Chronic occlusion of the thoracic aorta: a novel cause of pleuroperticardial effusions and pancytopenia

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ABSTRACT
A 57-year-old man had presented with a 6-month history of worsening dyspnea, renal failure, hypertension, pancytopenia, and a continuous machinery murmur. Imaging studies revealed pleuroperticardial effusions that recurred despite aspiration and suprarenal mid-thoracic aortic occlusion (AO) with extensive collateral vessels to the chest wall, rectus sheath, and diaphragm. A right axillofemoral bypass transformed his clinical course. The murmurs, renal failure, pleuroperticardial drainage, and pancytopenia resolved, and his hypertension had markedly improved. The association of chronic AO with pleuroperticardial effusions without peripheral edema or ascites was most likely due to increased supradiaphragmatic interstitial pressure, and the bone marrow hypoperfusion likely explains the pancytopenia. In addition to posing diagnostic challenges, chronic AO reveals unique insights into the pathogenesis of pleuroperticardial effusions and pancytopenia. (J Vasc Surg Cases and Innovative Techniques 2021;7:540-4.)

Keywords: Aortic occlusion; Bypass; Chronic; Pancytopenia; Pericardial effusion; Pleural effusion

Chronic aortic occlusion (AO) poses diagnostic challenges owing to its rarity and the diverse constellation of presenting symptoms. We have presented the case of chronic thoracic AO as an unusual cause of recurrent pleuroperticardial effusions, refractory hypertension, and pancytopenia.

CASE REPORT
A 57-year-old man had presented with a 6-month history of exertional dyspnea, progressive renal failure, pancytopenia, and a new murmur. His history included 30 years of hypertension, mild renal impairment (creatinine, 125 μmol/L), and rib fractures after a high-speed motor vehicle accident 30 years previously. He reported a 3-year history of postprandial abdominal pain and peripheral claudication. At presentation, he was hypertensive (190/80 mm Hg), with elevated jugular venous pressure and dull lung bases. A palpable right parasternal thrill was present, with a continuous machinery murmur that was loudest at the left and right second intercostal spaces and softer holo-systolic murmurs evident even at the lung bases. Lower limb pulses were absent. No ascites or peripheral edema was present. Blood tests showed pancytopenia (hemoglobin, 84 g/L; white blood cell count, 3.5 × 10^9/L; platelet count, 113 × 10^9/L) and worsening renal failure (creatinine, 192 μmol/L). The results from hematologic, hemolysis, and autoimmunity studies, urinalysis, and bone marrow biopsy were normal. Chest radiography showed moderate pleural effusions (Fig 1, A). Transthoracic echocardiography revealed a circumferential pericardial effusion (21 mm in depth) without evidence of tamponade (Fig 1, B).

Computed tomography of the chest and abdomen demonstrated descending thoracic AO that was >7 cm in length, with extensive collateral vessels to the posterolateral chest wall, rectus sheath, and around the diaphragm and esophagus (Fig 2, A-E). Also, marked calcified atheromatous disease involving the descending thoracic aorta was present for a distance of ~15 cm involving bilateral renal artery origins. The findings from the vertebral, carotid, and coronary arterial imaging studies were unremarkable.

Because the pericardial and pleural effusions were progressive despite initial drainage, an epigastric surgical pericardial window with biopsy and pleurocentesis and pleural biopsy were performed to exclude infective and neoplastic serosal pathology entities. Biochemistry revealed that both effusions were transudates. The pleural (~2000 mL/day) and pericardial (~400 mL/day) fluid accumulation remained very high after window formation and drain insertion, suggesting an increased transudative gradient into these supradiaphragmatic interstitial cavities to explain the ongoing serosal fluid accumulation.

Chronic AO was diagnosed. A right axillofemoral bypass (8-mm ring Gore Propaten graft; W.L. Gore and Associates, Flagstaff, Ariz) was performed to improve the subdiaphragmatic blood flow, with the primary intention to improve the mesenteric and renal blood flow (Fig 2, F). Surgery immediately transformed his clinical course. His blood pressure improved

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dramatically, with three antihypertensive agents ceased. In addition, his machinery murmurs, renal failure, and pancytopenia had resolved, and the pleural and pericardial drainage had stopped within 48 hours (Figs 3 and 4). At 24 months of follow-up, he was well with only minor renal impairment. The patient provided written informed consent for the report of his case details and imaging studies.

