A case of abnormal architecture.

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(Answer on page 52)

Just having settled in to your new office at the new Halifax Infirmary, you decide to unpack your things. You notice amid your belongings, a strange brown envelope marked "important", the contents of which is the single radiograph shown as Figure 1.

Q 1: With no other information, can you find the "important" features of this film?

Q 2: Can you give a quick differential with this limited information.

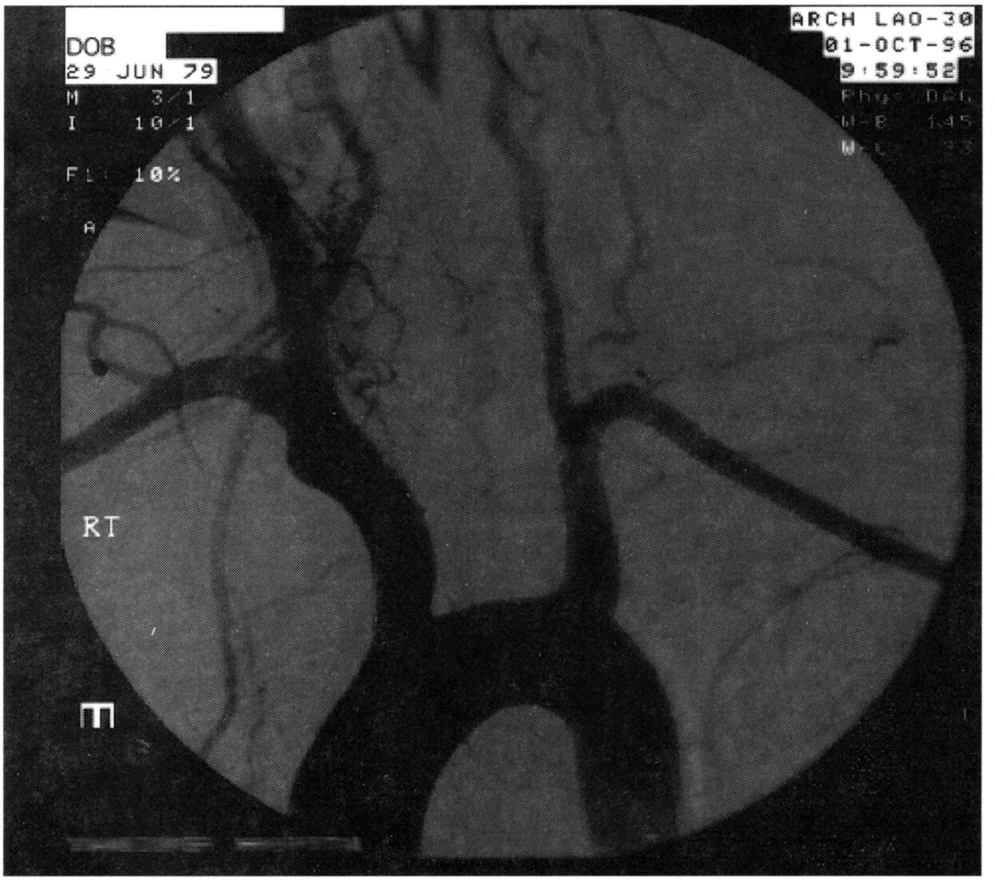

Figure 1.

On closer exam of the envelope you find a small piece of paper inside which reads:

> **"16 yr old female with hx. of L parietal stroke in 1988 and hypertension. Recent medullary infarct with good resolution."**

Q 3: What is your differential now?

Q 4: What is the structure marked "H"?

Q 5: What treatment / intervention would you offer this patient?

Q 6: Are there any other areas of concern that you would like to see radiographically?

