Four diagnostic exercises

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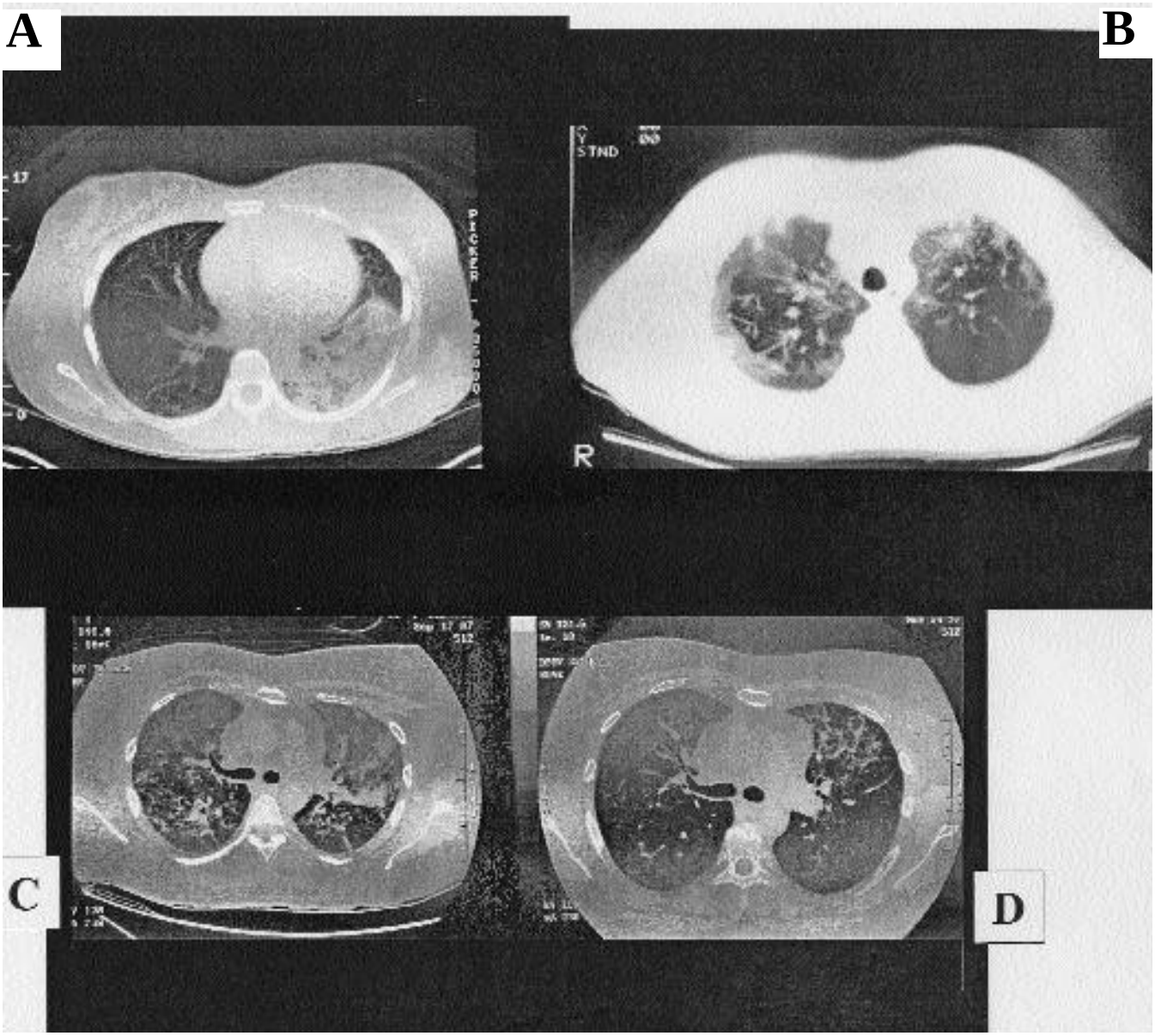
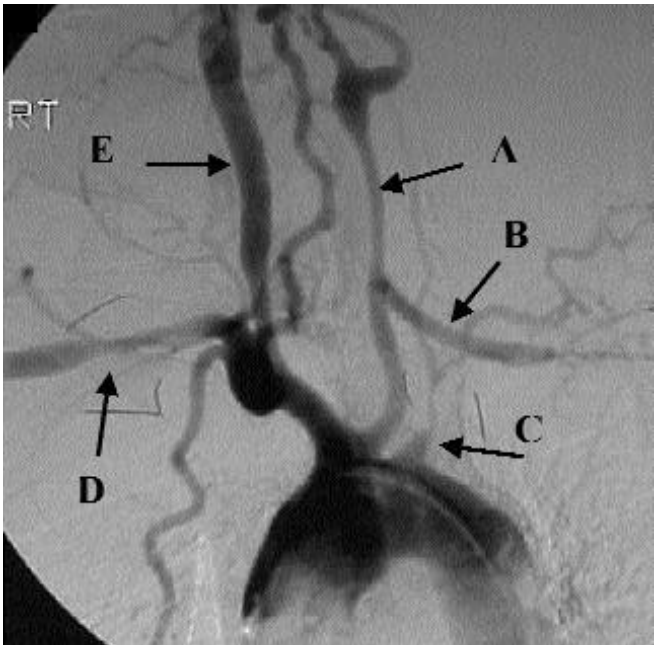
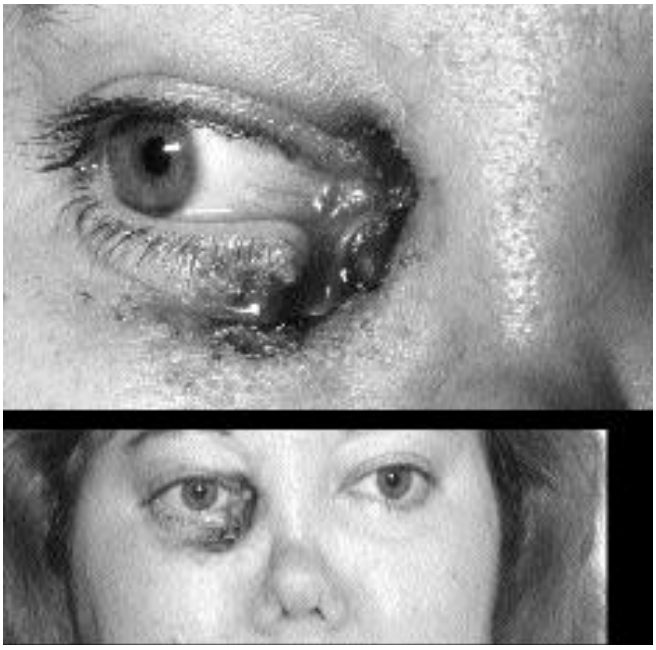
I. Examples of pneumonitis of different etiologies in 3 patients with vasculitis. (A) and (B) are two different patients. (C) and (D) are the same patient before and after a therapeutic decision was made – 2 months elapsed between C and D. Can you match each of these pictures with a diagnosis?

