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Histopathology

Primary Adrenal Teratoma- An Unusual Extragonadal Site

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Abstract | **Case Report**

Among mesenchymal neoplasms of Adrenal cortex, Myelolipoma constitutes the most common neoplasm that often diagnosed in patients in their fifth to seventh decade of life. Whereas Teratoma in retroperitoneal location, in Adrenal gland is rare. We present a case of 27year old woman who presented with pain abdomen, found to have a left adrenal mass. On imaging left adrenal mass had defined boundaries and fat predominant component with likely diagnosis of Myelolipoma. She underwent laparoscopic Adrenalectomy with histopathological diagnosis of Benign cystic teratoma. Primary adrenal teratomas are rare neoplasms, more commonly involving left adrenal gland. They are generally amenable to laparoscopic resection and have favorable prognosis.

Keywords: Adrenal teratoma, Histopathology, Imaging, Myelolipoma.

INTRODUCTION

Teratoma is a tumor composed of tissues derived from two or three germ layers (ectoderm, mesoderm and endoderm). These are usually cystic (Mature cystic teratoma), but rarely solid (mature solid teratoma) [1]. Most common sites being Ovaries, testes, sacrococcyx, mediastinum and rarely in retroperitoneum with incidence in Adrenal gland between 0.13 - 3% [2]. As documented in literature, primary adrenal teratomas most commonly found in females, frequently involving left adrenal gland. Most of the patients are asymptomatic or presented with nonspecific symptoms. Mature teratomas are benign neoplasms, although they may develop malignant transformation [3, 4]. The Imaging characteristics of adrenal teratoma are partly like those of myelolipoma, angiomyolipoma and adenoma, making it difficult to diagnose before surgery [5]. Diagnosis is confirmed on histopathology. Thus, surgical resection and follow up recommended in mature teratomas [4].

CASE HISTORY

A 27year old Indian female presented with complaints of pain abdomen and was found to have a left adrenal mass on US abdomen. Her laboratory investigations were normal (including adrenal hormone profile). Contrast CT imaging revealed a well-defined, lobulated fat predominant soft tissue lesion measuring 4.5x3.5x3cms, confined to adrenal gland and diagnosed as myelolipoma. Laparoscopic Left adrenalectomy was done. Grossly the adrenal mass was lobulated with cut surface showing predominantly adipose tissue with interspersed grey white firm areas and a cystic area with calcified wall.

Histologically the representative sections revealed derivatives from all the three germ layers that included mature epidermal structures, respiratory and glandular epithelium, neural tissue with mature ganglion cells, bone and cartilage and skeletal muscle fibers along with the adipose tissue. No areas of immature neuroepithelium or somatic malignancy seen.

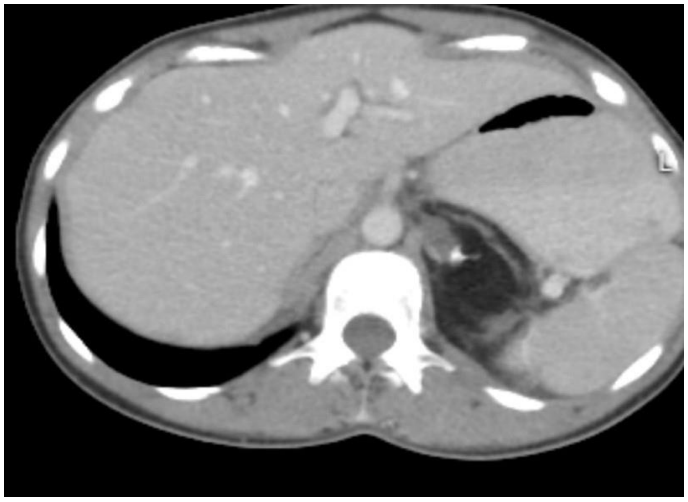


Figure 1: Axial contrast CT scan showed left adrenal heterogenous mass with calcification and fat component

