Case Report

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A Giant Angiomyolipoma of the Adrenal Gland: A Rare Case Report with Review of Literature

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Abstract

Introduction: Adrenal Angiomyolipoma is rare. Only four cases have been reported so far. These are commonly found in Kidney but extra renal sites are also mentioned. Angiomyolipoma arising in adrenal is very rare entity, usually asymptomatic, diagnosed incidentally on radiological investigation of abdomen for other conditions. *Case Report:* We report our experience with a 55-year-old woman who presented with pain in loin and abdominal mass. A computerised tomography (CT) scan showed an adrenal mass. Exploratory laparotomy with adrenalectomy was performed and the histopathological features confirmed the diagnosis of Adrenal Angiomyolipoma. The patient recovered without any complications following surgery. *Conclusion:* Our review is a brief summary about the adrenal Angiomyolipoma and should be kept in mind while diagnosing a lump in abdomen. Surgery should be considered as the definitive management for larger mass as to avoid associated complications.

Keywords: Adrenal Mass; Extra Renal Tumour; Adrenal Benign Tumour; Abdominal Tumour.

Introduction

Angiomyolipoma is a benign mesenchymal tumour consisting of varying amounts of mature adipose tissue, smooth muscle and thick walled blood vessels [1]. It arises from perivascular epithelioid cells and is commonly seen in the kidney. The extra renal sites reported include the bone, colon, heart, lung, parotid gland, skin, spermatic cord, gynaecologic organs and retroperitoneum with the most common extra renal site being the liver [2]. Only 4 cases of Angiomyolipoma of the adrenals have been reported in the English literature [1-3].

Case Report

A 55-year old male presented with a sudden onset of right loin pain with lump in right lumbar region. Abdominal examination revealed a vague mass in the right hypochondrium. His Ultrasound abdomen revealed a large mass with small calcification in hepatic-renal fossa suggestive of? Exophytic liver mass. A computerised tomography (CT) scan showed a well-defined heterogeneously enhancing large right adrenal myelolipoma mass measuring 24 x 10 x 8 cm with mass effect predominantly on portal vein, hepatic vein, right kidney, colon and head of pancreas but a clear fat plane was observed between these structures. CT angiogram did not show any definite arterial supply to the mass. Serum catecholamine, cortisol and urinary VMA were within normal limits.

Since it was symptomatic and malignancy could not be ruled out, he was advised for excision of the tumour. A laparoscopic adrenalectomy was attempted but the procedure was converted to open surgery due to large size of mass (Figure 1).

A right subcostal incision was then utilised for the adrenalectomy and tumour was excised (Figure 2).

The cut section of the tumour revealed an encapsulated mass with haemorrhagic and yellow necrotic areas within (Figure 3).

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Histopathological examination showed that the tumour was composed of mature adipose tissue, thick walled blood vessels with irregularly arranged smooth muscle cells. Multinucleated giant cells, hemosiderin laden macrophages and golden brownish granules were also present as well as extensive necrosis and areas of haemorrhage.

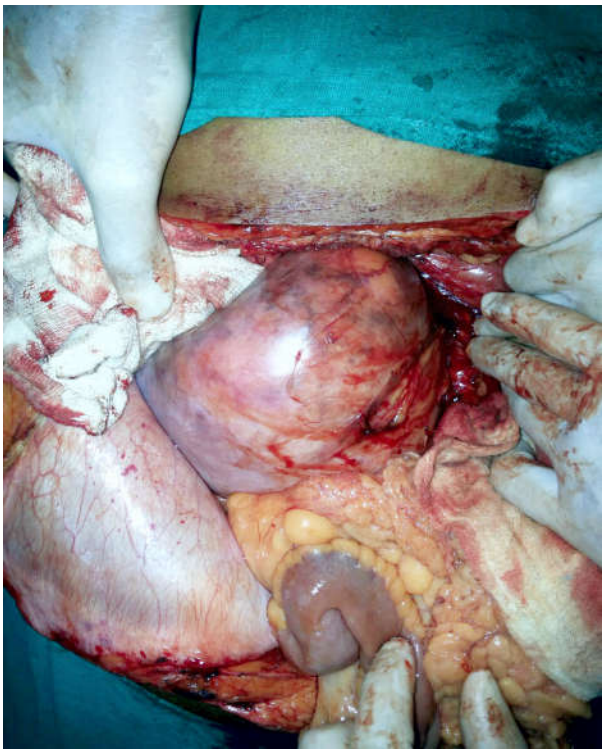

Fig. 1:

