sekaran and Brindley [26]</td>
<td>1</td>
<td>Teratoma</td>
<td>F 0, M 1</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td></td>
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<tr>
<td>Samarasekoon et al. [14]</td>
<td>1</td>
<td>Schwannoma</td>
<td>F 0, M 1</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
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<tr>
<td>Petrovic et al. [86]</td>
<td>1</td>
<td>Schwannoma</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Paul et al. [87]</td>
<td>1</td>
<td>Neurofibroma</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>0/1</td>
<td>n/a</td>
<td></td>
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<tr>
<td>Macafee et al. [44]</td>
<td>56</td>
<td>Variable</td>
<td>F 37, M 19</td>
<td>B 39, M 17</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>2/17 (malignant)</td>
<td>0/39</td>
<td></td>
</tr>
<tr>
<td>Huang and Cheng [88]</td>
<td>1</td>
<td>Parachordoma</td>
<td>F 1, M 0</td>
<td>B 0, M 1</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Du et al. [72]</td>
<td>93</td>
<td>Variable</td>
<td>F 61, M 32</td>
<td>B 72, M 21</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
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<tr>
<td>Bosca et al. [89]</td>
<td>40</td>
<td>Variable</td>
<td>F 25, M 15</td>
<td>B 17, M 23</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>8/9 chordomas recurred, 9/23 (malignant)</td>
<td>2/37</td>
<td></td>
</tr>
<tr>
<td>Lin et al. [15]</td>
<td>1</td>
<td>Teratoma</td>
<td>F 1, M 0</td>
<td>B 1, M 0</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>1/1</td>
<td>n/a</td>
<td></td>
</tr>
<tr>
<td>Lin et al. [5]</td>
<td>62</td>
<td>Variable</td>
<td>F 39, M 23</td>
<td>B 48, M 14</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>7/45</td>
<td></td>
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<tr>
<td>Li et al. [79]</td>
<td>33</td>
<td>Variable</td>
<td>F 13, M 20</td>
<td>B 29, M 4</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>2/33</td>
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<tr>
<td>Kye et al. [90]</td>
<td>15</td>
<td>Variable</td>
<td>F 13, M 2</td>
<td>B 14, M 1</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
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</tr>
<tr>
<td>Dozois et al. [47]</td>
<td>37</td>
<td>Sarcoma</td>
<td>F 17, M 20</td>
<td>B 0, M 37</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>23/37</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bartels et al. [27]</td>
<td>17</td>
<td>Congenital presacral tumours, majority teratomas</td>
<td>F 13, M 4</td>
<td>B 3, M 14</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td>n/a</td>
<td></td>
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</tr>
<tr>
<td>932</td>
<td></td>
<td></td>
<td>F 589 (63.2%), M 343 (36.8%)</td>
<td>B 601 (65.9%), M 307 (33.7%), 14</td>
<td>240</td>
<td>219/240 (91.3%)</td>
<td>0/141 (0.0%)</td>
<td>275/312 (88.1%)</td>
<td>72/365 (19.7%)</td>
<td></td>
</tr>
</tbody>
</table>

F, female; M, male; B, benign; M, malignant; I, indeterminate; SCC, squamous cell carcinoma; FDG, fluorodeoxyglucose; FNA, fine needle aspiration.

*Accuracy significantly less for definitive diagnosis (89/145; 61.4%).
T1, T2 and fat saturated T1 images help to determine the internal composition of the tumour, whether cystic, solid, fatty or mixed.

Features that predict a malignant lesion include heterogeneous signal intensity, irregular infiltrative margin, sacral destruction or remodelling and enhancement. Ossseous and neurogenic tumours usually remodel or destroy the sacrum (Fig. 2), neurofibromas have a target sign on MRI and haemangiomas have an increased T1 signal on MRI [7]. Malignant lesions have the MRI characteristics of heterogeneity, irregular margins, solid appearance, low T1 and high T2 intensity and gadolinium enhancement. They are usually larger and extend above S3 [24]. Spinal cord tethering has also been described as a sign of malignancy [32]. Merchea et al. [39] have suggested that the specificity and sensitivity for predicting malignancy in preoperative imaging for retrorectal tumours is 81% and 83% respectively.

