RENAL ANGIOMYOLIPOMA CAUSING FATAL RETROPERITONEAL HEMORRHAGE (A Case Report)

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Introduction

Angiomyolipoma is a specific and probably hamartomatous growth arising in the kidney [1]. Usual symptoms include abdominal pain, haematuria, chills and fever [2-4]. Hypertension has also been observed in cases of angiomyolipoma [5]. In this case report, we present an adult male who reported with agonising pain abdomen, was treated as acute pancreatitis and died. Post-mortem revealed a large retroperitoneal hematoma arising from an angiomyolipoma in the right kidney.

CASE REPORT

A 52-year-old male reported with acute pain in the epigastrium which later localised to the right side of the abdomen. Pain was agonising and non-colicky. He gave history of poorly controlled hypertension (with blood pressures ranging from 200-220 mm of Hg systolic and 140-180 mm of Hg diastolic) and was on regular antihypertensive medication (nifedipine, Inderal and Dytilde). On examination blood pressure was 120/80 mm of Hg, pulse 68 per minute. There was gross pallor. Abdomen showed guarding and rigidity in the right epigastric region. He was diagnosed as acute peritonitis. Being a peripheral hospital, neither ultrasonography nor angiographic facilities were available and patient was prepared and taken up for surgery. On laparotomy, the abdomen was seen to be filled with serosanguinous fluid and omentum was adherent to the posterior abdominal wall around the head of the pancreas. Peroperatively, patient suffered cardiac arrest and was revived by internal cardiac massage. In spite of active management his condition remained poor, followed a downhill course, and he expired on the same day.

At autopsy, external examination did not reveal shagreen patches, adenoma sebaceum or any stigmata suggestive of the tuberous sclerosis complex. Abdominal cavity was seen to be filled with hemorrhagic fluid. There

was a large retroperitoneal hematoma on the right side which weighed 2000 gm. It encompassed the great vessels, ureters, head of the pancreas and extended into the mesentery of the small intestine. The left kidney weighed 150 gm, while the right weighed 200 gm. The hematoma arose from a large gray tumor measuring 6 cm x 7 cm on the anterior surface of the right kidney. The tumor surface showed large, dilated and eroded vessels. The growth was well demarcated with a surrounding rim of renal tissue. It did not compress the renal artery. The cut section was gray-yellow. The aortic and paraortic lymph nodes appeared normal. There was no evidence of invasion into the renal vein.

Histopathology showed the tumor to be composed of blood vessels, spindle cells and mature lipocytes. The blood vessels were thick walled, with wide variation in luminal diameter, tortuous, showing an angiomatous arrangement with increased number of smooth muscle cells in the wall. Spindle cells arose from the blood vessel wall and radiated from it in a whorling pattern into the surrounding mature lipocytes (Fig 1). Some of the spindle cells showed hyperchromatic nuclei. Occasional mitoses were seen (Fig 2). The tumor was unencapsulated

Fig. 1 : The section shows thick walled blood vessels with numerous perithelial spindle cells, mature fat cells, and numerous spindle cells (H&E x 100).

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Fig. 2 : The spindle cells are plump with dark hyperchromatic nuclei (H&E x 400).

with no obvious compression of surrounding renal parenchyma (Fig 3).

The heart weighed 400 gm and left ventricular thickness was 2.5 cm. Microsections showed myocyte enlargement with increased transverse diameter. Adjacent cells varied in size. Nuclei were hyperchromatic. Anterior and posterior descending arteries showed luminal narrowing due to medial thickening. The blood vessels showed medial thickening. Paraortic and aortic lymph nodes showed normal histology. The cause of death was opined as a massive hemorrhage arising from an angiomyolipoma resulting in irreversible shock and death.

Discussion

Renal angiomyolipomas are hamartomatous growths in the kidney. Approximately half the patients of angiomyolipoma present with manifestations of the tuberous sclerosis complex [2] where it manifests at an earlier age with almost equal frequency in both the sexes.

In non-tuberous sclerosis, renal involvement is solitary and unilateral. Females are more commonly affected (80%) [6]. These hamartomas become usually manifest in the fourth to the fifth decade [7]. Renal angiomyolipomas may present as gross haematuria (33%), palpable flank mass (33%), while several reported with massive retroperitoneal or perirenal haemorrhage incidental to tumor rupture [3]. Flank pain has been reported in symptomatic patients (91%). The pain is often severe and agonising and has been ascribed to infarction. The complaint of chronic dull pain was less common [1,7]. Nonspecific features like fever and weight loss have also

Fig. 3 : The tumor margin shows replacement of renal parenchyma by the tumor with no notable reaction in the adjacent kidney tissue (H&E x 100).

been reported [4].

Angiomyolipomas have a characteristic polar location within the renal parenchyma. The tumors are usually small (less than 1 cm in diameter), single and unilateral, though multiple bilateral lesions may be seen. Hajdu et al in a series of 27 cases found 23 to be less than 1 cm diameter [7]. McCullough et al reported tumor size of 6 cm in two cases [2]. Gross appearance of the renal angiomyolipoma depends on the predominant cell type and may appear as yellow, gray and yellow mixture [1,8]. Tumors irrespective of the size were composed of fat, smooth muscle and blood vessels [1,7]. The adipose cells showed a small peripheral nucleus and large cytoplasmic vacuoles. The blood vessels composing the tumor were thick walled and tortuous. The smooth muscle component occasionally showed nuclear hyperchromasia and few mitoses. Distant metastases have not been reported. Regional lymph nodes have been shown to have identical tissue but these have been regarded as multicentricity rather than metastasis [9]. An association between renal angiomyolipomas and hypertension has been noticed [5]. The causes of hypertension in renal tumors has been ascribed to renal artery involvement, external compression of the artery by the tumor, and compression of kidney tissue adjacent to the tumor rendering it partially ischaemic [10]. The present case did not show any evidence of

invasion or compression of the renal artery. Ultrastructure in cases of renal angiomyolipoma have shown dense spherical granules and crystalline rhomboid bodies similar to those found in juxtaglomerular tumors [1]. Levels of renin in the affected renal vein and systemic plasma renin levels have been shown to be increased in patients of angiomyolipoma with hypertension [5].

This patient was a poorly controlled hypertensive on prolonged therapy. He presented with acute agonising pain over the right epigastric region. Perioperatively he was thought to have an acute pancreatitis. Appearance on CT scan and ultrasonography being characteristic could provide a clue to preoperative diagnosis [11]. However in this case the absence of any of the above investigative facilities precluded preoperative diagnosis. Treatment recommended varies from a radical nephrectomy to that of a conservative excision [2].

This case emphasizes the need to re-investigate old poorly controlled hypertensives, to exclude a cause of surgically correctable hypertension.

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