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Mature teratoma of the adrenal gland

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Abstract: Primary retroperitoneal teratoma in adrenal gland is exceedingly rare in adults. Here we report such a tumor in a 28-years-old-woman. Excision recommended either by open surgery or laparoscopic technique. Since there may be a risk of malignant transformation, close follow up is strongly recommended for a good outcome.
Keywords: Adrenal, Pluripotent, Retroperitoneal, Suprarenal, Teratoma.

INTRODUCTION:

Teratoma is tumor derived from pluripotent cells. They are made up of several parenchymal cell types originating from more than one germ cell layer of the developing embryo [1]. These tumors involve midline (paraxial) structures with the gonads being the commonest sites. Extragonadal teratoma is a rare entity [1]. Teratoma in adrenal region are exceedingly rare [1, 2] and accounts only 4% of all teratoma [3].They are more common in pediatric population and rarely occur in adults [2, 3]. Herein, we report a case of adrenal teratoma in a 28 years old female, who presented with right sided abdominal pain.

CASE REPORT:

A 28-years-old female presented with history of right flank pain for six months. On physical examination, there was a palpable, nontender lump with restricted mobility on the right flank. Routine hematological and renal biochemical parameters were unremarkable. Ultrasonography revealed a mixed echogenic mass in the right suprarenal area. Contrast-enhanced computed tomography (CECT) scan showed a large tumor in the region of right adrenal gland measuring 11cm × 9.1cm × 7.6cm and multiple cystic spaces with thin septations and coarse calcifications (Fig-1).

The huge space-occupying lesion displaced the right kidney inferiorly. The biochemical markers performed to identify a functional status of adrenal tumor, e.g. urinary 17-ketosteroids, plasma and urinary metanephrines and overnight dexamethasone suppression tests, were within normal limits. Laparoscopic adrenalectomy was done. The excised tumor was bagged and removed. The specimen measured 13 × 9.5 × 7.5 cm and weighed137 gm. The postoperative course was uneventful, and the patient was discharged on the fourth postoperative day. Grossly, a partially encapsulated, lobulated gray yellow specimen and cut section showed multilocular cystic spaces lined by whitish-gray wall, filled with pultaceous material. On cut section several hairs were noted within the cyst along with in-between solid fleshy areas with scattered areas of calcification (gritty sensation on cutting) (Fig.-2).

Microscopically, various proportions of mature well-differentiated parenchymal tissues derived from all three germ cell layers were present. No evidence of immature or malignant components was found.(Fig.-3&4) A diagnosis of mature (benign) retroperitoneal teratoma of the right adrenal gland was made. Till one year follow up the patient is fine without any recurrence.

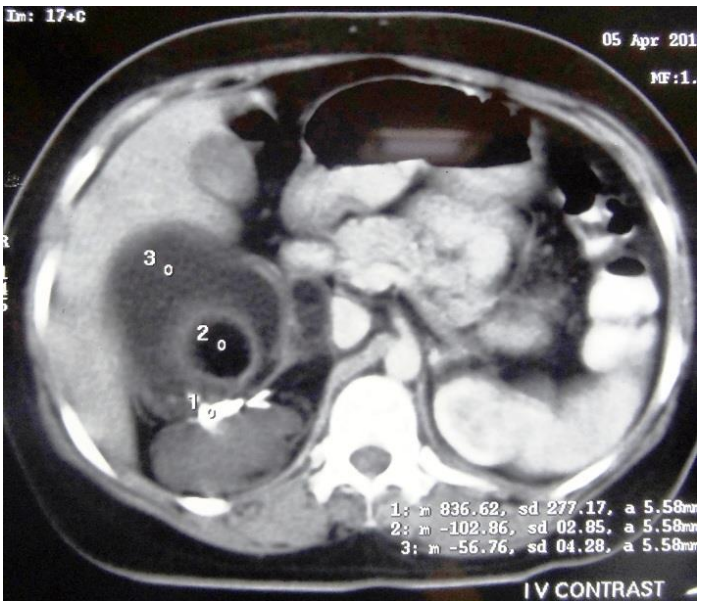

Fig 1: CECT showing a large tumour in right adrenal with multiple cystic spaces with ares of coarse calcifications

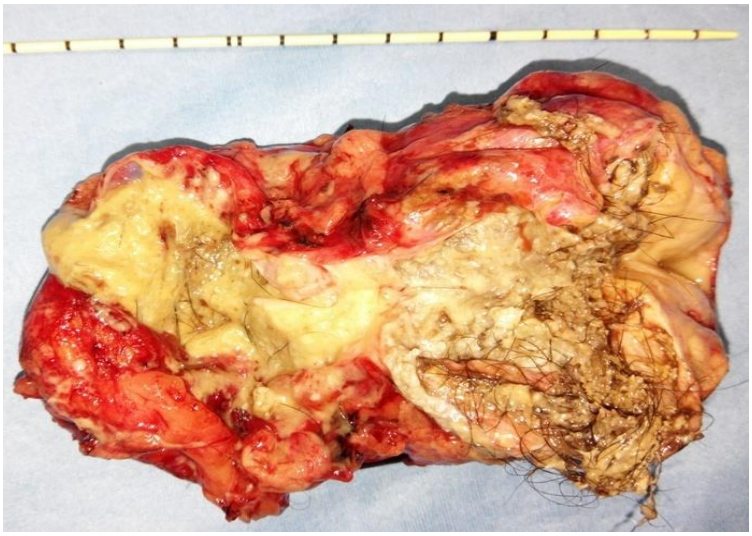

Fig 2: Cut section of the resected specimen showing multiloculated cystic spaces with hairs in between solid fleshy areas with scattered areas of calcification.

