ANGIOMYOLIPOMA OF THE KIDNEY

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Introduction

The angiomyolipoma is an unusual tumour, composed of mature adipose tissue, blood vessels and smooth muscle with supporting stroma. It is generally regarded as benign-hamartoma. Such tumour may grow to a large size.

There is a well recognized association between angiomyolipoma and tuberous sclerosis. In such cases tumours are invariably multiple and frequently bilateral. They may enlarge to produce palpable renal masses. This may also be found in patients lacking the stigmata of tuberous sclerosis complex. Inglis (1960) and Price and Mostofi (1965) gave an excellent resume of condition in 30 patients free from tuberous sclerosis. The commonest presenting symptoms are loin pain and renal mass.

Case Report

A 30 years old woman was admitted in Surgical A Ward, DHQ Hospital, Abbottabad, with left loin pain for 1½ years. She also had low grade fever and burning micturation but no history of haematuria.

On examination: left renal angle was tender and a big mass was palpable in left renal area. Haemoglobin was 11.6 gm/dl; white cell count 5700/cmm, blood urea was 40 mg/dl;

I.V.U. showed left kidney was grossly displaced downwards by a soft tissue shadow but both kidneys were functioning.

At operation a greyish, haemorrhagic mass measuring 12″ x 6″ was found arising from upper pole of left kidney, infiltrating peritomeum, and diaphragm. Left nephrectomy was done, becuase of suspicion of a renal cell carcinoma. Specimen was submitted for histopathology, which confirmed diagnosis of angiomyolipoma.

Discussion

Angiomyolipomas are thought to exist in two clinically distinct forms. 40 - 80% occur in association with tuberous sclerosis, a familio-hereditary disease characterised by the triad of mental retardation, epilepsy and adenoma sebaceum. They are multiple and bilateral in 80% of cases. They are often asymptomatic. The sex incidence is equal. Angiomyolipomas not associated with tuberous sclerosis are characteristically large, unifocal and symptomatic. There is a marked female preponderance with a female:

male ratio of 4:1. The peak incidence occurs in the fifth decade. Other authors disagree with this classification and consider that a solitary angiomyolipoma may be the only clinically evident manifestation in an incomplete variant of the tuberous sclerosis syndrome.

Macroscopically the tumour consists of friable, greasy tissue. Its colour ranges from yellow to grey depending on whether adipose tissue or smooth muscle predominates. It occurs predominantly in the parenchyma of the kidney although extension beyond the renal capsule and into the perirenal tissues occur in 25%. Invasion into the renal pelvis is uncommon. Haemorrhage, both inside the tumour and around it, is extrmely common. Histologically the tumour consists of an intimate admixture of adult adipose tissue, blood vessels and smooth muscle in varied proportions. The blood vessels are tortuous, thick walled and lack elastic tissue lamellae. The presence of these abnormal blood vessels explains the propensity of angiolipomas to bleed.

Angiomyolipoma is a benign tumour. There has been no documented case of distant metastasis and long survival after residual tumour has been left behind is well recorded. However progressive distruction of renal tissue in bilateral cases can lead to renal failure and a fatal outcome.

The tumour may be entirely asymptomatic and come to light only at autopsy or at operation for an unrelated condition. The commonest presenting symptom is loin or abdominal pain. The pain may be of abrupt onset and great severity when the tumour bleeds either intrarenally or retroperitoneally. There may be signs of acute blood loss and a loin mass may be palpable. In other cases the pain may be more chronic and intermittent. Other clinical features include haematuria, fever and hypertension.

Diagnosis in non-tuberous sclerosis cases is often difficult and preoperative diagnosis seldom possible. The main difficulty lies in differentiating angiomyolipoma from malignant tumours of the kidney. Plain abdominal film may show irregular areas of radiolucency not corresponding to bowel shadows, reflecting the high fat content of the tumour. This radiolucent fat sign may be obscured if there is perirenal haemorrhage. IVU may show a mass effect distorting the collecting system. Characteristic findings on angiography include multiple grape-like aneurysms of the inter-mediate renal arteries, a whorl-like appearance of the veins and the lack of arteriovenous shunting.

As the tumour is benign the aim should be the preservation of renal substance if possible. Surgical exploration should be reserved for those who are symptomatic and in whom the diagnosis remains in doubt. A frozen section biopsy may clinch the diagnosis and avoid an unnecessarily radical operation. Unilateral, solitary lesions should probably be completely excised as recurrent bleeding and progressive renal destruction can occur. If distinction from renal cell carcinoma is not possible a radical nephrectomy is justified.

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