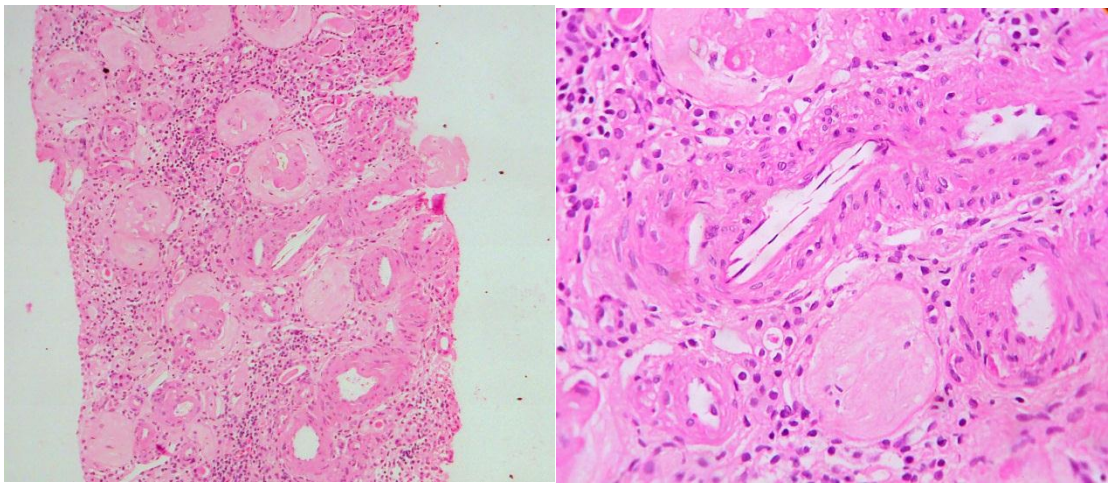
Image Quiz 2

Sixty year old male , a known case of systemic hypertension on regular medication and coronary artery disease presented with worsening of renal functions over the last three weeks. He was receiving oral anticoagulants for purplish discoloration of toes, prescribed elsewhere.

Serum creatinine at presentation 3.2 mg% (282.8 u mol/L). urine protein (dipstick) 1+, 24 hours urine protein 428 mg.



Answer: Atheroembolic renal disease (AERD) with benign nephrosclerosis.

Discussion:

Atheroembolic renal disease (AERD) is defined as renal failure secondary to the occlusion of renal vasculature by cholesterol containing atheromatous plaques dislodged from the aorta or other major arteries [1]. This occurs either spontaneously, or as an event following invasive vascular procedures such as angiographic catheter insertions, or after use of anticoagulant and thrombolytic agents [2].Incidence of AERD is difficult to estimate as it is often underdiagnosed owing to its propensity to coexist with as well as mimic several other renal disorders [3]. The reported incidence in biopsy series varies from 1.1 to 4.5 % in

various studies [5] and an approximately similar incidence in few autopsy studies [6].

The classic clinical manifestations of AERD include livedo reticularis (purplish rash over lower extremities and abdominal wall), purple toes, small nail bed infarcts and various other manifestations related to lodgement of atheroemboli in other organs. It is worth noting that many of these manifestations also associated with systemic vasculitis, which is often the first clinical differential diagnosis in patients with AERD [3], A further diagnostic difficulty arises in patients with pre-existing peripheral vascular disease, wherein livideo reticularis may be mistaken for manifestations of peripheral vascular disease, leading to a delay in diagnosis of the condition. In the present case, the finding of livedo reticularis (purplish discoloration of lower extremities and toes), was ascribed to peripheral vascular disease, and oral anticoagulants were prescribed, which possibly led to the sequence of events culminating in AERD.

AERD has a variable but definite temporal relationship to invasive vascular procedures and use of thrombolytic and anticoagulant therapy [2]. Overall it is estimated that about 30-85% of patients with AERD have a history of invasive vascular procedure in the preceding three months .Studies have shown that about 13-22% patients with AERD received anticoagulant therapy (heparin or warfarin), [1] as noted in the present case. The mechanism involves dissolution of superficial clots which destabilize the ruptured atherosclerotic plaques, resulting in showering of cholesterol atheroemboli into the circulation. The gold standard of diagnosing AERD remains the demonstration of atheroemboli in renal vasculature in a renal

biopsy. Delay or failure to diagnose this condition often stems from hesitation to perform renal biopsies, as the patients are elderly and have compromised kidneys due to co-existing morbidities. Cholesterol emboli appear as negative biconvex shadows appearing as “ghosts”. Involvement of renal vasculature is patchy and the diagnosis may be missed if few sections are examined in a biopsy.

Management of AERD is still empirical and no defined protocols have been developed. Medical management involves use of statins [7] or corticosteroids. Few studies have demonstrated the benefits of pulse steroid therapy in patients with AERD [8]. The rationale behind use of steroids in AERD is not clear but may be related to the suppression of reactive inflammation often seen along with the atheromatous plaques. Activation of the complement (C5) in vitro along with evidence of hypocomplementemia and eosinophilia in many patients suggest a possible role for inflammation in pathogenesis of AERD and perhaps the response to steroid therapy [1].

References:

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