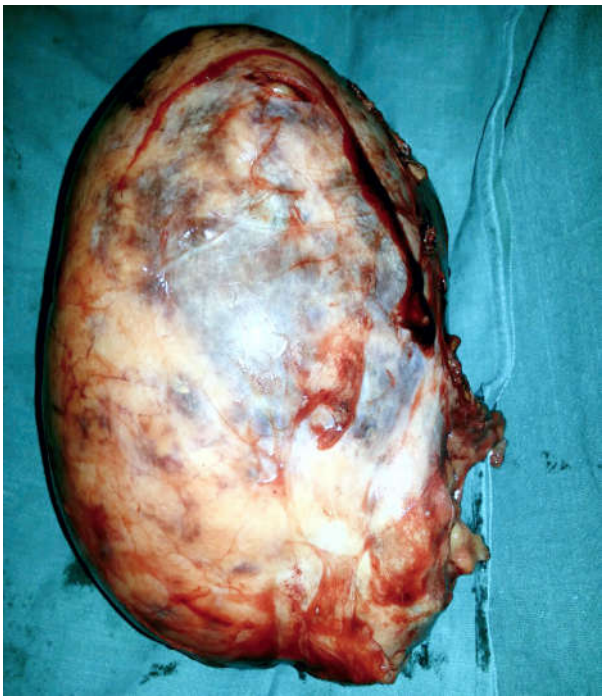

Fig. 2:

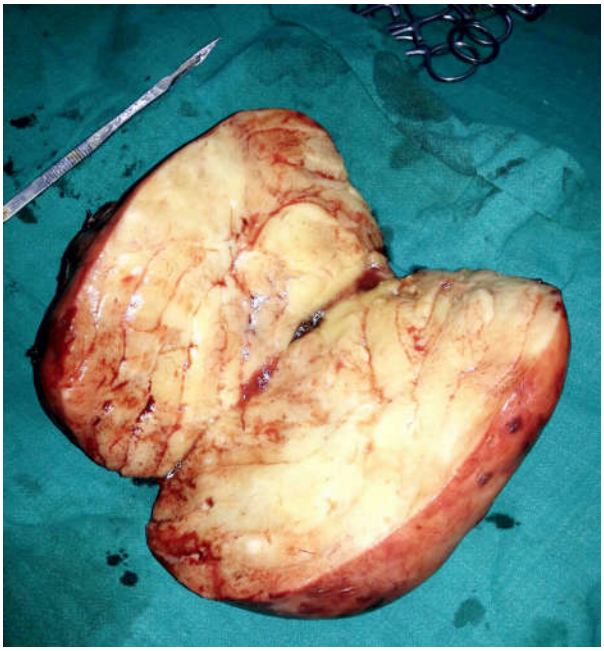

Fig. 3:

Immunohistochemistry were positive for smooth muscle antigen (SMA) and weakly positive for HMB45 and negative for Desmin. Based on these findings, the diagnosis made was Adrenal Angiomyolipoma. Post-operative recovery was uneventful and the patient was well at the one year follow-up.

Discussion

Angiomyolipoma is a benign hamatomatous tumour. About half of the Angiomyolipomas are associated with tuberous sclerosis and in these cases, they are usually multiple and bilateral [4]. It has been estimated that approximately 80% of the severe and complete forms of tuberous sclerosis have angiomyolipoma [5,6]. In the previous reported cases, two were associated with tuberous sclerosis, while the other three, including our case, were sporadic. A case of adrenal Angiomyolipoma in lymphangioleiomyomatosis has also been recently reported [7].

CT and MRI are the diagnostic tool. On CT scan, the presence of even a small amount of fat as evidenced by HU less than 10, suggests the diagnosis of Angiomyolipoma. On magnetic resonance imaging (MRI), the typical features of the fatty component include bright signal intensity on non-fat suppressed images, with dropout of signal on fat suppressed images [1]. However, these features can also be seen with other lipomatous tumours (lipoma, liposarcoma, teratoma or myelolipoma) and are not specific for

Angiomyolipoma. The absence of vascular supply to the tumour in this case initially made the diagnosis of Angiomyolipoma remote. The size of the tumour, despite the clear fat plane between it and the adjacent structures, could not rule out the malignancy.

Grossly, Angiomyolipomas appear well-circumscribed and depending on the relative amount of adipose tissue, they range from a glistening yellow (''fatty'') appearance to a more white-tan and firm appearance depending on the relative amount of adipose tissue [2]. Histology typically shows a variable mixture of mature adipocytes, thick-walled blood vessels and spindled and epithelioid stromal cells often radiating out from blood vessel walls. Most Angiomyolipomas show predominance of adipocytes but some contain mainly spindled stromal cells and thick-walled vessels with little adipose tissue. A diagnostically helpful feature is the staining of the stromal cells for HMB-45 by immunohistochemistry. In this patient, there were areas of haemorrhage and necrosis in the tumour, suggesting an internal haemorrhage which could have caused the acute pain when he presented.

Management should be the same as that for any adrenal mass. Assessment of functional status of the tumour should be done although all the five adrenal Angiomyolipomas reported so far were non-functional. Surgery is indicated if the patient is symptomatic or the tumour is more than 6 cm since the risk of malignancy increases with size [8]. Also, the risk of spontaneous rupture increases with size, owing to the presence of abundant and abnormal elastin-poor vascularity in the tumor [4]. Laparoscopic adrenalectomy is an option and had been successfully done for a 6 cm adrenal Angiomyolipoma but if the tumour size is more than 6 cm open adrenalectomy is ideal to be performed [7].

Since it is a benign disease, its prognosis is good. One of the four reported patients has been alive for eight years [2]. Nevertheless, follow up is recommended because of atypical morphology[3]. Currently, there is no agreed protocol on follow-up but an ultrasound three to six months following the surgery with annual clinical examination for large tumours is recommended.

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