

Mature cystic teratoma of adrenal gland: a case report

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Abstract: Primary adrenal teratoma is an exceedingly rare adrenal neoplasm which is more common in infants and children. It is usually considered as a benign neoplasm but malignant transformation is a known complication in teratomas. Thus, it is important to diagnose and treat this condition at the earliest. Diagnosis can be suggested by imaging but a definitive diagnosis requires histopathologic evaluation. Surgical excision is the treatment of choice and prognosis is excellent. We report a case of mature cystic teratoma of right adrenal gland in a 17-month-old female child who presented with swelling in the right flank. Characteristic features of teratoma were demonstrated on imaging and a definitive diagnosis was made on histopathology. The patient was managed with open surgical excision through retroperitoneal approach. There was no complication and the patient was subsequently discharged.

Keywords: Germ cell tumor, Teratoma, adrenal, cyst.

INTRODUCTION

A mature teratoma is a nonseminomatous primary germ cell tumor composed of well differentiated parenchymal tissues that are derived from more than one of the three germ cell layers i.e. ectoderm, endoderm and mesoderm [1, 2]. Teratomas most commonly occur in the gonads but can also occur in extragonadal sites such as intracranial, cervical, mediastinal, retroperitoneal, sacrococcygeal, large bowel and tongue [1, 3]. Most of the retroperitoneal teratomas are secondary to gonadal teratomas [4]. Primary retroperitoneal teratomas are very rare neoplasms accounting for 1-11% of all primary retroperitoneal tumors and occur mostly in neonates, infants and children [1]. Primary teratoma of the adrenal gland is an exceedingly rare neoplasm and accounts for only 4% of all primary teratomas [1, 3]. Among the cases of primary adrenal teratoma documented in the literature, left adrenal gland is found to be more frequently involved than the right and females are more commonly affected than males [5]. Most of the patients are asymptomatic or present with nonspecific complaints [1]. Some of the patients may present with complications such as gastrointestinal or genitourinary obstruction, lymphatic obstruction causing limb or genital swelling or rupture of the teratoma causing acute abdominal pain, ascites or peritonitis [1, 6]. Mature teratomas are considered as benign neoplasms, but malignant transformation into non-germ cell malignancies is known and malignant

teratomas involving adrenal glands have been reported in the literature [2]. Thus, surgical excision and follow-up is recommended in mature teratomas [2, 4]. Diagnosis is confirmed on histopathology but characteristic radiological findings can be demonstrated on all imaging modalities which can suggest a pre-operative diagnosis [2, 6]. The overall prognosis of mature teratoma is excellent with a 5-year survival rate of nearly 100% [1, 6].

CASE REPORT

A 17-month-old female child was brought to our institution by her parents with complaints of swelling in right flank and according to them it had slightly increased in size since they first noticed it few weeks ago. She was a product of non-consanguineous marriage born by normal vaginal delivery at full term. She weighed 10 Kg and her length was 79 cm. Her bowel movements and urine output were satisfactory according to the parents. There was no history of hematuria. Her vitals were stable with a blood pressure of 90/60 mmHg.

On examination, there was a vague, ill-defined swelling in the right lumbar region. It was non-tender and non-mobile. No movement with respiration was seen. Percussion note was dull over the swelling while it was tympanic over rest of the abdomen. Bowel sounds were normally heard and digital rectal examination was unremarkable.

Her laboratory investigations were normal. Liver function and kidney function parameters were within normal range. An ultrasound examination of the abdomen was performed which revealed a large, heterogenous, predominantly cystic mass measuring approximately 8.2 x 5.5 x 3.7 cm, superior to and separate from the right kidney which was displaced inferiorly by the mass. Few echogenic soft tissue components and calcifications were also noted within the mass.

On non-contrast CT scan, a large cystic mass measuring 8.5 x 5.9 x 4.5 cm was seen in the region of right adrenal gland. The right kidney was seen separate from the mass and displaced inferiorly by the mass. Within the cystic mass, fat attenuation areas with

attenuation values ranging from -120 to -180 HU were seen. Dense calcific foci with attenuation values ranging from 400 to 700 HU were also identified within the mass. On post-contrast study, enhancing soft tissue components could also be seen within the mass.

