Adrenal Angiomyolipoma: A Rare Entity

R Godara, M Vashist, S Singla, P Garg, J Sen, S Mathur

Citation

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Abstract

Angiomyolipoma is apparently part of family of neoplasms that derive from perivascular epitheloid cells. It is a rare mesenchymal tumor, usually found in the kidney. Extrarenal angiomyolipoma is uncommon and the most common extrarenal site is the liver. Only two cases of adrenal angiomyolipoma are reported in English literature. The authors wish to add one more case to the world literature. Because of its large size and symptomatic presentation this extremely rare tumor merits documentation.

CASE REPORT

A 45 year old female presented with epigastric discomfort off and on. Upper G.I Endosocopy was normal. Sonography for hepatobiliary system was normal but revealed a well defined 15x12cm mass in the retroperitoneum (incidentaloma). CECT abdomen further defined the mass as of left adrenal origin and a possibility of adrenocortical tumour (Fig - I). Laboratory investigations i.e Serum catecholamine, cortisol and urinary VMA were within normal limits. Exploratory laparotomy revealed 15x12x10cm mass, firm in consistency, quite separate from left kidney with no definable left adrenal gland. On cut section mass was grey-white and non-homogeneous in texture (Fig – II). Histopathological examination revealed mature fat cells, smooth muscle fibres and thin walled blood vessels with peripherally compressed adrenal cortical tissue suggestive of angiomyolipoma of adrenal (Fig – III & IV). The patient made uneventful recovery and was normal at 18 months follow up.

Figure 1

Figure 1: CECT abdomen showing well defined non homogeneous mass separate from left kidney.

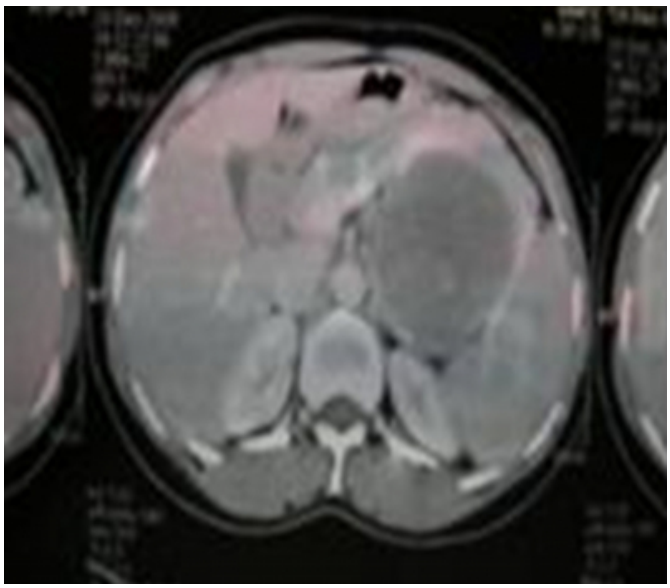


Figure 2

Figure 2: Cut section of removed mass

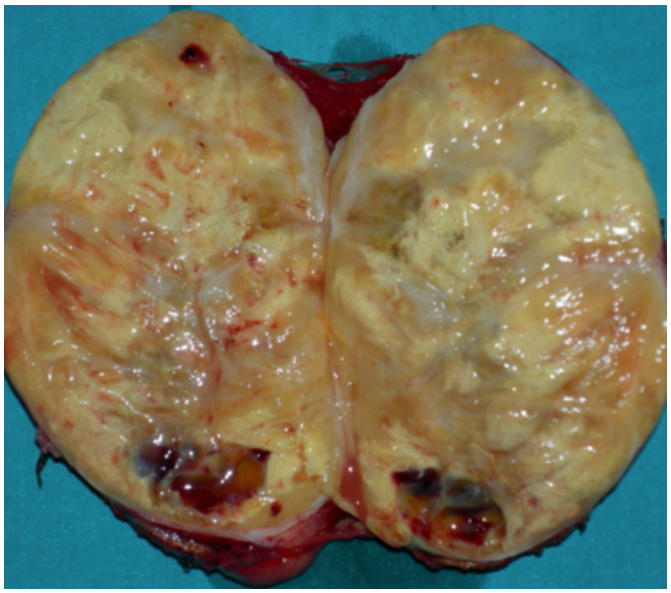


Figure 3

Figure 3: Photomicrograph (H&E staining x 10) revealing features of angiomyolipoma with peripheral compressed adrenal cortical tissue.

