Takayasu's disease in a young male

Natalie Smit 3rd year diagnostic radiography student, Department of Radiography and Nursing, Faculty of Health and Wellness Sciences, Cape Peninsula University of Technology, Cape Town, South Africa

Abstract: Takayasu's arteritis, which is also referred to as a pulseless disease or aortic arch syndrome, is an inflammatory and stenotic disease of the medium and large sized arteries [1]. The sequence of events that led to the diagnosis of Takayasu's disease in a twenty-five year old male patient and subsequent complications and treatment regimen used in the treatment of his disease are reported on. Complications associated with Takayasu's disease are highlighted. The epidemiology, patient management and radiographic appearance of the disease are discussed [1-9].
Keywords: aneurysms, stenosis, bruit, revascularisation surgery.

Case report

A 25 year old male, with a known history of Takayasu's disease, presented to a private health care facility with a three day history of increasing epigastric pain that radiated through to the back with some associated nausea. A lower chest and abdomen computed tomography [CT] scan demonstrated a dilated distal thoracic aorta just above the diaphragm. At the lower hiatus there was a dilation of the aorta with what appeared to be extramural thrombus present. The aneurysm extended down to the distal abdominal aorta. These findings led to the diagnosis that the patient had symptomatic Type 111 thoraco-abdominal aneurysm. He was immediately transferred to the intensive care unit where his blood pressure was controlled prior to him undergoing surgery to repair his thoraco-abdominal aneurysm. Two months following the surgery he presented with discomfort in his back and left lower chest and this was found to be related to a new aneurysm which had developed over the past few weeks. It was recommended that a stent graft of his aorta would be the best and safest option. He subsequently underwent a second endovascular repair of the thoracic aorta rupture (Figure 1). He recovered remarkably well.

When he was 12 years old he presented with a complaint of weight loss and pyrexia. After extensive clinical examinations he was diagnosed as having Takayasu's disease on the basis of absent pulse in his right arm. Two years later, after being treated with steroids, he returned to his general practitioner, presenting with hypertension. At this stage he was referred to a vascular surgeon for further management. It was found that his aorta was increased in diameter and a tight stenosis was demonstrated in the mid section of the right renal artery. His right kidney was smaller than the

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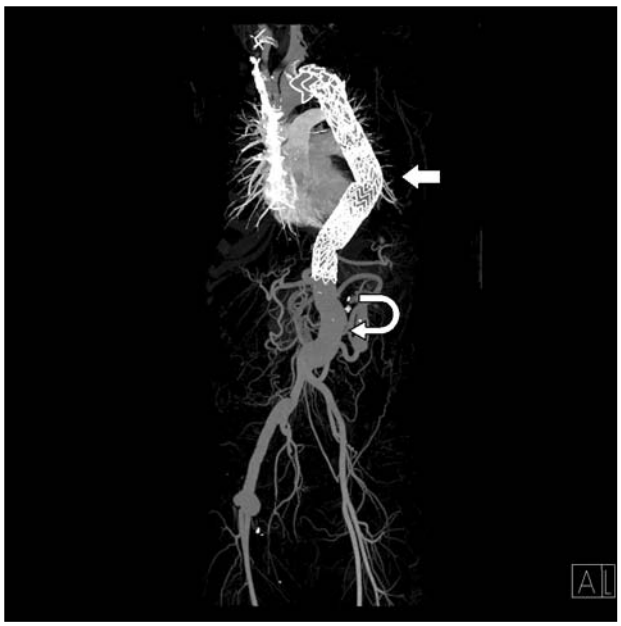

Figure 1: Computer Tomographic Angiogram of abdominal aorta and peripheral vessels. Demonstrates the thoracic aortic stent (white arrow). Also note the dilatation of the abdominal aorta (curved arrow).

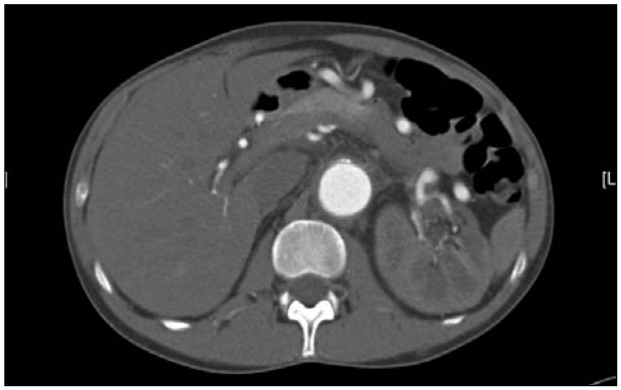

Figure 2: Computer Tomography of abdomen: Image demonstrating the absence of the right kidney, due to a right nephrectomy.

left one and it had some cortical thinning and decreased parenchymal blood flow on duplex. As a result of the right renal artery occlusion with reno-vascular hypertension he underwent a right nephrectomy (Figure 2). Post surgery his blood pressure stabilized and he was later discharged.