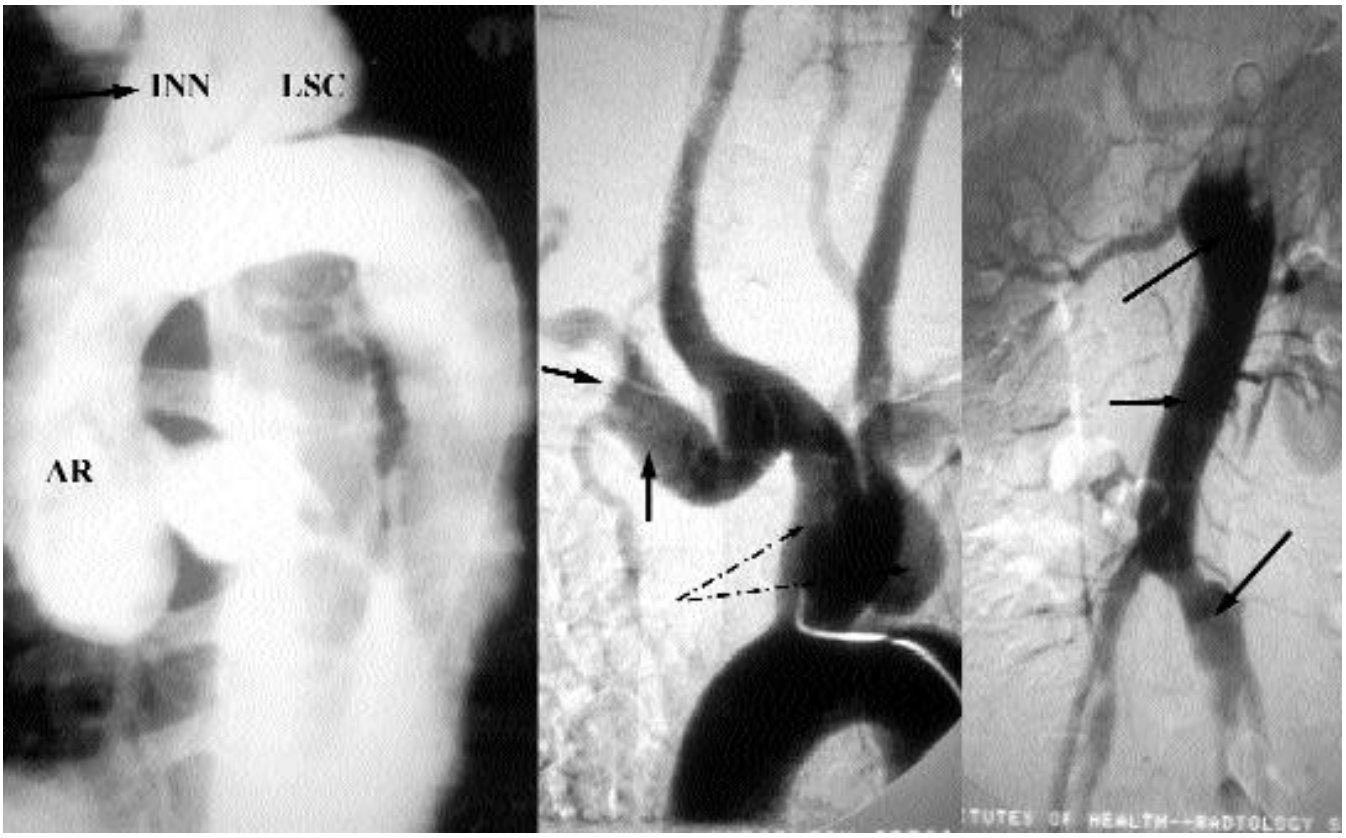




II. Can you name the vessels indicated by the arrows and describe their abnormalities?



III. What is the diagnosis, the abnormality seen, and its origin?



IV. What is the diagnosis?

Answers

I. Examples of pneumonitis of different etiologies in 3 patients with vasculitis.

(**A**) Alveolitis secondary to Churg-Strauss;

(**B**) *Pneumocystis carinii* pneumonitis in a patient with polyarteritis nodosa, treated with cyclophosphamide and glucocorticosteroids (not receiving *P.c.* prophylaxis);

(**C**) Cyclophosphamide-induced pneumonitis; first picture during treatment and second picture (**D**) two months after discontinuation of the medication.

In patients with vasculitides that may involve the lungs, the differential diagnosis of pneumonitis should include the disease process (e.g. Wegener's granulomatosis, Churg-Strauss, microscopic polyangiitis etc), opportunistic infections, and drug-induced pneumonitis. It is crucial to make the correct diagnosis because appropriate treatment for each is dramatically different. Increasing immunosuppressive therapy in a patient with an opportunistic infection may lead to significant morbidity or even death. The diagnostic work up frequently requires, at the very least, bronchoscopy with bronchoalveolar lavage.

II. Can you name the vessels?

(**A**) Left common carotid artery

(**B**) Surgical graft from left common carotid artery to distal left subclavian artery, which is occluded

(**C**) Occluded proximal left subclavian artery

(**D**) Stenotic right subclavian artery

(**E**) Dilated right common carotid artery

This patient with Takayasu's arteritis (TA) has bilateral subclavian stenosis, which on the left has led to placement of a graft from the common carotid to the distal subclavian. Because subclavian stenosis occurs in over 90% of patients (bilateral in ~40%) with TA, and the common carotid arteries are also frequently involved, grafts to the subclavian and common carotid arteries should originate from the ascending aorta. The aortic root and arch may be involved in TA (approximately 60% of the cases) but it is almost never stenotic. Aortic root lesions are aneurysmal, a condition that would not compromise graft patency.
Bilateral subclavian stenosis prevents accurate assessment of arterial blood pressure measured by arm BPcuffs. If the aorta is continuously patent from the root to the iliac arteries, the leg-BP cuff recordings may be a reliable reflection of central aortic pressure. Unrecognized central hypertension can be a serious source of morbidity and mortality.

III. What is the diagnosis, the abnormality seen, and its origin?

The primary abnormality in this patient with Wegener's granulomatosis (WG) is in the ethmoid sinus. The intense inflammatory, necrotizing process resulted in the development of an ethmoid-cutaneous fistula. Vision was not impaired. Fistulization of this type is a rare complication of WG.

IV. What is the diagnosis?

This 18-year-old male became ill at the age of 13. He presented with fever, uveitis, arthritis, Bell's palsy, erythema nodosum, and asymptomatic bruits. He subsequently developed aortitis, large aneurysms, and bilateral subclavian occlusion. Initially, he was thought to have Takayasu's arteritis but a synovial biopsy showed proliferative arthritis with non-caseating granulomas. A diagnosis of sarcoid vasculitis was made. His disease course was complicated by aortic dissection requiring grafting of the entire aorta. Sarcoidosis can involve vessels of all sizes and may therefore mimic many different types of vasculitis.

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