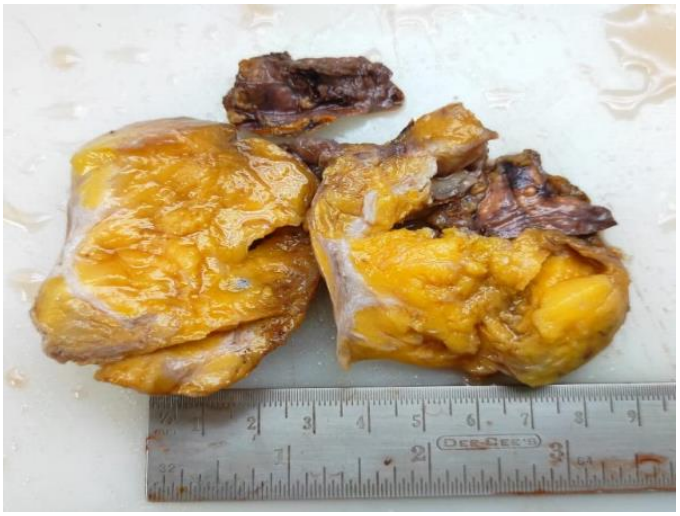
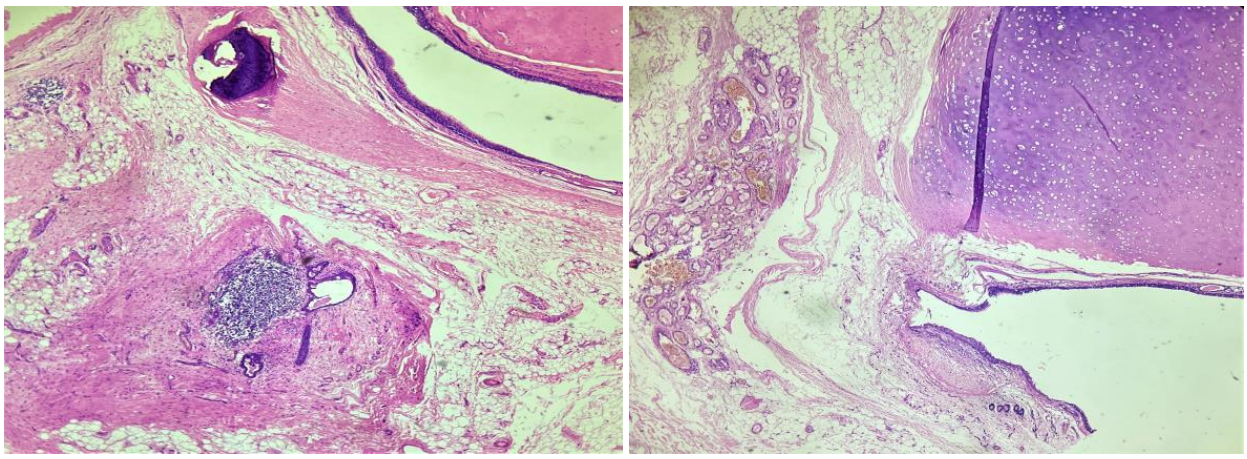


Figure 2: Gross image showing fat predominant mass with a cystic change, grey white firm area and peripherally compressed adrenal tissue