Four years following his nephrectomy he had a stent placed in his remaining renal artery. The following year he was diagnosed as having left renal artery occlusion confirmed on angiography; renal revascularisation surgery to the left kidney was performed. During this procedure it was found that the diseased abdominal aorta had no palpable pulse present at the origin of the superior mesenteric artery thus the saphenous vein was anastomosed to the inferior mesenteric artery resulting in a clearly palpable pulse in the renal artery beyond the vein to artery anastomosis and a pulsatile kidney. The inferior mesenteric artery was noted to be patent at the time of the angiogram.

Radiographic findings

A follow-up CT scan, a month after the second thoracic aortic rupture repair, demonstrated that the graft was well placed with the proximal stent positioned down to just above the celiac origin from his previous placed surgical graft. The patient has since stabilized and is required to have a follow-up CT angiogram scan annually. The latest CT scan demonstrated a false aneurysm in his right femoral artery arising from the ileo-femoral graft and he will require repair to this artery in the foreseeable future (Figure 3). This study also demonstrated that the subclavian aneurysm was unchanged in size (Figure 4).

Discussion

Takayasu's disease is a chronic inflammatory and stenotic disease which primarily affects the medium and large sized arteries characterized by a strong predilection for the aortic branches. It causes varying degree of stenosis, occlusion, or dilation of the involved vessels as well as aneurysm formation [1-4]. Takayasu's disease is an uncommon disease; it is prevalent in adolescent girls and young women, with the onset usually occurring between the ages of 10 to 40 years[1,5]. The female to male ratio is 8:1[6]. Takayasu's disease is distributed world wide, but is more common in Asia and in parts of the world where there is a high incidence of tuberculosis [3, 6]. Takayasu's disease is a systemic disease with a variety of symptoms which vary from generalized symptoms to local symptoms. The general symptoms which may occur, include night sweats, anorexia, malaise, fever, increased blood pressure and weight loss. These symptoms may occur months before vessel involvement is apparent [1,5]. Localized symptoms, related to pain over the involved vessels, followed by

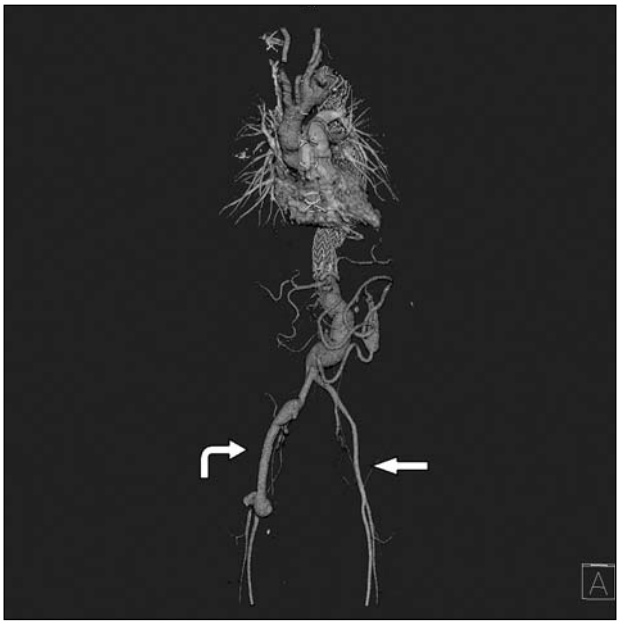

Figure 3: Computer Tomographic Angiogram of abdominal aorta and peripheral vessels shows a false aneurysm in the right femoral artery (curved white arrow). The left femoral artery appears normal (white arrow).

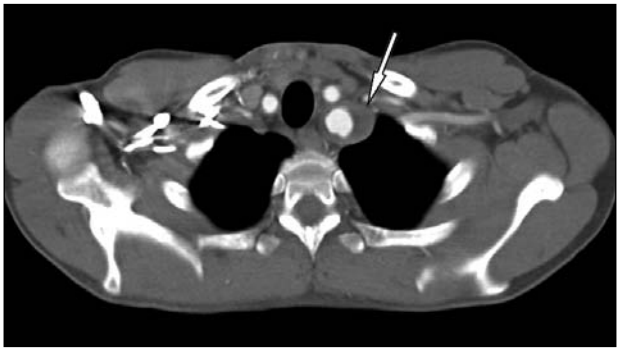

Figure 4: Computer Tomography axial view of chest: An aortic aneurysm in the left subclavian artery is noted (see arrow).

