CASE REPORT

Takayasu Arteritis with predominantly pulmonary involvement diagnosed by CT angiography

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Summary
A 19-year-old female was diagnosed with Takayasu’s Arteritis (TA) following CT angiography (CTA). The striking feature, to our knowledge not previously described in the literature, was marked pulmonary arterial (PA) involvement with a cuff of tissue up to 1 cm thick encasing lobar arteries whose lumen centrally was reduced to 4 mm. Clinical and plain film respiratory manifestations are often absent in TA but PA involvement is common (50–80% prevalence). CTA has superseded conventional angiography as the investigation of choice, allowing the acquisition of enhanced thin section images, in multiple phases of vessel opacification, showing enhancing circumferential mural thickening as well as luminal narrowing. Radiological evaluation of the pulmonary arteries should be performed even in the absence of respiratory symptoms.

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Case report
A 19-year-old female student of African descent, who had lived her entire life in the UK, presented with a one week history of flu like symptoms, including fever, sore throat and night sweats. She complained of sub-sternal chest pain radiating to her arms and shortness of breath on exertion. Her only past medical history was of an ostium secundum atrial septal defect, which had been diagnosed and surgically corrected when she was 8 years old.

On examination she had mild cervical and groin lymphadenopathy, pharyngeal injection and conjunctival pallor.
and an early diastolic murmur. She was initially referred for a cardiological opinion. A chest radiograph demonstrated prominent hila. An ECG displayed longstanding RBBB. She had a hypochromic microcytic anaemia, elevated acute phase proteins, lymphocytosis and thrombocytosis. Investigations for haemoglobinopathies, atypical pneumonia, T-cell dysfunction, infectious mononucleosis, malaria, mycobacterial infection and coeliac disease were all negative. A transthoracic echo-cardiogram merely demonstrated evidence of her previous surgery.

Because of her anaemia she was referred to a haematologist. The patient underwent a contrast-enhanced multi-detector computed tomography (MDCT) scan of the thorax. This demonstrated bilateral peri-hilar soft tissue cuffing of the main pulmonary arteries extending down the bronchovascular bundles, particularly involving the lower lobes (Fig. 1). This was initially thought to represent lymphatic tissue. There was no other significant lymphadenopathy elsewhere but there were some patchy areas of ground glass attenuation in the lower lobes and the spleen was slightly bulky. A radiological differential diagnosis of atypical pneumonia or sarcoidosis was made and the patient was commenced on broad-spectrum antibiotics and iron supplements.

The patient continued to suffer relapsing symptoms of fever, dyspnoea and lethargy. Serum calcium, angiotensin converting enzyme, vasculitic screens, bronchoscopy and video assisted thoracoscopic biopsy were normal. A bone marrow aspirate did not support the differential diagnosis of lymphoma.

She was referred for a respiratory opinion. Repeated examination demonstrated bilateral carotid bruits. The patient had complained of Raynaud’s phenomenon for several years. The peripheral pulses were now hard to elicit and blood pressure difficult to record. A CT angiogram (CTA) was performed eight months after initial presentation. This demonstrated a soft tissue cuff from the root of the ascending aorta to the lower descending aorta involving the aortic arch at the origins of the head and neck vessels with narrowing of the origins of the left common carotid artery and left brachiocephalic artery (Fig. 2). The apparent hilar nodal enlargement described on the initial CT was shown to be centred on the pulmonary arteries, particularly involving the lower lobe vessels (Fig. 3). Some of these arteries were attenuated peripherally.

The striking feature in this case was the particularly marked pulmonary arterial involvement with a cuff of tissue up to 1 cm thick encasing significantly narrowed lower lobe pulmonary arteries with a lumen centrally reduced to only 4 mm diameter. To our knowledge this degree of pulmonary arterial involvement has not previously been reported. The radiological appearances with the clinical presentation met the diagnostic criteria for Takayasu’s Arteritis (TA). The patient was commenced on oral steroid therapy and 75 mg Aspirin daily and gradually improved.

Discussion

Takayasu’s Arteritis is an idiopathic inflammatory arteritis that primarily affects the aorta and large vessels. First described in 1908, it is usually a disease of young women of East Asian ancestry. It remains rare in Western countries. There is no definitive diagnostic test and, as in this case, there is frequently a delay in establishing a firm diagnosis with alternative pathologies initially considered (median delay to diagnosis of 10 months). Presentation can vary, from those who are asymptomatic, to death from stroke. Vessel inflammation results in mural thickening, fibrosis and subsequent stenosis and thrombosis. By the time of diagnosis patients often have extensive vascular

Figure 1 Initial contrast-enhanced MDCT of chest demonstrating bilateral peri-hilar soft tissue cuffing of the main pulmonary arteries initially thought to represent lymphatic tissue.

Figure 2 Diagnostic coronal CTA of the chest demonstrating a soft tissue cuff from the root of the aorta to the lower descending aorta, involving the origins of the head and neck vessels, with narrowing of the origins of the left common carotid artery and mural thickening of the left pulmonary arteries.
disease. They may describe symptoms due to ischemia but presentation is often non-specific and features including fever, night sweats, malaise, arthralgia, myalgia and anaemia are commonly experienced first. The diagnosis of TA is made on the basis of clinical signs together with radiological evidence of large vessel arteritis.2,6

Pulmonary arterial involvement

Pulmonary arterial involvement was first described in 1940 but was regarded as a rare manifestation until recently. It now appears to be more common than initially recognised with a prevalence of between 50% and 80%.2,3,5,7 The absence of respiratory clinical manifestations with few plain film manifestations may explain why pulmonary arterial involvement was historically considered unusual. For this reasons radiological evaluation of the pulmonary arteries should be considered even in the absence of respiratory symptoms.

PA involvement has traditionally been demonstrated at pulmonary angiography. Advances in CT angiography (CTA) allowing the acquisition of well-enhanced thin section images, in multiple phases of vessel opacification, mean this modality is now the investigation of choice.7–10 CTA has the additional benefit over conventional angiography of showing the mural thickening as well as luminal changes. The primary CT finding in TA, which is particularly well demonstrated in this case, is circumferential wall thickening. Additional aortic findings which are described but which were not present in this case include high attenuation on pre-contrast images and contrast enhancement of the thickened wall. Concentric low attenuation inside the aortic wall associated with the mural enhancement is sometimes seen on delayed phase images.

Conclusion

Pulmonary arterial involvement is more common than previously appreciated in TA and, as in the case we report, can be severe. Clinically this may result in misdiagnosis. Mural thickening or localised thrombosis from TA can lead to misdiagnosis of embolic thrombus on CT. A high level of clinical suspicion is required for both the clinician and radiologist to make an accurate diagnosis. Radiological evaluation of the pulmonary arteries should be considered even in the absence of respiratory symptoms if TA is being considered. CTA incorporating evaluation of the pulmonary vessels as well as the systemic arteries has replaced conventional angiography as the investigation of choice.

Conflicts of interest statement

The authors have no conflict of interest.

References


Figure 3 Axial view of diagnostic CTA of the chest demonstrating mural thickening with luminal narrowing of the left pulmonary arteries.