A sagittal T2-weighted image is commonly used to determine the upper level of the sacrum reached by the tumour. This aids surgical planning, with the middle of S3 being the main landmark [8]. This is because higher tumours are considerably more difficult to resect via a posterior perineal approach as the sacrum limits the surgical exposure from the perineum. Recently, the sacrococcygeal sinus angle (SSA), an angle formed by a tangential line from the anterior surface of the S1 vertebra and a line drawn from the sacral promontory to the coccyx, has been proposed as the best landmark to help with determining whether the best approach would be anterior or posterior. Kaplan et al. [45] showed that children have a 20° smaller angle which favours a posterior approach.

**Examination under anaesthesia and flexible sigmoidoscopy**

A flexible sigmoidoscopy is essential to exclude a primary rectal cancer and to determine if the presacral tumour has invaded into the rectum. Transrectal ultra-
sound with or without biopsy has also been described in case reports. Although MRI has good accuracy in establishing the diagnosis, retrorectal tumours are rare and may present with non-specific symptoms. An examination under anaesthetic is often warranted. In asymptomatic patients incidentally diagnosed with a retrorectal tumour on laparoscopy or imaging, a flexible sigmoidoscopy is performed not only to appreciate the bulge sign (Fig. 3), which may be subtle, but also to assist in determining rectal tethering or involvement and local invasion by the tumour. While MRI can adequately assess tissue planes and depth of invasion, examination under anaesthetic and flexible sigmoidoscopy may clearly demonstrate rectal involvement and thereby determine the surgical strategy.

**Neoadjuvant and adjuvant therapy**

There is a definite role for neoadjuvant imatinib in the management of GIST and of chemoradiotherapy for lymphomas. There is some evidence that adjuvant treatment may have a useful effect in sarcoma, but the prognosis is poor. Because of its rarity, there are no randomized controlled trials or high level of evidence for neoadjuvant therapy. Some groups including Neale [46] advocate neoadjuvant therapy to reduce the chance of local recurrence, but the evidence is poor.

There is also very limited evidence for the value of adjuvant therapy. In a study of adjuvant therapy for 17 retrorectal sarcomas, there was no statistically significant improvement in disease-free survival or overall survival for patients who received surgery with adjuvant chemotherapy, radiotherapy or chemoradiotherapy over surgery alone [47]. Similarly, myelipomas are frequently resistant to chemoradiotherapy [48]. There is some evidence that aggressive chemoradiotherapy for osteosarcoma and Ewing’s sarcoma may be beneficial, although this is controversial [49]. Adjuvant radiotherapy for an incompletely excised locally invasive presacral tumour may defer but not prevent recurrence. As most patients with a presacral tumour are women with a median age of 45, the evidence for benefit for neoadjuvant or adjuvant therapy must be shown clearly before it is adopted as standard practice due to its impact on fertility [50].

**Surgical techniques and strategies**

The incidental retrorectal cyst or mass

If a retrorectal cyst or mass is diagnosed incidentally during laparoscopy, an attempt should not be made at immediate resection. It is vital to determine whether it is solid, cystic, mixed or fatty, and, as previously discussed, to assess its extent in relation to S3 and the SSA to determine the surgical approach. The retrorectal mass may simply be a common Tarlov perineural cyst, whose management is entirely different. Resection should only be attempted after adequate imaging. If a Tarlov cyst is suspected, myelography should be performed [10].

**En bloc resection for malignant tumours**

Because revisional surgery for retrorectal tumour is difficult and not optimal, *en bloc* resection for malignant tumours with the aim of achieving an R0 resection should be the strategy wherever possible. Simultaneous coccygectomy, sacral resection, rectal resection [51] and vaginal resection [52] may be required to achieve this. For benign tumours, adjacent organs do not have to be resected even for recurrent benign tumours. Resection for presacral sarcoma, which is usually locally invasive, is associated with significant morbidity and mortality. Despite the aim of achieving an R0 resection, Dozois *et al.* [47] showed no statistically significant difference in disease-free survival between R0 and R1 resections. The prognosis is poor even with an R0 resection.

**Nerve-sparing resection for benign tumours**

A nerve-sparing technique should be used for benign tumours because it preserves postoperative urogenital, anorectal and lower-extremity neurological function [53]. While local recurrence of benign tumours can occur with incomplete excision, Pongsthorn *et al.* [54] reported only one recurrence after resection of giant presacral schwannoma. In their series, three patients had subtotal piecemeal excision and two had a partial exci-
cision. None of the patients who had a piecemeal excision developed recurrence and only one patient having a partial excision had recurrence which was removed on a second operation. Importantly, none of the patients had pain or obvious neurological deficit at the final follow-up.