ischemia in the organs that are supplied by the compromised vessels, usually develop [1]. Absence of normal pulses commonly occurs in the involved vessels and the vessel most often affected is the subclavian artery. Decreased pulsation may also occur in one or both brachial arteries. Bruit over subclavian arteries or aorta may be audible on auscultation, which may occur in one or both subclavian arteries or abdominal aorta [5]. The development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities, may occur [1, 5]. The disease may also affect the pulmonary artery. Involvement of the major branches of the aorta is much more marked at their origin than distally [1]. Inflammation initially appears around the vaso vasorum in the media and adventitia, but fibrosis, scarring and vascularization of the media, as well as disruption and degeneration occur of the elastic lamina. This results in the narrowing or obliteration of the lumen, which may occur with thrombosis [1,3,5]. As a direct result of the compromised blood flow through the involved vessels, pathological changes often occur in various organs [3].

Takayasu's disease has been divided into various groups, based on angiographic morphology. Type1 involves the aortic arch and its branches, Type 11 involves the thoraco-abdominal aorta and its branches. Type 111 which is the type discussed in this report, involves lesions of both Types 1 and 11 [3,4]. Takayasu's disease does not usually affect the infra-renal aorta and the iliac vessels. The inferior mesenteric artery is rarely involved [3]. The modalities of choice in the diagnosis of Takayasu's disease are CT, ultrasound and contrast enhanced magnetic resonance imaging which provides information of mural changes of vessels [3,8]. It is believed that CT is superior to that of aortography, although stenosis and aneurismal changes are evaluated equally by both modalities. Mural changes are demonstrated better on CT [4]. Initial non-contrast studies on patients with suspected Takayasu's disease demonstrate a high-attenuation aortic wall. Aortic wall enhancement and delayed enhancement have been described with contrast studies [4].

Diagnosis of Takayasu's disease is based on clinical findings and should be suspected in young women who present with decrease or absence of peripheral pulses, abnormal blood pressure readings and arterial bruits [1]. The disease is confirmed by the characteristic patterns seen in arteriography studies. These patterns may include irregular vessel walls, stenosis, post-stenotic dilation, the formation of aneurysms, and occlusion [1,3]. Complications that may arise due to the disease include hypertension, aortic regurgitation, aneurysms and possibly cardiac involvement [3].

The prognosis of Takayasu's disease has improved over the years due to advances in interventional procedures for the treatment of renal and aortic stenosis. In the acute phase the disease is treated with corticosteroids. Reconstructive vascular surgery of stenosed vessels has improved survival and reduced the morbidity rate because it reduces the risk of strokes and improves blood flow which is especially beneficial in patients suffering from hypertension secondary to aortic or renal lesions [7]. Reconstructive surgery should be avoided during periods of active inflammation [1,3,6].

Conclusion

Takayasu's arteritis is a fairly uncommon disease. Although many complications may arise in a patient the prognosis of Takayasu's disease has improved over the years due to improved interventional procedures for the treatment of renal and aortic stenosis. With the consideration of modern medicine and the efficient medical treatment by specialist doctors the possibility of the patient living a fairly normal life is enhanced.

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ANSWERS to the CPD questions of the April 2008 issue:

Question 1. Pituitary macroadenomas are a common intracranial mass lesion.
Question 2. Pituitary macroadenomas are classified according to whether they produce one of the following substances: hormones.
Question 3. Which of the following is not considered a complication of macroadenomas: abscess formation.
Question 4. The mortality rate for patients with follicular thyroid cancer ranges between 10 - 40%.
Question 5. Follicular carcinomas accounts for about 25% of thyroid malignancies.
Question 6. A synovial sarcoma is a malignant disease originating from soft tissue.
Question 7. The majority of synovial sarcomas occur in the extremities.
Question 8. Synovial sarcomas occur mostly equally in males and females.
Question 9. Radiation therapy is indicated for a meningioma when the tumour was partially removed.
Question 10. A patient with an intraosseus meningioma suffers from a loss of vision because the meningioma compressed the optic nerve.
Question 11. Meningiomas are common in the following anatomical site: Arise from the dura or arachnoid.
Question 12. The patient with an intraosseus meningioma suffered from right-sided proptosis since there was a prominence on the greater wing.
Question 13. Schwannomas are best imaged and diagnosed by MRI with gadolinium-based contrast medium since it provides excellent soft tissue resolution.
Question 14. The treatment of Schwannomas aims to relieve symptoms and monitor Schwannoma development.
Question 15. A Schwannoma originates from Schwann cells.
Question 16. Schwannomas are common in the brain and spine.
Question 17. Schwannomas can develop on the Peripheral, cranial and spinal nerves.
Question 18. Which pathology is not a complication of follicular tumours? Abscess formation.
Question 19. Raised intracranial pressure may be caused by mass lesions of the brain.
Question 20. Raised intracranial pressure may lead to displacement of the brain.