

Adrenal Angiomyolipoma: A Case Report and Review of Literature

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Abstract: Angiomyolipoma of adrenal is extremely rare with few cases reported in literature. It usually presents as incidentiloma. We report a case of a 42 years old male patient presenting with pain abdomen, nausea and vomiting. USG abdomen showed features of acute calculus cholecystitis with right adrenal tumour. CECT abdomen revealed heterogenous non enhancing hypodense fatty lesion in right adrenal gland. Biochemical investigations were negative for functioning adrenal tumour. After conservative management of acute cholecystitis, interval cholecystectomy with resection of adrenal tumour was done. Histopathological examination of adrenal tumour showed features of adrenal angiomyolipoma.

Key words: Adrenal angiomyolipoma, abdomen, CECT, cholecystitis, fatty lesion

INTRODUCTION

Angiomyolipoma is a rare tumour first described by Morgan in 1951 in Kidney (Martignoni *et al.*, 2000). It is a relatively rare, benign tumor that appears in 0.3% of general population and accounts for 3% of solid renal masses (Nelson and Sanda, 2002). It belong to the family of lesions characterized by epithelioid perivascular proliferation of cells and consists in varying proportions of three cell lines; mature fatty tissue, smooth muscle and irregular blood vessels (Frago *et al.*, 2006). Case report of extra-renal angiomyolipoma are rare with fewer than 50 reported cases. Liver is most common site for extra-renal angiomyolipoma (Sajima *et al.*, 1999). It has also been reported in retro-peritoneal soft tissue (Friis and Hjortrup, 1982), colon (Abdulkader *et al.*, 2005), spleen (Hulbert and Graf, 1983), lung (Guinee *et al.*, 1995), heart (Shimizu *et al.*, 1994). Only few cases of adrenal angiomyolipoma are reported in literature (Sutter *et al.*, 2007; Lam and Lo, 2001; Godara *et al.*, 2007; Kim *et al.*, 2007).

CASE REPORT

A 42 years old male patient presented in emergency department with pain in upper abdomen, nausea and vomiting for 1 day. There was history of dyspepsia and recurrent abdominal pain in past. On physical examination, there was abdominal guarding and tenderness in right hypochondrium without any palpable lump. Total Leucocyte count was raised (18000/cumm). Urgent abdominal ultrasound (USG) showed echorefective multiple calculi in gallbladder with gallbladder wall oedema



Fig. 1: USG abdomen showing pericholecystic oedem, echorefective multiple calculi in gall bladder and hyperechoic space occupying lesion in right adrenal

and pericholecystic fluid. In addition to features of acute calculus cholecystitis, there was a hyperechoic mass lesion in right adrenal gland, measuring 70×63 mm (Fig. 1). Contrast Enhanced Computerized Tomography (CECT) abdomen was done which confirmed the diagnosis of acute calculus cholecystitis and also showed heterogenous non enhancing hypodense lesion measuring 8.0×5.5×4.5 cm in right adrenal gland consistent with fatty tissue (Fig. 2).

Patient was managed conservatively with nil-per-mouth, intravenous fluids, antibiotics and analgesics, etc., his pain subsided. Biochemical investigations for adrenal



Fig. 2: CECT abdomen showing heterogenous non enhancing hypodense lesion measuring 8.0x5.5x4.5 cm in right adrenal consistent with fatty tissue

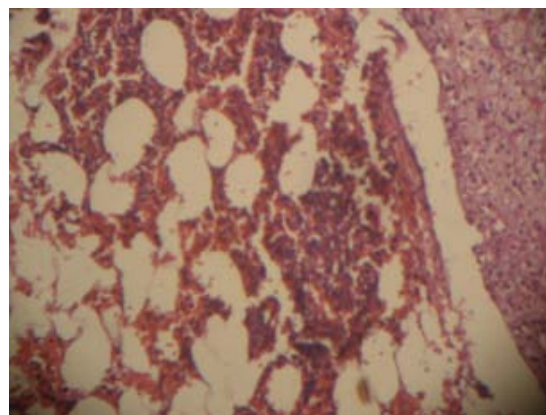


Fig. 4: Photomicrograph of tumour showing adipose tissue, blood vessels and smooth muscles

