Rupture of a renal angiomyolipoma (hamartoma)

M. STAVOROVSKY
M.D.

B. MORAG
M.D.

D. IRGE
M.D.

E. SCHUJMAN*
M.D.

**Department of General and Vascular Surgery and Pathological Institute,
Municipal-Governmental Medical Center Tel-Aviv/Yaffo, Ichilov Hospital
and Tel-Aviv University Sackler School of Medicine, Tel-Aviv, Israel*

Summary

A 54-year-old woman with spontaneous rupture of angiomyolipoma is presented. The literature on the 2 types of angiomyolipoma, without and with association of tuberose sclerosis, their rarity and the difficulties in their diagnoses, is reviewed.

Introduction

Angiomyolipoma (hamartoma) of the kidney is a rare finding. Hajdu and Foote (1969) recorded an incidence of 0.31% in 8501 post-mortem cases. McCullough, Scott and Seybold (1971), Becker *et al.* (1973), Honey and Honey (1977) consider that 50-80% of renal hamartomas appear in patients with tuberose sclerosis. Angiomyolipomas without tuberose sclerosis are usually solitary and unilateral. Rupture may be a complication in 4.5% of the solitary angiomyolipomas (Vasko, Brockman and Bomar, 1965).

To the author's knowledge, only 9 rupture cases have been reported (Vasko *et al.*, 1965; Wapnik *et al.*, 1975). An additional case is now presented which was diagnosed intra-operatively.

Case report

A 54-year-old woman was admitted with acute abdominal pains of 8 hr duration accompanied by vomiting. The pain radiated to the back and right lower abdomen. Over the past 2 years the patient had lost approximately 30 kg and had recurrent episodes of atrial fibrillation. On admission she was pale and sweating with a blood pressure of 90/60 mmHg and sinus tachycardia of 124/min. Exophthalmos was marked and an enlarged thyroid was palpable. There were decreased breath sounds at the left base. There was rebound tenderness and guarding in the right abdomen. The patient's Hb was 10.5 g/dl with a haematocrit of 21%. The leucocyte count was $20.7 \times 10^9/l$, platelet count $110 \times 10^9/l$. Glucose, urea, diastase, prothrombin and urine

tests were normal. The ECG showed sinus tachycardia. A plain abdominal X-ray revealed absence of the right psoas shadow. The IVP was normal but the right kidney was displaced upwards by a retroperitoneal mass which caused an area of depletion in the lower pole. Despite blood and electrolyte transfusion, the patient's condition deteriorated. Her blood pressure dropped to 75/70 mmHg and the pulse rate rose to 140/min. Therefore, an urgent laparotomy was performed under general anaesthesia. Fresh blood was found in the abdominal cavity. In the right retroperitoneum a fatty haemorrhagic necrotic mass was detected attached to the kidney and the suprarenal gland, all surrounded by blood clot. The bleeding was controlled. The right kidney with the attached necrotic fatty mass and suprarenal gland was removed. Excluding episodes of atrial fibrillation, the postoperative period was uneventful. Endocrinological investigations, including elevated T3, T4 and high value of the ^{131}I uptake, indicated hyperthyroidism. The patient was discharged in good condition and advised to receive further treatment with carbimazole. Following appropriate treatment for 6 months, her general condition improved and she gained 8 kg. At follow-up, no neurological or X-ray signs of tuberose sclerosis were observed.

Histology

The kidney weighed 380 g. At the lower pole a spherical, lobulated yellowish-grey tumour of 8 cm in diameter was found (Fig. 1). No capsule was apparent on gross inspection. The cut section disclosed extensive foci of haemorrhagic discoloration and areas of softening. Microscopically, the tumour was highly cellular and composed of 3 main components (Fig. 2): areas of typical vacuolated adult fat cells, demonstrable only by the thin rim of cell membrane separating them from adjacent cells; spindle smooth muscle cells; and numerous

FIG. 1. Right nephrectomy specimen: A, kidney; B, attached angiomyolipoma; C, suprarenal gland; D, haemorrhagic perirenal adipose tissue.

thick walled, tortuous blood vessels surrounded by large numbers of smooth muscle cells which extended into the fat tissue. The tumour was sharply demarcated from the adjacent renal tissue. Vascular invasion was not found.