On the basis of the imaging findings, a diagnosis of right adrenal cystic teratoma was made and the patient was taken up for surgical excision. Surgical excision was performed by retroperitoneal approach. A well-defined 8.7 x 4.8 x 5.0 cm mass was excised. Grossly, the mass was lobulated in appearance and rubbery in consistency (Figure 1). Cut section of the mass revealed multi-loculated cystic spaces with interspersed yellow adipose tissue and foci of cartilaginous areas (Figure 2).

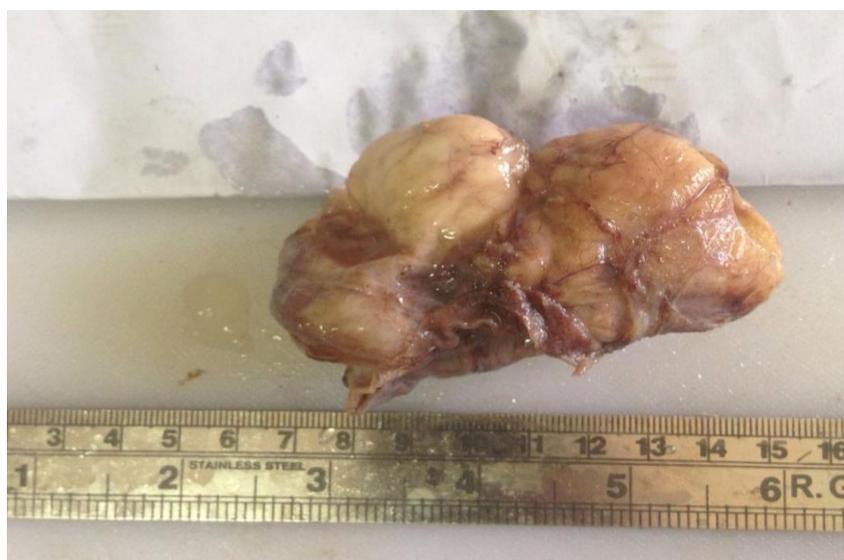


Fig 1: Gross specimen of surgically excised right adrenal mass. The mass appears lobulated in appearance and was rubbery in consistency.



Fig 2: Cut section of the mass shows multi-loculated cystic spaces within and yellowish areas of adipose tissue interspersed in between. Cartilaginous areas can also be identified within the mass.

Histologically, the mass was seen to be lined by stratified squamous epithelium with few interspersed goblet cells (Figure 3). Mature adipocytes were also seen in clusters. Areas of intestinal epithelium and glandular tissue were also identified (Figure 4). Hyaline

cartilage and smooth muscle were also identified within the mass (Figure 5). Neural and glial tissue was also seen. No evidence of dysplasia or anaplasia was seen within the cells.

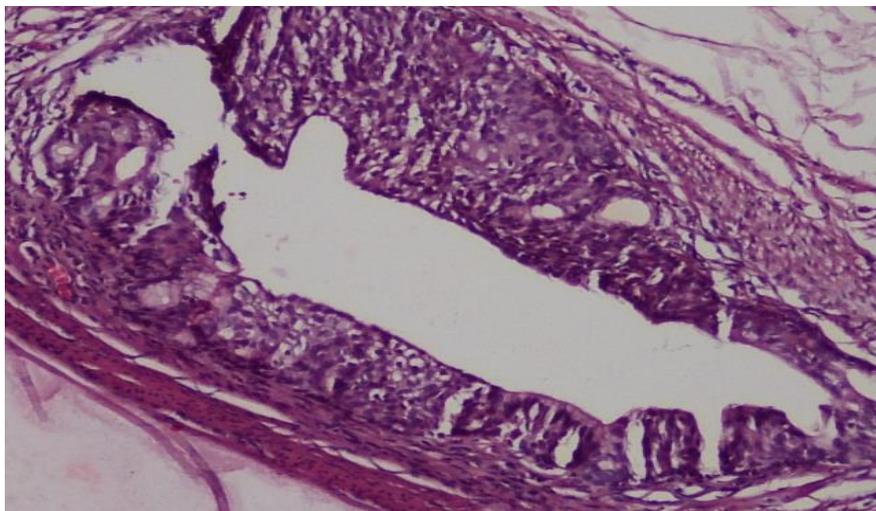


Fig 3: Histopathological examination (H&E stain) of the mass showing stratified squamous epithelium with interspersed goblet cells.

