P425

P425-Ruptured renal artery aneurysm leading to the diagnosis of Behçet´s disease

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BACKGROUND:
Behçet´s disease is a rare systemic vasculitis, with unknown aetiology, involving both arteries and veins of all sizes, which follows a chronic and relapsing course. Renal involvement is uncommon and shows a wide variation. Renal amyloid A amyloidosis, glomerulonephritis and vascular involvement are the main causes of renal Behçet´s disease. [1-3] Among vascular complications, arterial involvement is less common than venous involvement and occurs in 1-7% of patients with Behçet´s disease. Arterial disease includes aneurysms, stenosis and thrombus formation, and is associated with high morbidity and mortality. Renal arteries are uncommonly involved. [4]

CASE REPORT:
A 28-year old male patient of Turkish ethnicity initially presented with an acute renal haemorrhage due to a ruptured left lower segmental artery aneurysm which was successfully treated with coil embolisation. 6 months later he presented again with an acute left-sided renal haemorrhage and acute kidney injury. A transfemoral renal angiogram was performed and demonstrated a ruptured left upper segmental artery aneurysm, two microaneurysms of the right lower segmental artery and bilateral renal microinfarcts. Coil embolisation of the left upper segmental artery was performed successfully. After taking extended history and including a systemic enquiry the patient reported recurrent oral aphthous ulcers for 10 years, genital ulcers and recurrent superficial thrombophlebitis after having blood samples taken. He fulfilled the international criteria for Behçet´s disease (ICBD), scoring 6 points (oral aphthosis, genital aphthosis, skin lesions, vascular manifestations), and thus was diagnosed as having Behçet´s disease. The patient was commenced on high dose glucocorticoids (1mg/kg/day), azathioprine (2mg/kg/day) and Tumor necrosis factor-alpha (TNF-α) inhibitor (Infliximab – 40mg fortnightly). He was discharged with normal kidney function and remains relapse free.

CONCLUSION:
There are no pathognomonic laboratory tests in Behçet´s disease and diagnosis is made on the basis of clinical findings. Renal artery involvement in Behcet´s disease is very rare and can be life-threatening. In a patient with renal artery aneurysms, Behçet´s disease should be one of the differential diagnoses. When diagnosed it should be treated with immunosuppressive therapy and surgical intervention if needed.