CASE REPORT

Thromboatheromatous coarctation of the aorta diagnosed with intraoperative TOE during emergent open aneurysm clipping

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SUMMARY

A woman in her 50s presenting for emergent intracranial surgery was discovered to have a large incongruity in blood pressure between her right arm and her other extremities. Intraoperative rescue transoesophageal echocardiography (TOE) revealed a large thromboatheromatous burden in her descending aorta resulting in a functional coarctation. Usually diagnosed via CT imaging, we present what we believe to be the first published case diagnosed intraoperatively using TOE. After the diagnosis was made, blood pressure goals were adjusted to provide sufficient perfusion distally and her surgery was completed otherwise uneventfully.

BACKGROUND

Acquired thromboatheromatous coarctation of the aorta, or coral reef aorta, is a rare condition found almost exclusively in hypertensive patients with peripheral arterial disease and tobacco use. Usually diagnosed via CT imaging, we present what we believe to be the first published case diagnosed intraoperatively using transoesophageal echocardiography (TOE). This report highlights the importance of rescue echocardiography whenever unexplained haemodynamic instability is encountered in the operating room and also reviews a rare vascular condition that should be considered whenever discrepancies in extremity blood pressures are encountered.

CASE PRESENTATION

A woman in her 50s with a history of hypertension, tobacco abuse and peripheral arterial disease presented to an outside hospital complaining of the worst headache of her life. Following presentation, she had a syncopal event and subsequent imaging revealed a Fisher grade 4 subarachnoid haemorrhage and an anterior communicating artery aneurysm. She became combative and was intubated and then transferred to our institution for neurosurgical intervention. She presented to the operating room for emergent open aneurysm clipping. Following induction of anaesthesia, no palpable left radial pulse was found and a right radial arterial catheter was inserted. The initial systolic blood pressure from the right arterial line was 200 mm Hg; concurrent non-invasive blood pressure (NIBP) measurement on the left arm revealed a systolic pressure of 120 mm Hg. Her preoperative CT scan showed left

subclavian origin occlusion with later reconstitution so it was assumed that the pressure measured from the right arm represented systemic pressure and that left arm measurements were decreased secondary to subclavian artery disease. However, multiple NIBP measurements taken from both lower extremities were also low with a similar 70–90 mm Hg systolic discrepancy from the right radial pressure. An NIBP cuff was placed on her right arm, and readings correlated with the ipsilateral radial catheter.

The neurosurgical team requested that her systolic blood pressure be maintained below 100 mm Hg to facilitate surgical exposure and clipping of the aneurysm; however, with uncertainty regarding her true systemic pressure, this was felt to be unsafe without further evaluation. Given the incongruity in blood pressure readings between her right arm and the rest of her extremities, aortic coarctation was considered and a TOE probe was inserted to visualise the aorta and any possible vascular pathology. Views of the descending thoracic aorta taken from the midoesophagus showed large calcified plaque that nearly occluded flow (figures 1 and 2). The surgeons were notified and the systolic blood pressure goal was adjusted to 140 mm Hg, resulting in blood pressures of approximately 50/30 mm Hg in her lower extremities and left upper extremity. She remained stable throughout the case, with adequate urine output and no evidence of metabolic acidosis. At the conclusion of the complex aneurysm clipping, she was transferred to the intensive care unit intubated in stable condition.

INVESTIGATIONS

Non-contrast CT scan is the diagnostic test of choice. CT in patients with acquired thromboatheromatous coarctation shows dense, serpiginous, exophytic calcifications of the aortic wall protruding into the aortic lumen as opposed to typical calcifications seen following the curve of the vessel wall.¹² The majority of plaques occur on the posterior surface of the aorta. These lesions cause significant occlusion, most commonly in the juxtarenal or suprarenal aorta.

Coarctation of the aorta can be diagnosed with either transthoracic (TTE) or transoesophageal (TOE) echocardiography combined with Doppler imaging. TOE findings in coral reef aorta are similar to those seen on CT with dense exophytic calcifications of the aorta resembling a coral reef

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Figure 1 Transoesophageal midoesophageal descending aorta short axis view showing dense atheromatous burden of the posterior surface of the descending aorta measuring approximately 1 cm.

found primarily on the posterior wall of the aorta (figure 1). The area of coarctation is identified with colour flow Doppler by the presence of turbulent flow consistent with a narrowing of the lumen (figure 2, video 1). Pulsed wave Doppler is used to assess flow velocities proximal and distal to the coarctation while velocities and gradients across the narrowed portion of the aorta are best evaluated by continuous wave Doppler. Unfortunately, the severity of coarctation is classically underestimated by Doppler techniques due to collateralisation and for precise analysis the exact location, luminal narrowing and presence of collateral vessels should be confirmed by either CT or MRI.² Also, commonly seen on cardiac imaging is concentric left ventricular hypertrophy secondary to remodelling by the left ventricle to adapt to abnormally high afterload.³

DIFFERENTIAL DIAGNOSIS

Aortic coarctation, aortic aneurysm rupture, severe peripheral vascular disease, thromboembolic disease; equipment malfunction.