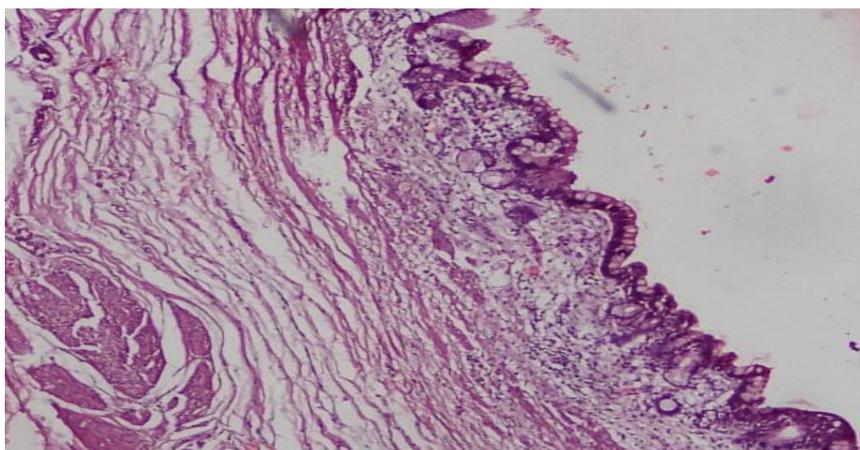


Fig 4: Histopathological examination (H&E stain) from a different area of the mass showing areas of keratinized squamous epithelium, glandular tissue, intestinal type of epithelium and clusters of adipocytes.

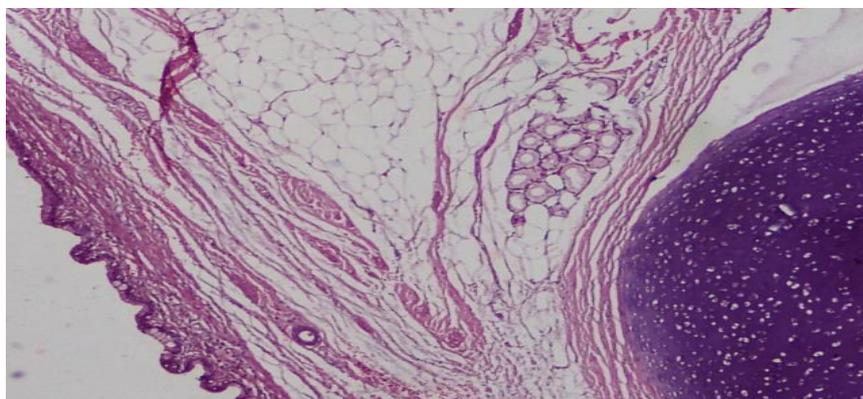


Fig 5: Histopathological examination (H&E stain) of the mass showing areas of hyaline cartilage, smooth muscle, clusters of glandular tissue, adipocytes and intestinal type epithelium.

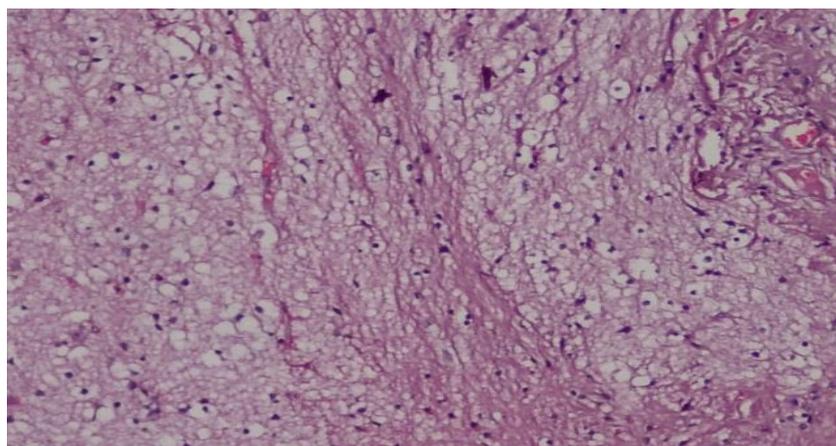


Fig 6: Histopathological examination (H&E stain) showing tissues with neural and glial differentiation within the mass.

A final diagnosis of mature cystic teratoma of right adrenal gland was made. The post-operative period remained uneventful. The patient was subsequently discharged and put on regular outpatient follow-up.