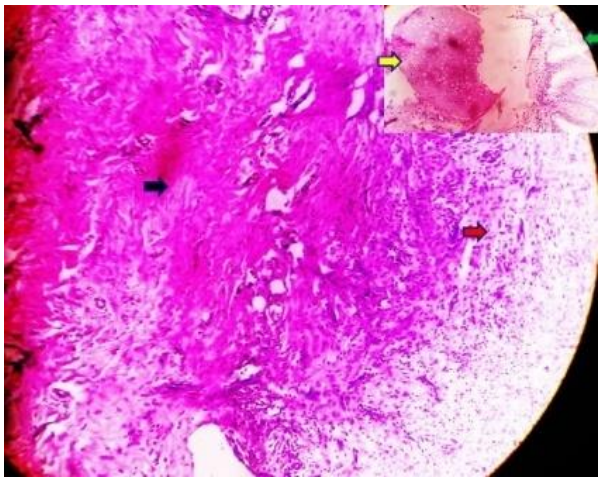

Fig 3: Histopathological picture showing normal adrenal cortex (red arrow), smooth muscle (black arrow), mature cartilage (red arrow) and in inset intestinal mucosal epithelium (green arrow). H & E X 100

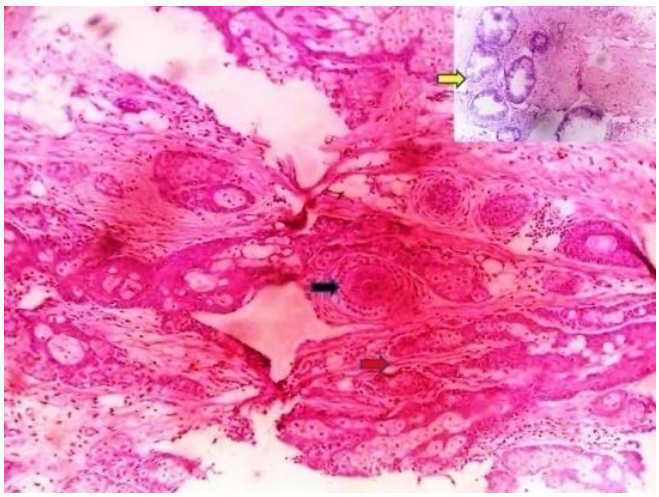

Fig 4: Histopathological picture showing mature ectodermal components like hair follicles (black arrow) and sebaceous gland (red arrow). Areas of intestinal glands (yellow arrow) are shown in the inset. H & E X 100

DISCUSSION:

Teratomas are congenital tumors arising from uncontrolled proliferation of pluripotent cells, either germ cells or embryonal cells [1, 2]. The type of pluripotent cell greatly influences the age of presentation and site of origin. Teratomas originating from germ cell mostly found in gonads (testes and ovaries) which may be congenital or acquired and mainly found in adults. In contrast, teratomas of embryonic cell origin are always congenital and are usually found in extragonadal locations, such as intracranial, cervical, retroperitoneal, mediastinal and sacrococcygeal sites. They are mostly arises in the pediatric population [2, 3]. Teratoma may be mature or immature on basis of the maturation of their tissue constituents. Mature teratomas are made up of well differentiated parenchymal tissue. They are usually benign and commonly found in females. They can be highly variable on histology and can be solid, cystic or mixed in consistency. In contrast, immature teratomas are mostly solid and made up of undifferentiated parenchymal tissue which can be benign or malignant. Any constituent of immature teratomas can undergo malignant transformation [4, 5].The majority of retroperitoneal teratomas are secondary neoplasm with male preponderance. Primary mature teratomas of adrenal origin are exceedingly rare, representing 4% of all primary retroperitoneal teratomas [2, 3], mainly found in the pediatric age groups [1, 3]. Location wise, the tumors have a left-side predilection [1].In our case, the tumor arose from the right adrenal of a young adult female. The majority of retroperitoneal teratomas are asymptomatic and detected incidentally on routine sonography. Sometimes the presentation may be with flank pain, dysuria or lower limb or genital swelling due to lymphatic obstruction [3].Patient may present rarely with complications such as secondary infections with abscess formation, traumatic rupture leading to acute peritonitis, or malignant transformations [4]. Several imaging modalities aid in the diagnosis of teratomas. Ultrasonography (USG) can detect a solid, cystic or mixed echogenic mass but its diagnostic value is limited due to poor identification of fat and calcification which are pathognomonic of teratomas [5]. CT scan can differentiate between fat, proteinaceous fluid and calcific densities leading a better role than USG. The presence of fatty portions of the tumor in the horizontal interface with dependent fluid often clinches the diagnosis of a teratoma. CT can also demonstrate the extent of the tumors well as its relation to the surrounding structures [6]. MRI is superior to both ultrasound and CT as it can accurately define the anatomical relationship between the tumor and the adjacent organs, as well as the local tumor spread. MRI also differentiates the components better and helps in the definitive diagnosis of teratoma [7].Surgery either laparoscopic or open is the mainstay of treatment and the prognosis is excellent with nearly 100% survival after complete excision. Regardless of the benign nature of mature teratomas, malignant transformation is a possibility and, therefore, close follows up is required.

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