TREATMENT

Acquired thromboatheromatous coarctation often requires surgical intervention due to a high risk of embolism. It is primarily treated surgically with thromboendarterectomy, resection of thrombus and aortic graft placement or via thoracoabdominal bypass graft, largely determined by the location of the lesion.⁴ Unfortunately, open aortic surgery has a high rate of operative mortality (8.711.6%) and postoperative complications (13.915.9%).⁴ Recent improvements in endovascular techniques have enabled patients to be successfully treated with uncovered, self-expanding or rarely covered stent grafts. Covered stent grafts

have limited utility in this disease secondary to an inability to expand and complications including paraplegia from occlusion of the side branch.⁵ The decision to intervene is highly patient dependent and is often based largely on symptoms, comorbidities and surgical risk. Alternatively, patients not thought to be candidates for surgical intervention may be managed medically with statins, smoking cessation, blood pressure control and possibly anticoagulation.⁶ Medical management should be initiated as soon as the diagnosis is made.

OUTCOME AND FOLLOW-UP

A vascular surgeon was consulted and recommended a CT angiogram, which confirmed our TOE findings. The surgical team ultimately decided to defer addressing her flow obstruction until her acute neurological problems resolved. Unfortunately, her neurological function never improved and she ultimately succumbed after an exploratory laparotomy for mesenteric ischaemia. It is unclear if emboli from her aorta contributed.

DISCUSSION

Coral reef aorta is a dense atherosclerotic disease of the juxta-renal or suprarenal aorta in which heavily calcified plaques grow in the aortic lumen causing significant occlusion and eventually functional coarctation. Occurring in 0.6%–1.8% of the population, the disease shows no gender or ethnic preference and is usually diagnosed around age 50.⁷ Most patients have a history of poorly controlled hypertension, peripheral arterial disease and tobacco use. Common presenting symptoms include neurological symptoms (headache, visual changes, vertigo), renovascular hypertension, claudication from limb malperfusion and peripheral blood pressure discrepancies. In advanced stages,

Figure 2 Transoesophageal midoesophageal descending aorta long axis view at the level of maximal atheromatous disease with colour flow Doppler placed across the lesion. Turbulent flow is visualised around the atheromatous burden.

renal failure, visceral ischaemia, heart failure and ultimately death may occur.^{7,8} Low serum levels of ferritin-A and uncarboxylated matrix gla protein, both inhibitors of calcification, appear to play a role in the disease process.⁷

TOE is a valuable diagnostic tool particularly when unexplained haemodynamic changes occur and TTE is unfeasible. Several centres have established a rescue echocardiography protocol that recommends nine standard views that can be obtained by any provider with basic echocardiography training.⁹

Video 1 Video of the transoesophageal midoesophageal descending aorta long axis view at the level of maximal atheromatous disease with colour flow Doppler placed across the lesion and visualisation of turbulent flow across the atheroma.

Views of the ascending and descending aorta, both in long and short axis are an important component of any rescue TOE examination, as demonstrated in this case where imaging the heart exclusively would not have led to the diagnosis. Additionally, intraoperative TOE is very safe and practical for diagnosis, particularly in patients without risk factors for complications.¹⁰

Learning points

- Acquired thromboatheromatous coarctation, or coral reef aorta, is a rare cause of functional coarctation that should be considered whenever blood pressure discrepancies are noted in patients with risk factors (hypertension, peripheral arterial disease, tobacco use).
- Although most commonly diagnosed with CT, ultrasound (either transthoracic echocardiography (TTE) or transoesophageal echocardiography) can also be used effectively particularly in the perioperative arena.
- Definitive treatment requires vascular surgery, usually open, although surgeons are increasingly using endovascular approaches. Medical treatment includes lipid-lowering agents, smoking cessation and antihypertensive agents.
- TEE is a valuable diagnostic modality that should be considered whenever unexplained haemodynamic changes or abnormalities are encountered in the perioperative arena.

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