DISCUSSION

Chronic thoracic AO is a rare presentation, with suprarenal occlusion accounting for only 12% of cases. Patients have classically presented with hypertension, renal failure, and claudication. The etiologies include atherosclerotic disease progression with subsequent aortic thrombosis, inflammatory conditions such as Takayasu’s arteritis, trauma, and radiotherapy to the pelvis resulting in progressive arterial wall inflammation and stenosis. The phenomenon of “coral reef aorta” has been described, comprising eccentric heavily calcified atherosclerotic lesions with areas of focal heterotrophic ossification causing aortic obstruction. Our case was unusual because of the mid-thoracic occlusion that had occurred in a patient in middle age, the remarkable improvement in blood pressure, claudication, postprandial abdominal pain, and renal function, and the resolution of pancytopenia and pleuropericardial effusions after bypass of the occlusion. The occlusion was likely chronic, given the presence of the extensive collateral vessels. The underlying etiology has remained unclear but might have been progressive atherosclerosis around sites of previous unrecognized aortic injury after his motor vehicle accident, resulting in calcification, stenosis, and subsequent occlusion. Progressive atherosclerosis alone is another possible etiology, although this tends to occur most often in smokers and those with diabetes and hypercholesterolemia, generally with multilevel infrarenal aortic involvement.

The association of chronic AO with the presence of both pleuropericardial effusions and pancytopenia has not been previously reported, with the almost immediate resolution postoperatively in our patient establishing causation. Persisting transudative effusions can be attributable to increased supradiaphragmatic hydrostatic pressure in the parietal pleuropericardial microvessels and, thus, increased filtration across the serosal membranes. The high cardiac output from the collateral circulation is also likely to have contributed. In contrast to the pleuropericardial effusions described in the present patient, interstitial pulmonary edema has been described in case reports of those with coral reef aortic occlusion or neonatal coarctation. The continuous murmur was generated by the continuous blood flow in the collateral vessels (predominantly dilated intercostal arteries) shunting from the high-pressure aortic circulation to the low-pressure circulation throughout systole and diastole. This has been described previously in patients with severe coarctation (<3 mm) in which the developed collateral circulation was not sufficient to ensure normal flow in diastole. In our patient, radiation of the murmur to the back was present, arising from the extensive collateral circulation to the posterolateral chest wall. Resolution of the continuous murmur in our patient after surgical bypass and the reduction in blood pressure presumably resulted from the loss of the pressure gradient difference. The hypertension and renal failure observed in our patient are readily explained by the renal hypoperfusion.

Pancytopenia in aorto-occlusive disease is, to the best of our knowledge, a novel association and a diagnosis that was carefully reviewed by our consultant hematologists. Although the mechanism is unclear, it might be
attributable to bone marrow hypoperfusion, especially given the prompt resolution after surgery. Additionally, renal failure and anemia secondary to chronic disease might have also been exacerbating factors owing to the release of inflammatory cytokines (eg, interleukin-1β, tumor necrosis factor-α, interferon-β), impaired erythropoiesis due to decreased erythropoietin production and bone marrow responsiveness to erythropoietin.10

However, our patient’s mild to moderate renal impairment would not normally be associated with the degree of anemia seen at presentation. Management will vary depending on the location and severity of the aorto-occlusive disease but should include anatomic or extra-anatomic bypass or endovascular revascularization. Extra-anatomic bypass, as used for our patient, will be performed when general or abdominal
Contraindications to an anatomic bypass are present. Extra-anatomic bypass has been associated with 3-year patency rates of 40% to 80% and mortality rates of 20% to 60%. Endovascular stenting is an emerging approach limited to case series but has been associated with the risk of acute renal ischemia, spinal cord ischemia, distal embolization, side branch jailing, and access site complications. In our patient, the highly calcified stenotic lesion

Fig 3. Graphs showing time course from admission to 6 months after the axillofemoral bypass. A, Systolic blood pressure had improved postoperatively. B, Renal function had improved postoperatively. C, Hemoglobin levels had increased after blood transfusion but had returned to normal levels at 2 years postoperatively. D, Platelet levels had increased postoperatively owing to reactive thrombocytosis and had returned to normal levels at 2 years postoperatively. E, White blood cell count had returned to normal limits postoperatively.
was deemed unsuitable for stenting, especially given the high risk of distal embolization. For our patient, the axillo-femoral approach was considered the option with the least short-term risk. He was considered high risk owing to his multiple comorbidities at the time of surgery. Open thoracic reconstruction was later discussed with the patient, who opted for careful surveillance of his axillo-unifemoral bypass graft as an alternative.

We have presented an unusual case of chronic mid-thoracic occlusion that had presented in a middle-age patient, with widespread multiorgan involvement. Revascularization profoundly improved our patient’s hypertension, claudication, and renal failure, with complete resolution of pancytopenia and serosal effusions, revealing unique insights into the importance of the vascular supply in these pathologic entities.

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