Discussion

Renal hamartoma is a mesenchymal tumour composed of a mixture of blood vessels, smooth muscle and adipose tissue in varying proportions; thus the name angiomyolipoma. Tuberose sclerosis associated

with renal hamartomas was first described by Bourneville in 1880 (Vasko *et al.*, 1965). It is caused by an autosomal dominant hereditary gene and is characterized by epilepsy, mental retardation and skin manifestation (adenoma sebaceum cyst). According to various authors, 50–80% of patients with tuberose sclerosis have additional renal hamartomas (McCullough *et al.*, 1971; Becker *et al.*, 1973; Honey and Honey, 1977).

Hamartomas associated with tuberose sclerosis are often found in various organs; chiefly in the

FIG. 2. Renal tissue (left upper corner) and the 3 main components of the tumour are seen: mature fat cells, thick walled blood vessels and smooth muscle fibres (HE, $\times 24$).

kidneys, chest, heart and skeletal bones. They are mostly small, multiple, multifocal and involve both kidneys. In patients without tuberose sclerosis they are usually unilateral, solitary and relatively larger (Becker *et al.*, 1973). Histologically the two forms are identical (Hajdu and Foote, 1969). Renal hamartomas associated with tuberose sclerosis are found in both sexes and appear at an early age, the average age being 30 years (Becker *et al.*, 1973). Renal hamartomas which are not associated with tuberose sclerosis appear at an average age of 48 years and are more common in women with a ratio of 23 : 4 (Becker *et al.*, 1973; McCullough *et al.*, 1971).

Seabury, Ensor and Wolfe (1967) compiled from the literature 150 cases of angiomyolipomas without tuberose sclerosis. Vasko *et al.* (1965) discovered that 15.3% of the cases were symptomatic.

The diagnosis of angiomyolipoma is difficult. Pain, haematuria, a palpable mass and continuous fever assist the diagnosis. However, when bleeding and signs of an acute abdomen appear, the diagnosis is quite impossible. The main complaint is flank pain which is found in 73.7–91.3% of the cases (McCul-

lough *et al.*, 1971; Klapproth, Poutasse and Hazard, 1959; Hajdu and Foote, 1969). A palpable mass was found in 33–47% of the cases (Price and Mostofi, 1965; McCullough *et al.*, 1971). Macrohaematuria usually appears with solitary tumours and is found in 16–40% of the patients (Becker *et al.*, 1973; Hajdu and Foote, 1969; Price and Mostofi, 1965; McCullough, 1974). Necrosis in the hamartoma may result in continuous fever (Campbell *et al.*, 1974).

A fatty mass which can be seen in a plain abdominal X-ray is, in 9% of cases, suggestive of an angiomyolipoma. In 6% of the cases calcification can be found (Becker *et al.*, 1973); IVP, sonography and selective renal angiography are not sufficiently diagnostic and may confirm only that this mass is not cystic (Lee, 1977; Becker *et al.*, 1973). The differential diagnosis of an angiomyolipoma and malignant tumour of the kidney is difficult. However, 26 cases of hamartomas without tuberose sclerosis were diagnosed by selective renal angiography (McCallum, 1975; Smulevitz and Fafreshi, 1977) in which no arterio-venous shunts were found. False negative diagnoses were recorded (Becker *et al.*, 1973; McCullough, 1974; Smulevitz and Fafreshi, 1977).

Treatment

In cases of a solitary unilateral angiomyolipoma unassociated with tuberose sclerosis, removal of the mass and total nephrectomy are indicated. Partial nephrectomy may be possible according to the histology of intra-operative frozen section. Asymptomatic, multiple and small angiomyolipomas associated with tuberose sclerosis should be treated conservatively. Surgical intervention is reserved for cases where the tumour increases in size or is symptomatic. The surgical procedure should be according to the result of histological frozen section. A partial nephrectomy is indicated rather than total nephrectomy and is usually adequate (McCullough, 1974). Exploration of the retroperitoneum and frozen section study of the mass is indicated when malignancy is suspected. Ruptured angiomyolipoma, as in the case presented, requires total nephrectomy and removal of the attached fatty mass.

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