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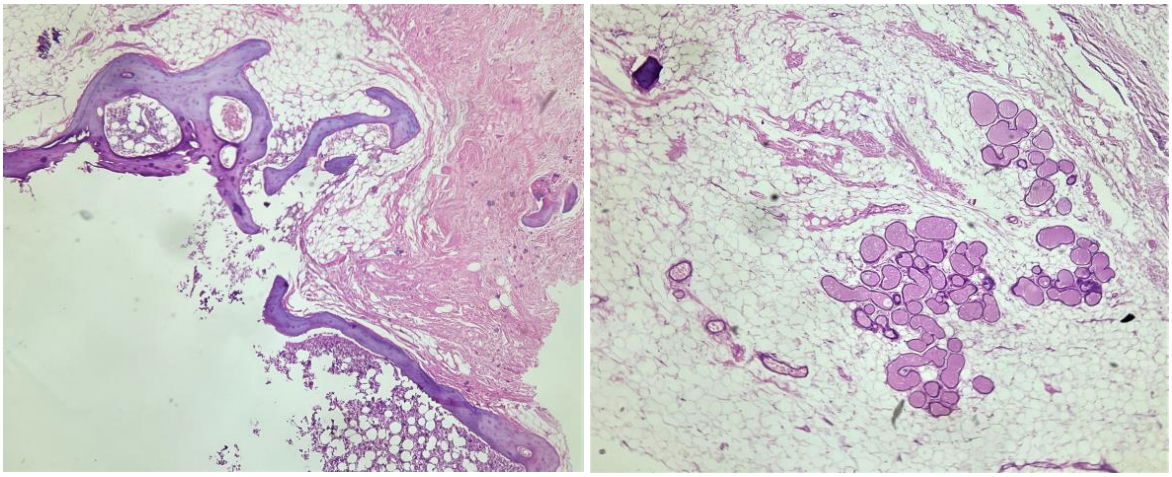

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Figure 3: Microscopic image (A). Respiratory and glandular epithelium with lymphoid aggregates, (B). Mature cartilage with squamous epithelium, (C). Mature bone with marrow elements, (D)Sweat glands (H&E stain).

DISCUSSION

Teratoma is a type of germ cell tumor originating from peripheral pleuripotent stem cells, which composed of tissues derived from the ectoderm, mesoderm and endoderm. The tumor can be solid or cystic. These are predominantly located in gonads, extragonadal sites amounting to 15%, retroperitoneum 4% and adrenal gland being even more rare [6].

The close anatomic association of adrenal cortex with the urogenital apparatus and its embryological development from coelomic epithelium make it a possible site of development of teratoma [7]. Adrenal mature teratomas are generally non-functional and the tumor exhibits latent growth. During early stages the tumor will be nearly asymptomatic. Later, as it increases in size, patient might experience pressure associated symptoms like abdominal discomfort, backpain, nausea, vomiting, urinary retention, intestinal obstruction and sometimes swelling of lower limbs/ genitals due to lymphatic obstruction [8].

CT or MRI findings of adrenal teratoma show fat predominant mass, as like other lipomatous adrenal tumors like myelolipoma, angiomyolipoma or liposarcoma. The typical radiological features of adrenal teratoma are heterogenous mass in adrenal region containing fluid, adipose tissue with calcification [2].

A definitive diagnosis of adrenal teratoma requires histopathological evaluation, which shows tissue elements derived from all the three germ layers. These are further categorized into mature and immature teratomas. Mature teratomas are benign and show differentiated tissues. Immature teratoma composed of undeveloped/ undifferentiated tissues and is classified as benign, possibly malignant or frankly malignant [9].

Surgical excision is usually curative. The goal is the complete removal of tumor without damaging adjacent structures and to relieve clinical symptoms and prevent malignant transformation. Chemotherapy and radiotherapy are recommended only if there is malignant transformation. Overall prognosis is excellent with 5yr survival rate is 100%. A close follow up is recommended in mature as well as immature teratomas [10].

CONCLUSION

In conclusion, primary adrenal teratomas are rare, nonfunctional and preoperative diagnosis would be difficult as it radiologically resembles other common lipomatous adrenal tumors. Histopathological examination of the resected tumor is mandatory for the diagnosis and a careful search for solid areas during grossing is essential for the detection of malignancies as the prognosis differ. Complete surgical excision is the mainstay of treatment with excellent prognosis and overall 5yr survival rate of 100% [10].

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