Preoperative vascular embolization
Preoperative vascular embolization may be useful for vascular lesions such as gastrointestinal tumours [55]. Hosaka et al. [56] reported resection of a giant schwannoma more than 10 cm in diameter without a blood transfusion after embolizing vessels feeding the tumour 2 days before resection. There is minimal information on the role of preoperative embolization for retrorectal tumours. Risks of embolization include tumour necrosis, sepsis and complications related to angio-embolization. Further studies are required to determine the safety and role of vascular embolization.

Anterior abdominal approach
An abdominal approach is recommended for tumours above the middle of S3, or if preoperative investigation has indicated pelvic or pelvic side wall involvement. Ureteric stenting, early control of iliac vessels and an en bloc approach to malignant lesions or benign lesions with no obvious plane of dissection is useful [33]. The anterior approach may be intraperitoneal or extraperitoneal. Senoglu et al. [57] described an anterior extraperitoneal approach for resection of a presacral schwannoma.

Posterior Kraske approach
A posterior (Kraske) approach (Fig. 4) is indicated for tumours below S3 without any involvement of the sacrum or side wall of the pelvis and not involving other viscera [44]. It is also more likely to be successful in patients with a small SSA as can be demonstrated by MRI in younger children [45]. The patient is placed in the prone jack-knife position and a midline or parasagittal incision is made with excision or elevation of the coccyx and division of the levators to provide adequate access to the tumour. If benign, the mass should be dissected from the rectum. A finger in the anal canal and rectum can help with this but, if there is no plane, an en bloc resection including removal of the rectal wall may be required. A leak test with methylene blue or water is important to ensure that there is no leak. If the tumour is malignant, the rectum should be resected en bloc with the tumour. Chereau et al. showed that the Kraske approach has a low morbidity and good oncological outcome. In 42 patients of whom 25 (60%) had resection of the coccyx, there was minimal operative morbidity and a good 5-year disease-free survival. Only one patient required reoperation for haematoma and overall four patients developed complications (two haematomas and two abscesses). Overall, the Kraske procedure has been reported with low morbidity and good oncological outcome [6,24].

Posterior modified Osaka approach
Angelini and Ruggieri [58] proposed a new posterior-only technique for en bloc resection of malignant retrorectal tumours which they named the modified Osaka technique. This allows en bloc excision of sacral tumours with protection of the nerve roots. The technique involves a posterior midline incision followed by osteotomies performed laterally by a threadwire saw and Kerrison rongeurs through the sacral foramina to avoid sacral root damage. The lateral portion of the sacrum is then mobilized to achieve adequate access to the presacral structures without damage to the sacral nerve roots. At a 3 year follow-up, 69% (9/13) remained disease free. The advantage of this technique is that it allows resection of tumours proximal to S3, protects the sacral roots and may be associated with decreased blood loss and operation time.

Combined approach
Malignant lesions requiring en bloc resection of adjacent organs often require a combined abdominal and per-
incal approach. The operation time is greater for a combined approach and repositioning the patient if required can be difficult. It has been shown, however, that it is not always necessary to reposition the patient to the prone jack-knife position, since it is feasible to do both abdominal and perineal approach with the patient in the modified Lloyd Davies position [59].

**Other surgical considerations**

**Laparoscopic vs open**

Laparoscopic removal has been shown to be safe for resection of retrorectal lesions, with no increase in morbidity or intra-operative complications [10,60–62]. Duclos et al. showed that while it is feasible to perform laparoscopic resection of presacral tumours, it is not without risk [63]. In their series of 12 patients, two procedures were converted to open, one patient had incomplete resection (R1) and required reoperation with a laparotomy to completely remove the tumour and one patient needed a temporary diverting ileostomy for rectal injury during the laparoscopic procedure. The main predictors of open conversion include huge retrorectal tumours, obesity, narrow pelvis and high ASA. In Nedelcu et al.’s [64] series of nine laparoscopic excisions, only one patient with a tumour greater than 7 cm in diameter required open conversion.

**Single-port laparoscopic surgery**

There is limited information on single-port surgery for retrorectal tumours. A recent case report on the use of single-port access laparoscopic surgery for excising a presacral schwannoma showed that the technique was feasible [65].

**Robotic surgery**

There are reports evaluating the role of robotic surgery for the removal of presacral tumours. These have shown that robotic surgery is useful for patients with large presacral tumours over 10 cm in diameter. In a case series of nine patients presented by Oh et al. [66], robotic rectal dissection in five resulted in shorter length of hospitalization, bleeding and operation time compared with open resection in four cases.