DISCUSSION

Germ cell tumors (GCTs) most frequently occur in gonads and are relatively rare in extragonadal sites such as pineal gland, neurohypophysis, mediastinum and retroperitoneum [7]. The overall incidence of germ cell tumors has been estimated to be about 0.9/100000 population [4]. There are two main categories into which germ cell tumors can be divided: seminomatous and nonseminomatous GCTs [1]. Teratomas are the most common of all nonseminomatous GCTs [1,4]. There are two different origins of extragonadal GCTs based on embryologic and histopathologic consideration: metastasis from gonadal GCTs and primary extragonadal GCTs originating from the pluripotent primordial germ cells which are misplaced during their long trip to the gonads [7]. The time of presentation and location of teratoma is dependent on the type of pluripotent cell from which they develop: teratomas developing from germ cells tend to be gonadal and may be congenital or acquired whereas those developing from embryonal cells are always congenital and usually extragonadal [1]. The usual sites of extragonadal teratoma in descending order of frequency are sacrococcygeal, mediastinal, retroperitoneal and pineal gland [1]. While primary retroperitoneal teratoma is very rare, primary adrenal teratoma is exceedingly rare [1-8]. Li *et al.*; reported a series of five cases of primary adrenal teratoma from March 2009 to February 2014. All the five patients in their case series were females between 16 to 51 years of age with solitary lesions found on routine physical examinations. They concluded that adrenal teratoma does not exhibit typical clinical manifestations or adrenal-related lab abnormalities and pre-operative diagnosis depends on imaging characteristics of heterogenous lesion with fat components and

calcifications [8]. CT and MRI can reliably detect fat within a teratoma [2]. Calcifications may be punctate, shard-like or linear-strand high density. Attenuation of calcification higher than cortical bone is highly suggestive of teeth within the lesion [2]. The differential diagnosis of adrenal teratoma includes other lipomatous masses arising from the adrenal gland such as Myelo lipoma, lipoma, liposarcoma, angiomyolipoma as well as tumors with extensive necrotic component such as pheochromocytoma [2, 8]. A predominantly cystic component with congealed or shard-like calcifications distinguishes mature teratoma from other lipomatous tumors of the adrenal gland [2]. Calcifications can be demonstrated on 62% of plain radiographs [1]. A definitive diagnosis of adrenal teratoma requires histopathological evaluation. Histologically, teratomas contain elements derived from more than one germ cell layer i.e. endoderm, mesoderm and ectoderm and different tissues such as fat, hair, skin and teeth can be seen within a specimen of teratoma [1,2]. A mature teratoma may have partially or completely developed organ systems and is usually benign. An immature teratoma, on the other hand, has undeveloped or undifferentiated tissues and may be benign, possibly malignant or frankly malignant [1]. A group of exceptionally rare teratomas known as teratomas with malignant transformation have a tendency to undergo malignant transformation and have an increased tendency to metastasize. Malignant transformation usually occurs in stratified squamous epithelial components. Somatic malignancies such as carcinoma, sarcoma and leukemias may also occur within a teratoma [1].

Surgery is the method of choice for treatment of mature teratoma [5]. Surgery may be open or laproscopic. Laproscopic approach may be transperitoneal or retroperitoneal [5, 6, 8]. Teratomas are resistant to radiotherapy and chemotherapy and these are recommended only if there is evidence of malignant transformation on histopathological examination [1]. The overall prognosis is excellent with

a 5-year survival rate of nearly 100% [1, 6]. A close follow-up after surgery is recommended in mature as well as immature teratoma [8].

Our case was a 17-month-old girl child who presented with a swelling in right flank. She was otherwise asymptomatic with normal laboratory parameters. A diagnosis of right adrenal teratoma could be established in this case with high diagnostic confidence on the basis of imaging findings. The radiological findings were subsequently confirmed histopathologically and a definitive diagnosis of mature cystic teratoma of right adrenal gland was made. The patient underwent an open surgery through retroperitoneal approach. Considering the age of the patient and relative size of the tumor, laparoscopy was not considered as a safe option in this case. There were no complications and the patient was subsequently put on regular post-operative follow-up.

CONCLUSION

Mature cystic teratoma of adrenal gland is an extremely rare condition with literature limited to few case reports and case series. A relatively confident pre-operative diagnosis can be made on the basis of characteristic imaging findings; however, a definitive diagnosis can be established only on histopathological evaluation. Surgical excision is the treatment of choice for adrenal teratoma. Considering the malignant potential of teratoma, patients should be closely followed in the post-operative period.

ABBREVIATIONS

CT- Computed Tomography
GCT – Germ Cell Tumor
HU – Hounsfield Unit
H&E – Hematoxylin and eosin
MRI – Magnetic Resonance Imaging

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