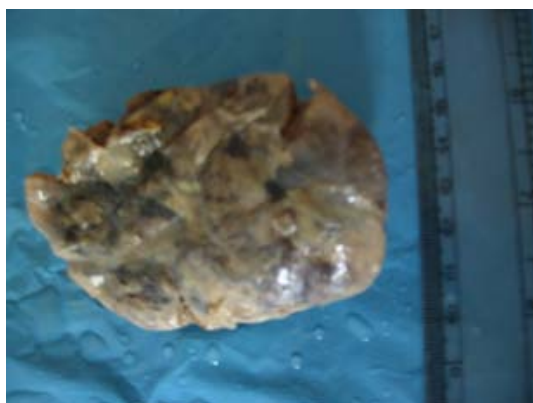


Fig. 3: Grayish white, solid, smooth, firm and globular adrenal tumour revealed measuring 8.0x5.5x5.0 cm

tumour such as urinary Vinyl Mandelic Acid (VMA), serum catecholamines and cortisol were within normal limits, the diagnosis of calculus cholecystitis with non functioning right adrenal incidentiloma, possibly angiomyolipoma was made. On laparotomy gall stones were present for which cholecystectomy was done. A well encapsulated, firm, globular mass, separated from right kidney and involving right adrenal gland was found which was excised.

Histopathological examination of gall bladder revealed features of chronic cholecystitis. Post operative gross evaluation of adrenal tumour revealed a grayish white, solid, smooth, firm and globular mass measuring 8.0x5.5x5.0 cm (Fig. 3). Microscopic histopathological examination revealed adipose tissue, blood vessels and smooth muscles, suggestive of adrenal angiomyolipoma (Fig. 4).

DISCUSSION

Angiomyolipoma is a rare, well-known soft tissue tumour involving the kidneys, liver and other organs. It was previously considered to be a hamartoma or choristoma but now known to be neoplastic (Eble, 1998). They are part of a group of tumours with a diverse appearance and evidence of dual melanocytic and smooth muscle differentiation known as PEComas (tumours of Perivascular Epithelioid Cell origin). The typical lesion is composed of an admixture in varying proportions of thick walled blood vessels, bundles of smooth muscle and mature adipose tissue. In recent years and largely through the research of the group of pathologists of Verone (Italy), it has become evident that most distinctive component of this tumour is the HMB-45 positive epithelioid smooth muscle cell (Chan *et al.*, 1993).

Most commonly involved organ is kidney. Two types are described; isolated angiomyolipoma and angiomyolipoma that is associated with tuberous diseases (Rakowski *et al.*, 2006). Isolated angiomyolipoma occurs sporadically; it is often solitary and accounts for 80% of cases. Angiomyolipoma that is associated with tuberous sclerosis accounts for 20% of angiomyolipomas (Cohen, 1992). The male to female ratio for isolated angiomyolipoma ranges from 1:4-1:8 where as it is 1:1-1:2 in case associated with tuberous sclerosis (Dahnert, 1999). Most patients with isolated angiomyolipoma are in the age group of 27-72; the mean age being 43 years. The mean age of patients in the cases which are associated with tuberous sclerosis is 17 years.

Although, angiomyolipoma are considered benign, rare cases that are possibly related to muticentric disease have been reported, regarding extension into renal vein,

Inferior Vena Cava (IVC) or both. Deposits in regional lymph nodes have also been reported (Baert *et al.*, 1995; Ricketts *et al.*, 2008; Williams and Oakes, 2008). Martignoni *et al.* (2000) reported a case of renal angiomyolipoma in 50 years female. About 7 years after surgical removal of the lesion, the patient developed a local recurrence and successive lung and abdominal metastasis that showed morphologic and immunohistochemical features, overlapping those of the epithelioid area of the previously removed angiomyolipoma. Sato *et al.* (2008) reported a case of metastatic angiomyolipoma. A 16 years female was diagnosed to have bilateral renal angiomyolipoma. Left nephrectomy was done. Few years after, the patient died suddenly of cardiac arrest. At autopsy massive necrosis of right kidney tumour was found, invading the IVC, the tumour was not hemorrhagic. Metastatic lesion was identified in the right lung, liver, diaphragm and mesentery.

