Fatal coronary sinus thrombosis due to hypercoagulability in Crohn's disease

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Original Article

A 27-year-old male, admitted to hospital for a psoas muscle abscess secondary to Crohn's disease, suddenly died while in hospital. A medicolegal autopsy showed coronary sinus thrombosis. Coronary sinus thrombosis is rare and is usually associated with invasive cardiac procedures. Coronary sinus thrombosis associated with Crohn's disease has not been reported. Autopsy examination of the coronary sinus is advocated, especially in individuals with hypercoagulable states.

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1. Introduction

Coronary sinus thrombosis is a rare and a highly lethal medical event. Previous cases are most frequently associated with cardiac procedures particularly those involving the right heart including central venous catheterization, cannulation during cardiopulmonary bypass, pacemaker wire placement, electrophysiologic ablation for supraventricular tachycardia, ventriculostial shunt insertion for hydrocephalus, and cardiac surgery. Previous reports of noniatrogenic coronary sinus thrombosis have been documented in association with preexisting disease processes including atrial fibrillation, tricuspid regurgitation, chronic obstructive pulmonary disease, chronic cor pulmonale due to pulmonary fibrosis, Kawasaki disease, and acute lymphoblastic leukemia. We present a case of a 27-year-old male hospitalized for a psoas muscle abscess secondary to Crohn's disease, who developed spontaneous, lethal coronary sinus thrombosis.

2. Case description

This 27-year-old male, who had a medical history significant for Crohn's disease, was admitted to hospital with abdominal pain, nausea, vomiting, diarrhea, and weight loss. A psoas muscle abscess was diagnosed and was treated with an in situ drain and antibiotics. Additional medications in hospital included azathioprine, infliximab, prednisone, and dalteparin. He experienced right leg pain, and an ultrasound demonstrated partial thrombosis of the right popliteal and trifurcating veins. He improved in the hospital, and plans for hospital discharge were made. On the date of his discharge, he was well, with no voiced complaints. Two hours later, he was found unresponsive in his hospital bed by the nurse. Despite cardiopulmonary resuscitation and administration of a thrombolytic, he died. A medicolegal autopsy was requested.

At autopsy, the decedent was a thin male weighing 68 kg and measuring 190 cm in height. His body mass index was 18.8 kg/m². Therapeutic intervention included an endotracheal tube, defibrillator pads, electrocardiogram leads, and intravenous lines. The heart weighed 420 g, with right-dominant circulation. The coronary arteries had normal course and normal origin, without significant atherosclerosis. The coronary sinus was grossly dilated and filled with thrombus measuring 2.0×0.5×0.5 cm (Fig. 1). The thrombus did not extend into the coronary sinus ostium or into the right atrium. Histology demonstrated an acute antemortem thrombus with layered red blood cells, platelets, and fibrin (Fig. 2A, B). No thrombus organization was seen.

The right psoas muscle abscess was confirmed grossly and microscopically. No malignancy was present. Toxicology was negative.
3. Discussion

Coronary sinus thrombosis in the absence of instrumentation or manipulation of right heart structures is exceedingly rare. In this case, coronary sinus thrombosis is associated with Crohn’s disease. The decedent’s suspected cause of death was pulmonary thromboembolism; coronary sinus thrombosis was an unexpected finding.

The mechanisms of coronary sinus thrombosis have been postulated to be the same for those associated with thrombosis at other sites including endothelial damage, hypercoaguability, and venous stasis [1,3]. In this case, there was no cardiac instrumentation (i.e., no iatrogenic endothelial damage). An acquired hypercoagulable state is most likely. Individuals with inflammatory bowel disease (i.e., Crohn's disease and ulcerative colitis) are at increased risk of venous and arterial thromboses and thromboembolic events due to fluid depletion, prolonged immobilization, surgery, steroid therapy, hyperhomocysteinemia, increased levels of certain coagulation factors such as fibrinogen, prothrombin and factor V, impairment of fibrinolytic system factors, endothelial dysfunction, and abnormalities of platelets [17,18]. Coronary sinus thrombosis has not been previously described in the literature in association with inflammatory bowel disease. There was no reported family history of primary thrombophilia. Infection (i.e., psoas muscle abscess) and prolonged immobilization may have been additional risk factors. His hypercoaguable state is corroborated by the partial thrombosis of the right leg veins.

Clinically, coronary sinus thrombosis may present with chest pain, shortness of breath, hypotension, electrocardiogram changes, pericardial effusion, cardiogenic shock, or sudden cardiac death [2,8]. Some individuals are asymptomatic. Mortality is high due to the nonspecific clinical presentation, rapidity of clinical deterioration, and rarity of the condition [8]. Acute occlusion of the coronary sinus leads to ischemia and eventual infarction of the myocardium. This may precipitate a cardiac arrhythmia and sudden death [1,2,6,9]. Partial or incomplete occlusion of the coronary sinus by chronic thromboses may not cause immediate death due to the formation of adequate collateral circulation [1,9].

The right ventricle demonstrated minute foci of myocyte necrosis with associated rare inflammatory cells. The differential diagnosis includes myocarditis or ischemic changes. Myocarditis may precipitate cardiac arrhythmias and sudden death. However, myocarditis is also a well-documented incidental finding at autopsy [19,20]. In this case, the coronary sinus thrombosis is the more compelling cause of death.

4. Conclusions

Coronary sinus thrombosis should be included in the differential diagnosis of sudden unexpected death in individuals with hypercoagulable states including those with inflammatory bowel disease. Coronary sinus examination may be neglected during postmortem examination. This case proves the necessity of meticulous examination and sampling of the coronary sinus to diagnosis this rare cause of sudden cardiac death.

Fig. 1. Posterior surface of the heart showing the coronary sinus distended by thrombus.

Fig. 2. Acute coronary sinus thrombosis. (A) Hematoxylin and eosin stain. The coronary sinus wall is indicated by the arrow. (B) Martius Scarlet Blue stain for fibrin. On the color image, fibrin is red (star), erythrocytes are yellow, and connective tissue stains blue. The coronary sinus wall is indicated by the arrow.
Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Acknowledgments

None.

References
