Takayasu Arteritis: The Role of Multimodality Imaging

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Takayasu arteritis is a rare, chronic granulomatous inflammatory disease of the large arteries. The disease mainly affects females of reproductive age. Patients present with a wide range of non-specific symptoms. Diagnosis mainly relies on imaging features as laboratory investigations are non-specific.

We present a case of a 31-year-old Indian female who presented with chest pain and uncontrolled hypertension. Chest X-ray revealed aneurysmal thoracic aorta, which prompted further diagnostic investigations and was ultimately diagnosed as type V Takayasu arteritis.

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The disease is named after the Japanese ophthalmologist Mikito Takayasu, who described the first case in 1908[1]. The highest incidence is found in Japan of 30 cases per million per year[2]. It is mainly a disease of young females; however, male to female ratio varies around the world[2]. The disease has a predilection for the aorta, aortic branches, pulmonary and coronary arteries. Patients from Japan and South America are more likely to have thoracic aorta involvement while involvement of the abdominal aorta is more common in other Asian and Middle Eastern patients[3].

The etiology of the disease is still not well established; however, it is thought to be a T-cell mediated autoimmune process. Inflammation starts from the tunica adventitia proceeding to the tunica intima[4]. The process eventually leads to narrowing of the lumen and impairment of blood flow.

Symptoms vary upon the location of the stenosed arteries. Symptoms include, but are not limited to, neck pain, arm numbness, chest pain, dizziness, vision impairment and abdominal pain[2]. Systemic symptoms include low-grade fever and easy fatigability[2].

Diagnosis is usually based on radiological findings of CT angiography or MR angiography (MRA). Other valuable imaging modalities include angiography, fluorodeoxyglucose (FDG) scan and carotid sonography[5].

The aim of this study is to present an overview of the role of different imaging modalities in the diagnosis and follow-up of Takayasu arteritis.

THE CASE

A previously healthy 31-year-old Indian housemaid presented with episodes of atypical chest pain and headache. The BP was constantly elevated measuring ≥180/110 mmHg despite compliance with multiple antihypertensive medications. The patient had a weak left radial and bilateral femoral pulse with radio-femoral delay. Electrocardiogram revealed normal sinus rhythm with changes indicating left ventricular hypertrophy. Laboratory results were unremarkable, apart from elevated erythrocyte sedimentation rate (ESR) of 40 mm/h and C-reactive protein (CRP) of 8 mg/l. A chest X-ray revealed a saccular non-calcified aneurysm of the thoracic descending aorta, see figure 1.

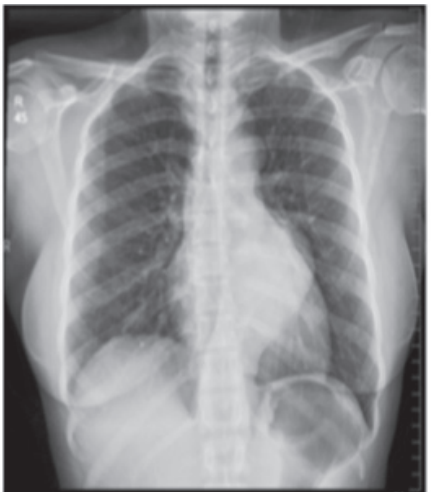

Figure 1: Plain X-ray Revealed Aneurysmal Dilatation of Thoracic Aorta with Irregular Contour

CT Aortogram was obtained using 64 slice CT scanner (GE optima CT660). A 70 ml of intravenous nonionic contrast (Optiray 350, ioversol 74 %) was injected automatically at a rate of 5 ml/sec. Multi-planner reconstruction was generated along with 3D reconstruction using volume rendering and maximum intensity projection techniques. Images demonstrated stenosis at the origin of the left subclavian artery followed by multiple areas of saccular dilatation (maximum caliber of 18 mm) and stenosis ending by a thread-like stenosis up to the proximal axillary artery. A tortuous fusiform aneurysmal dilatation of the descending aorta (maximum caliber of 5.8 cm) was seen extending downwards into the abdominal aorta, proximal to

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the renal branches, see figures 2 and 3. Multiple areas of wall thickening (maximum thickness of 5.5 mm) were seen in the descending and abdominal aorta which showed appreciable enhancement at the post-contrast study. A slit-like stenosis of the left renal artery was seen with the ischemic manifestation of the left kidney in the form of atrophy and significant hypoperfusion of the lower half. In addition, significant stenosis was also noted at the origin of the celiac trunk and superior mesenteric artery followed by a long segment of fusiform dilatation, surrounded by multiple tortuous collaterals. Gross tortuosity with multiple saccular aneurysmal dilatations of abdominal aorta below the level of renal vessels was found. Furthermore, a mural thrombus (maximum thickness of 27 mm) was seen extending downwards to the level of bifurcation into the left common iliac artery, causing almost complete occlusion. No evidence of sizable dissection or gross leakage was found. CT carotid angiogram was performed; however, it was negative for carotid or cerebral vessels involvement.

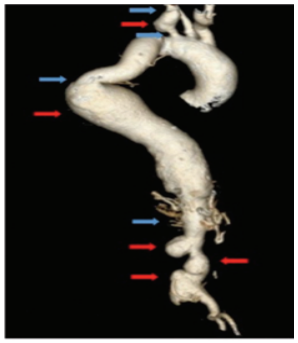

Figure 2

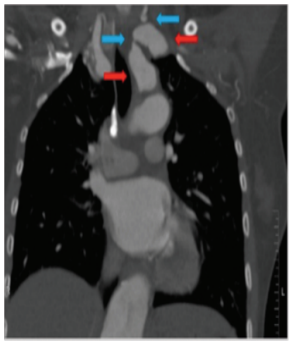

Figure 3

Figures 2-3: CT Angiogram with Coronal Reconstruction and 3D Volume Rendering Revealed Multiple Areas of Stenosis (Blue Arrows) and Aneurysmal Dilatation (Red Arrows) Involving Aortic Arch, Great Vessels and Descending Aorta

A decision was made to refer the patient to vascular surgery. Unfortunately, she was lost to follow-up after discharge.