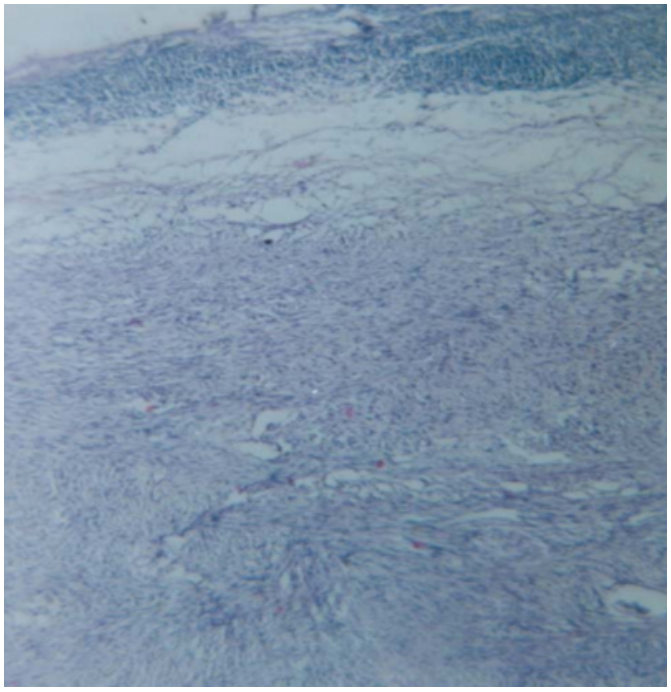
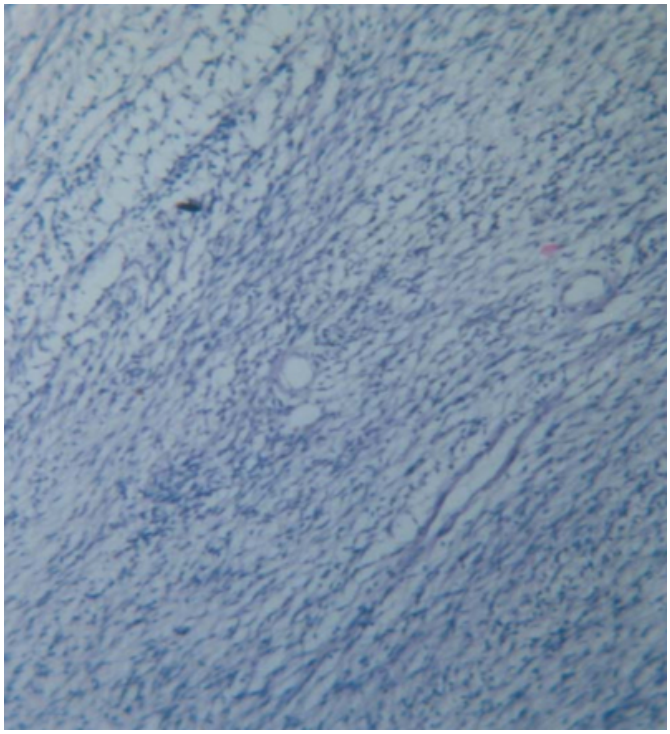


Figure 4

Figure 4: Photomicrograph (H&E staining x 40) showing mature fat cells, smooth muscles & thin wall blood vessels.



DISCUSSION

An increasing number of incidental adrenal lesions have been reported recently, which may be attributable to the increasing use of better imaging techniques. Fatty tumors of the adrenal gland are uncommon and their features have received little attention in the literature. These include myelolipoma, lipomas, teratoma, liposarcoma and angiomyolipoma[1]. Angiomyolipoma of adrenal gland is an extremely uncommon tumour detected incidentally at investigations for other reasons.

Angiomyolipomas are rare lesions, often arising in the kidney, and are part of a group of tumors with a diverse appearance known as PEComas (tumors of perivascular epitheloid cell origin). Angiomyolipoma most commonly occurs in kidney. Next common site is liver. Extrarenal angiomyolipoma are extremely rare and have been reported in liver, colon, suprasellar region, small intestine, skin, intranodal, omentum, breast and adrenal gland[1,2,3,4,6,7]. Adrenal angiomyolipoma is extremely rare and only three cases have been reported, including the present case. One case was reported in setting of tuberous sclerosis and other two were sporadic (one present case). The previous sporadic case was of 8 cm size in a 46 year old female. Both previously reported cases were in left adrenal gland like the

present one. The case in setting of tuberous sclerosis was very small while one sporadic was larger i.e 8cm. In our case the tumor size was 15x12cm, the largest being reported. Angiomyolipomas predominately composed of smooth muscle cells are known diagnostic challenges to pathologists. They are often misdiagnosed as sarcomatoid carcinoma, carcinoma or sarcoma. Some of these tumors have malignant potential and recur locally. A diligent search for adipocytes and abnormal blood vessels may help in confirming the diagnosis.

Large angiomyolipomas even if asymptomatic should be removed to avoid complications like spontaneous rupture owing to presence of abnormal elastin and poor vascularity in the tumour5. Nevertheless, follow up is necessary because of atypical morphology.

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References

1. Abdulkader M, Abercrombie J, McCulloch TA, Kaye PV. J Clin. Colonic angiomyolipoma with a monotypic expression and a predominant epitheloid component. Pathol 2005 Oct; 58 (10): 1107-9.
2. Kahng HC, chin NW, Opitz LM, Pahuja M, Goldberg SL. Cellular angiolipoma of the breast: immunohistochemical study and review of the literature. Breast J. 2002 Jan - Feb; 8 (1): 47 - 9.
3. Kazakov DV, Hes O, Hora M, Sima R, Michel M. Primary intranodal cellular angiolipoma. Int J Surg Pathol 2005 Jan; 13 (1): 99-101.
4. KY Lam, CY Lo Adrenal Lipomatous tumours: a 30 years clinicopathological experience at a single institution. J. Clin. Pathol. 2001; 54: 707-712.
5. Makino E, Yamada J, Tada J, Arata J, Iwatsuki K. Cutaneous angiolipoleiomyoma J Am Acad Dermatol 2006 Jan; 54 (1): 167 - 71.
6. Manner M, Scholz E, Whermann M, Stickel W, Stickel W. Invagination caused by angiolipoma of small intestine - a rare cause of occult gastrointestinal hemorrhage. Chirug 2001 Mar; 72 (3): 305 - 7.
7. Shuangshoti S, Wangsuphachart S. Angiolipoma of suprasellar region. J Med Assoc Thai 1995 Nov; 78 (11): 631-4.

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