**Transanal endoscopic microsurgery (TEMS)**

TEMS may be safe for the resection of retrorectal benign tumours [13,67] but information in the literature is limited. Approaching a retrorectal tumour through the rectal wall does not follow oncological principles for malignant tumours. The concern is that it may be difficult to exclude malignant degeneration of a benign appearing cyst. It has been argued by some that even with careful preoperative assessment malignancy cannot be completely excluded [8,32,33,35]. This was highlighted in the TEMS series presented by Duck et al. [13], where a patient having TEMS was initially thought to have benign disease but had squamous cell cancer within the cyst wall on final pathology. On reviewing the preoperative assessment retrospectively, abnormalities of the wall of the lesion and solid component were apparent on imaging. Careful preoperative evaluation with a high index of suspicion for malignancy is required as TEMS should not be performed for malignant tumours. The mean tumour size reported in TEMS case series has been small (3–4 cm) and the maximum distance from the anal verge approximately 8 cm. Whether larger retrorectal tumours that are further away from the anal verge can be resected by TEMS needs further evaluation.

**Reconstruction**

Simultaneous or staged pedicle or free flap transfers after extensive soft tissue dissection in the retrorectal space may avoid chronic sinus formation, fistulation and mucous secretion [68]. Simultaneous use of Permacol mesh to reconstruct the posterior pelvic wall has also been described [69]. Staged flap transfers may provide more oncological safety pending full histological analysis and pathological evaluation. Reconstruction is usually performed as a staged procedure by an oncoplastic surgeon.

**Discussion**

This review of the recent literature on presacral tumours assessed the pathology, the role of preoperative biopsy, various imaging modalities, operative approaches, complications and the role of neoadjuvant and adjuvant therapy. The quality of studies presented was poor due to the rarity of presacral tumours.

The current study showed that retrorectal tumours are more likely to be benign. Cystic lesions are mostly benign, except in cases where solid wall or heterogeneous components are demonstrated on imaging [13,36]. A preoperative biopsy should not be performed for a cystic retrorectal lesion. This is not only because most are benign, but also because biopsies of cystic lesions are inaccurate and malignancy may be present even if not detected on biopsy. Wall abnormalities, solid components or heterogeneity on MRI should raise the suspicion of malignancy. Although there are concerns with the risk of seeding during a preoperative biopsy, the studies assessed in this review showed no case of tumour within the biopsy track. A transgluteal, transperineal or transsacral approach
where the biopsy track is tattooed and is later resected with the tumour reduces the risk of biopsy induced recurrence.

The review found that a preoperative biopsy should be performed for solid tumours. Confirming a definitive diagnosis of malignancy should result in an aggressive en bloc resection. A biopsy for solid retrorectal tumours reported to be benign has a low false negative rate. A negative biopsy is valuable preoperative information, and is helpful when obtaining informed consent. Furthermore, it may guide neoadjuvant therapy and may direct management to a non-operative approach in the case of a lymphoma or palliative treatment in the case of metastatic disease.

MRI has a greater sensitivity and specificity than transrectal ultrasound and CT. While CT may diagnose an abnormality in the presacral region, it cannot distinguish between benign and malignant tumours as accurately as MRI. CT is useful, however, as a preliminary investigation for non-specific abdominal symptoms. Chereau et al. [24] recently demonstrated 100% accuracy with MRI and reported significantly lower accuracy with other imaging. MRI, however, is not as accurate as preoperative biopsy in providing a histopathological diagnosis.

Most studies indicate that surgery is nearly always indicated when a retrorectal tumour is diagnosed, but patients with metastatic disease, lymphoma and those with a benign tumour who are too frail may avoid surgery. There are reports of successful non-operative treatment of diffuse large B-cell lymphoma with CHOP (cyclophosphamide, hydroxydaunorubicin, oncovin, prednisolone), rituximab and radiotherapy [70]. Patients with metastatic disease can be referred to a palliative route, with surgery for a defunctioning stoma if the retrorectal tumour is associated with a large bowel obstruction. Although recent studies such as that by Hopper et al. [71] have demonstrated that cystic lesions may be managed non-operatively, the present review suggests that provided the patient is fit for surgery benign lesions should be resected. This is because a benign lesion can be complicated by infection and cause compression of the presacral nerves or obstructive symptoms and vague abdominal discomfort. Benign lesions may contain malignancy, particularly teratomas which are well known to degenerate. There have also been a significant number of reports in the literature of cystic lesions harbouring malignancy. Benign lesions may also continue to grow and symptoms of local compression may worsen. Delaying surgery may increase the difficulty of subsequent removal. S3 is a very useful landmark to decide on an abdominal or a posterior approach, and the SSA [45] is an interesting concept deserving further consideration. The authors demonstrated that younger patients had a significantly narrower SSA (approximately 20°), were more amenable to the posterior approach and required less sacral resection.