DISCUSSION

Takayasu arteritis is a rare, large-vessel granulomatous arteritis. The disease mainly affects Asian and Middle Eastern patients; however, it can also be found in other races[2]. Japan reported the highest incidence of 30 cases per million per year compared to 2.6 cases per million per year in North America[2].

It preferentially affects females in the second or third decade. The inflammation is thought to be an autoimmune mediated T-cell reaction causing panarteritis[4]. Although the disease is not a genetic disorder, susceptibility to the disease is transmitted. Approximately 50% of Japanese patients carry HLA B25 allele; however, risk alleles are different in each country[2].

The disease has a long asymptomatic phase. Nonspecific symptoms of low-grade fever, chronic fatigue and weight loss precede symptoms of ischemia. Patients can present with complications such as vision loss, stroke, myocardial infarction, extremity or bowel ischemia and refractory hypertension[2]. Physical signs include absent pulses, vascular bruits and hypertension[2]. There is no specific blood test for Takayasu arteritis. Non-specific markers like CRP and ESR are often raised and therefore, used for monitoring disease activity[2].

The latest radiological classification was proposed by Numano in 1996 based on angiographic findings of involved vessels. Involvement of the branches from the aortic arch is considered type 1; involvement of the ascending aorta, aortic arch and its branches is considered type IIA; involvement of the ascending aorta, aortic arch and its branches, and thoracic descending aorta is considered type IIB; involvement of the thoracic descending aorta, abdominal aorta and/or renal arteries is considered type III; involvement of the abdominal aorta and/or renal arteries is considered type IV; involvement of the combined features of types IIB and IV is considered type V. Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively[6].

Our patient had type V according to this classification. In addition, our patient had involvement of the celiac, mesenteric arteries and its collaterals. The available classification schemes are anatomical and do not give information about prognosis.

Conventional digital subtracted angiography has been the gold standard for diagnosis. It is a precise method to show lumen of the vessels and measure aortic pressure[5]. It is also important for planning endovascular treatment. Alternating segments of narrowing and dilatation of the aorta and its branches above and below the diaphragm is the typical pattern. However, it does not show mural thickening and may give false negative results in early stages[5]. It is an invasive method that requires use of contrast and has significant radiation dose.

CT angiography (CTA) is a noninvasive tool to show wall thickening, luminal constriction and dilatation. It is also useful to measure the wall thickness. It has a comparable accuracy to conventional angiography for detection of large vessel involvement[7,8]. It has been observed that wall thickening is a very important sign in early stages that would give a clue for diagnosis. Post-contrast images would show the appearance of double ring, where the inner layer is poorly enhanced and the outer layer is intensely enhanced. The inner ring represents the swollen intima and the outer ring represents the inflamed outer layers. Therefore, the ring sign can be useful to follow-up disease activity[9]. The value of CTA in the follow-up of patients is not well demonstrated since studies had contradicting results[5]. CTA cannot show changes of small vessels. In addition, CTA carries the risk of contrast induced nephrotoxicity and excessive radiation[9].

MRI, similar to CTA, could also show luminal and wall changes. MRI in particular has the advantage of better demonstration of soft tissue changes. It could show arterial wall edema and increased vascularity as hyper intense signals on T2 weighted fat suppressed images. Active inflammation is seen on gadolinium enhanced; T1 weighted images as enhanced arterial wall, with progressive contrast accumulation and delayed washout[10]. 3D MRA has a 100% sensitivity and specificity for the detection of vascular lesions in the systemic and pulmonary circulation compared to conventional angiography[11]. It is an accurate, reproducible method to assess cardiac morphology

and function in patients with co-existent aortic regurgitation[12]. The main advantage of MRI is that it can be repeated without exposing the patient to radiation hazards.

US is a quick non-expensive method for the assessment of superficial vessels. US evaluation (carotids, subclavian, and common femoral arteries) was able detect changes suggestive of Takayasu in 98% of patients compared to CT angiography[13]. The typical finding is smooth concentric homogenous long segment thickening of the arterial wall. In contrast, atheromatous plaques are irregular calcific lesions. It can show intima media complex thickening in early disease stages before lesions are detectable on angiography[5]. Assessment of other vasculature is limited due to presence of superimposing tissue or bowel. Another major drawback, it is operator dependent.

18F-Fluorodeoxyglucose Positron Emission Tomography CT or MRI can show an increased uptake in areas of active inflammation in vessels larger than 4 mm[14]. It can detect areas of inflammation in the pre-stenotic phase. The presence of atherosclerosis would give a similar picture, but Takayasu is a disease of young age unlike atherosclerosis. Studies have shown correlation between reduction in FDG uptake and clinical response to treatment and reduction of wall thickness[14]. However, it has low resolution which prohibits small vessel evaluation. Moreover, PET/CT is not widely available and quite expensive.

CONCLUSION

Takayasu arteritis is a rare disease with constellation of non-specific symptoms and laboratory findings. Imaging would play a major role for early diagnosis, if clinicians had a high index of suspicion. Radiologists must be aware of its imaging features to guide clinicians in their diagnosis and follow-up of patients.

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