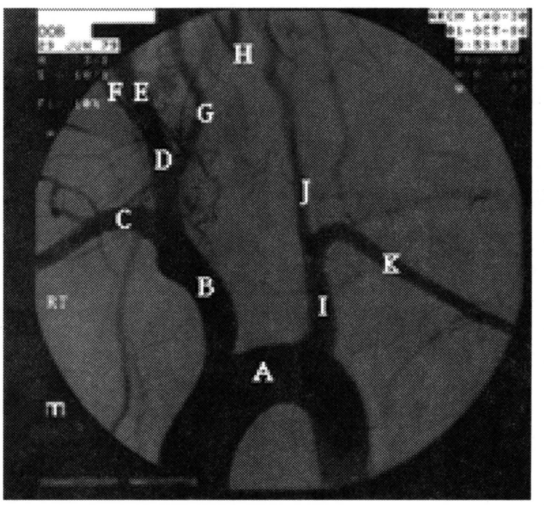

Figure 2: A- aortic arch, B- brachiocephalic trunk, C- R subclavian, D- R common carotid, E- internal carotid, F- external carotid, G- R vertebral, H- ???, I- L subclavian trunk, J- L vertebral artery, K- L subclavian

Answer: Takayasu's Arteritis

Takayasu's Arteritis (TA) is an acute periarteritis that most commonly presents in young or adolescent females. This inflammatory disease has historically been seen most commonly in an Asian population but recent studies have shown that more cases are presenting around the world. In North America, the estimate of incidence is 2.6 per million persons per year (1). TA tends to affect both the proximal aorta and its branches (2) but involvement of the abdominal aorta and its branches has also been studied (3).

Among the varied signs and symptoms that TA can present with include fever, sweating, arthralgia, myalgia, cough, hemoptysis, pleural effusion, elevated ESR, and leg ulcers (4). More characteristic symptoms of TA involve a history of TIAs, stroke, cool extremities, headaches, dizziness, amaurosis fugax or diplopia (2). Even vague signs, such as intra-abdominal pain or unexplained hypertension may be important clues to mesenteric or renal artery stenosis. Physical signs (incidence percentages in patients with TA shown in parenthesis) like vascular bruits (80%), claudication (70%), aortic regurgitation (20%), carotodynia (30%) and diminished or absent pulses (60%) can often be elicited from patients with TA (1).

The disease presentation of TA typically comes in two distinct stages. In the "pre-pulseless" (early) or systemic phase, TA usually presents with the vague constitutional symptoms. This is a contrast from the obliterative or "pulseless" (late) stage where the diagnosis is usually made and the patient shows ischemic and inflammatory changes in their vasculature (5). There are no specific Human Lymphocyte Antigen markers that directly correlate with TA in the North American population (6). ESR is the only consistently elevated lab result seen in active forms of TA.

Diagnosis is generally confirmed by arteriography (especially digital subtraction angiography) as is shown in Figure 1. Now both CT and MRI are being used to show luminal narrowing and mural thickening to support the arteriographic findings of afflicted vessels (7, 8). The five year survival rate has been reported to be over 90% (2).

A differential diagnosis could include the following pathologies: 1) chronic aortitis with involvement of the common carotid artery, 2) giant cell arteritis, 3) ankylosing spondylitis, 4) congenital absence of left common carotid, 5) arterial occlusion/ embolism, 6) thromboangitis obliterans (Buerger's Disease).

Treatment usually involves the use of glucocorticoids (with 60% remission rate). Additional benefits are gained with the administration of cytotoxic agents in those who have relapse of their disease on glucocorticoids (2). Surgical treatment with bypass of significantly stenosed vessels (9) and transluminal balloon angioplasty in those with aortic obstruction (3) has been shown to be protective in the development of stroke or other thromboembolic events.

Returning to this case, the mysterious y-shaped object seen at the top of the angiogram is the bifurcation of the obliterated left common carotid. It is interesting to note the smoothness of the arch. Most initial impressions of the film tend to favor a congenital absence of the great vessel to explain the anomaly. However, the appearance of the bifurcation leads to the diagnosis of TA.

Although TA predominantly affects the aorta, upper extremity vessels and the cerebral blood supply involvement of the abdominal aorta is not uncommon (4). Examination of the renal arteries, the superior mesenteric artery and the femoral-iliac system should be undertaken, especially in the case of this patient who has a history of hypertension.

In truth, this patient did have TA involving her right renal artery that was successfully treated with balloon angioplasty at age 9. There has been no relapse of stenosis in this vessel since then. The patient had a well developed collateral blood supply through the Circle of Willis and no attempts to bypass the carotid stenosis were being considered at the time of the investigation.

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