The review has identified small case series of robotic surgery, single-port surgery and TEMS in the management of presacral tumours. The benefit is not clear. Robotic surgery and single-port surgery are feasible, but the learning curve may be steep and operation time significantly longer than conventional laparoscopic surgery. The use of TEMS is controversial as it is not oncologically safe and even benign appearing retrorectal lesions with benign biopsies have been reported as malignancy on final histopathology [13]. While Duck et al. did not report any increase in short-term recurrence rates for TEMS, the lack of long-term follow-up and the limited cohort of patients assessed were inadequate to determine the safety of TEMS for presacral tumours. The review has identified one case of preoperative angiembolization for a presacral giant schwannoma. This procedure has not been adequately assessed. It is not currently recommended but it may be useful for vascular or large tumours. Further studies are needed to determine its safety and efficacy.

Overall, the postoperative complication rate of surgery for retrorectal tumours was around 20%, including neurological and non-neurological complications such as bleeding, pelvic haematoma, sepsis and bowel injury [72]. There are not many data on bladder and bowel dysfunction after surgery in the presacral space [73]. It has been shown, however, that when sacral nerves are divided, a high rate of urinary and bowel dysfunction ensues, with over 20% of patients developing both bladder and bowel dysfunction and nearly 50% bladder dysfunction alone — much higher than the rates reported for the entire group (15% bladder dysfunction, 7% anal incontinence) [74]. There may be some evidence that the rate of bladder and bowel dysfunction is lower in laparoscopic excision or removal via a posterior approach [24,64,75].

The recurrence rate for malignant presacral tumours in the literature is approximately 30% and for benign lesions the rate is significantly lower [23]. Benign tumours usually recur locally whereas malignant tumours recur locally and in addition often metastasize. Presacral tumours can metastasize to internal iliac nodes through the neurovascular lymphatic space, bypassing the external iliac nodes [76]. Tumour metastases have been reported to gluteal muscle [77], liver [78], lung [79], brain [80], and malignant recurrences and metastases usually result in death whereas benign recurrences are usually associated with a good prognosis after
repeated excision [23,24]. The prognosis has been shown to be significantly worse with presacral sarcomas and recurrent malignant teratomas.

Neoadjuvant therapy is currently given for retrorectal GIST, Ewing’s sarcoma, osteosarcoma and desmoid type fibromatosis. The indication for chemotherapy after resection depends on the histology, invasiveness, pathology and grade of the tumour. The main advantage of using imatinib for GIST in a neoadjuvant instead of an adjuvant setting is rapid tumour shrinkage which may facilitate a sphincter preserving operation. Both neoadjuvant and adjuvant therapy for GIST with imatinib has been shown to reduce the risk of recurrence, as even after a macroscopically complete resection with negative margins there is a significant recurrence risk without imatinib. There are reports of variable radiological appearance of GIST which can be distinguished on biopsy [39,81].

There are many limitations to this review. Being rare, most of the studies on presacral tumours assessed were retrospective and most were cohort studies, case series or case reports. Most were poor in quality but were included on the premise that case reports and case series are never consensus guidelines. Nevertheless, some guidance towards a coherent, logical strategy and algorithm which improves the understanding and management of presacral tumours.

The diagnosis, investigation and surgery for presacral tumours are complex. MRI is the most useful imaging modality to differentiate benign from malignant tumours. Preoperative biopsy is safe, useful and feasible for solid presacral but not for cystic tumours. Patients with a benign appearing tumour should undergo a nerve-sparing surgery approach whereas malignant tumours should have an en bloc resection. There is no consensus guideline for neoadjuvant and adjuvant therapy, but it has been shown to be beneficial in retrorectal GIST, various forms of sarcoma and desmoid type fibromatosis. S3 is the main landmark in deciding between anterior or posterior surgical approaches with SSA as a useful adjunct. The use of TEMS for presacral tumours is controversial due to concerns that benign appearing tumours may harbour malignancy. Robotic and single-port surgery is feasible, but the benefit is currently unclear.

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J. W. T. Toh & M. Morgan

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