Extra-renal angiomyolipomas are rare. It has been reported in liver, retroperitoneum, heart, lung, spinal cord, skin, nasal cavity, parotid gland, fallopian tube, vaginal wall, spermatic cord, penis and breast (Damiani *et al.*, 2002). Gastrointestinal involvement is rare with only seven cases reported in colon and 1 each in duodenum, appendix and stomach (Toye and Czarnecki, 2002; Maluf and Dieckgraefe, 1999; Pelz *et al.*, 2003; Chen *et al.*, 2003; Maesawa *et al.*, 1996; Verzaro *et al.*, 1992; Hikasa *et al.*, 1989; Prasad *et al.*, 2000). Only few cases of adrenal angiomyolipoma are reported in literature. Lam and Lo (2001) reviewed adrenal lipomatous tumours in 20 patients (12 men, 8 women) and they accounted for 4.8% of the primary adrenal tumours reported. They found eleven myelolipomas, three lipomas, three teratomas, two angiomyolipoma and one liposarcoma. Godara *et al.* (2007) reported a case of adrenal angiomyolipoma a 45 years old female. The patient presented with non specific epigastric discomfort. Upper GI endoscopy was normal. USG abdomen revealed a well-defined 15×12 cm mass in left adrenal (incidentaloma) which was confirmed on CECT. Histopathological examination of resected specimen revealed adrenal angiomyolipoma (Godara *et al.*, 2007). Kim *et al.* (2007) published a case of right adrenal incidentaloma which was angiomyolipoma. Vasanthakumar reported a case of adrenal angiomyolipoma. The patient presented with complaints of vague abdominal pain of 3 months duration. CECT abdomen revealed a large fat containing mass lesion of approximate size 8×9 cm in the right suprarenal area arising from the lateral limb of adrenal gland. The right kidney was displaced infero laterally due to the mass effect.

Renal angiomyolipoma are commonly found as incidental finding on cross-section imaging, performed for other indications (Wagner *et al.*, 1997). On USG, they usually cast an acoustic shadow and appear homogenous

with reflectivity due to their high fat contents. On CT angiomyolipoma are characteristically well marginated, cortical based, predominantly fatty lesions (-16 to -20 H) with heterogenous soft tissue interspersed thought. On MR imaging, lesion are bright on T-1 weighted images and dark on fat suppressed images. Radiological diagnosis of extra renal angiomyolipoma is difficult because of the rarity of the condition. A lipoma would be more probable diagnosis in the radiological differentiation of a fatty bowel lesion. A high index of suspicion is required for diagnosis.

CONCLUSION

The management of renal angiomyolipoma is widely discussed in the literature. The most widely accepted is the therapeutic algorithm of Oesterling *et al.* (1986) based on clinical presentation, the size of the tumor and bilaterality. Thus in asymptomatic tumors, evaluation with abdominal USG and/or CT-scan, every 6 or 12 months depending on the size of the tumor, greater or less than 4 cm, respectively is necessary. In symptomatic and/or bilateral tumors, selective kidney artery embolization or conservative surgery (nephron sparing) are the treatments of choice. Radical nephrectomy is reserved for those cases with hemodynamic instability due to massive bleeding, large tumors or coexistence with carcinoma in the same kidney. Large angiomyolipomas, even if asymptomatic should be removed for the fear of spontaneous rupture and retroperitoneal haemorrhage. Stolle *et al.* (2006) reported a case of retroperitoneal haemorrhages due to spontaneous rupture of an adrenal angiomyolipoma (Stolle *et al.*, 2006).

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