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PRIMARY ADRENAL MATURE TERATOMA- A RARE PAEDIATRIC TUMOUR

Radiology

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ABSTRACT

Introduction- Primary adrenal Teratoma is rare neoplasm which arises from one or all the three germ layers. Mature teratomas are usually benign but may possess malignant potential, chances of which is lesser in paediatric population than in adults.

Case report- We report a rare case of a 1 year old male infant who presented with abdominal distension, more on right side. Abdominal X-ray showed soft tissue opacity in right hypochondriac region with calcification for which Computed tomography done which showed an adrenal mass for which patient underwent major surgery at GCRI.

Conclusion- Diagnosis of Adrenal teratoma can be made radiologically with characteristic findings on CECT. Definitive treatment is surgical removal with excellent prognosis.

KEYWORDS

Mature adrenal teratoma, Extragonadal teratoma, Germ layers

Introduction

Incidence of germ cell tumors and of these, teratoma has been reported as the leading fetal and neonatal neoplasm (1, 2). Teratomas are unusual tumors that are derived from totipotent cells with their origin from more than one or usually all three germ cells, i.e., ectoderm, mesoderm, and endoderm incidence of which has been estimated at about 0.9/100,000 population (3). Most common sites are gonads, sacrococcygeal, mediastinal, and pineal region (4). Teratomas that occur in infancy and early childhood are usually extragonadal. Most teratomas in retroperitoneal region are secondary to germ cell tumors

of the testicles or ovaries. Specifically, in male patients, retroperitoneal germ cell tumors are more likely to have metastasized from the testes than to present as primary tumors (2). We report the case of a primary adrenal mature teratoma presenting in an infant who was treated at GCRI.

Case Report

A male infant born by normal vaginal delivery was found to have abdominal distension for 3 months, more on right side. There was no history suggestive of any adrenal, liver and renal dysfunction or family history related to familial syndromes. On physical examination, general condition of patient was good and he was normotensive. Dullness is noted on right side of abdomen compared to tympany on left side. For which Abdominal X-Ray done which showed soft tissue opacity with few calcific foci in right hypochondriac region (Figure 1). Patient underwent CECT examination which showed large homogenous mass in right adrenal measuring 4.4x7.4 cm with few cystic areas, fat content and calcifications (Figure 2). Mass was well defined, homogenous in architecture, capsule was intact, do not infiltrates adjacent liver, kidney or vascular structures supports benign nature of mass. Fat and calcification favours diagnosis of mature adrenal teratoma with high certainty. Based on radiological diagnosis surgical resection was carried out. The postoperative period was uneventful, and he was discharged on 5th postoperative day.

The specimen was sent for histopathological examination. Mass showed features of all three germ line derivatives. Normal adrenal gland was not identified. No immature component was seen. Hence, the final diagnosis was given as primary mature adrenal teratoma (Figure 3).

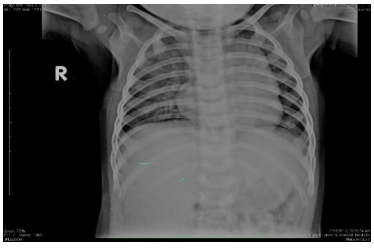

Figure 1- CXR examination shows soft tissue opacity in right hypochondriac region with few (2) foci of calcification.

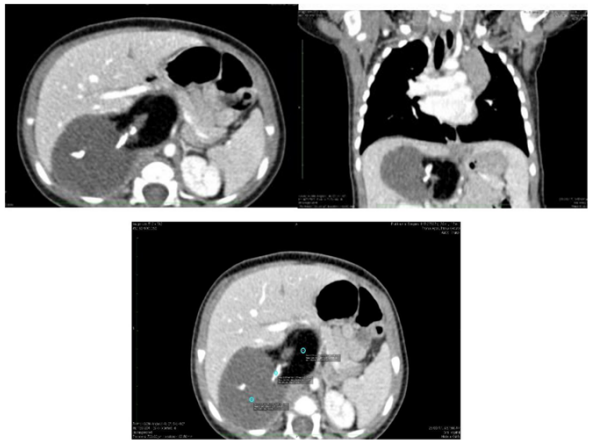

Figure-2 CECT examination shows mixed density lesion (Fat, cystic, soft tissue) in right adrenal gland region, right adrenal gland is not seen separately from lesion.

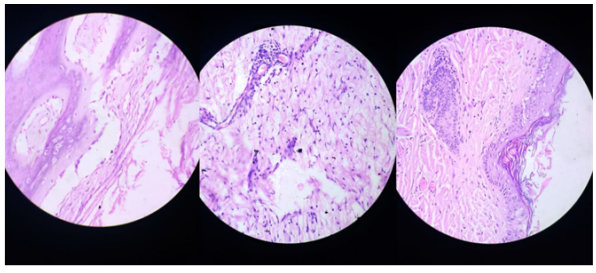

Figure 3- Histopathology specimen shows components of all 3 germ line.

Discussion

Teratoma is neoplasms comprising tissues derived from more than one germ layer. Primary extragonadal teratoma in retroperitoneal locations are very rare and constitute only 4% of primary teratomas.

Most patients present with an incidentally detected non-functional solitary adrenal mass. Sometimes there may be vague symptoms such as abdominal distension, dull flank/epigastric pain, or it may also present as intestinal obstruction caused by compression (5).

Radiographic investigations play an important role in diagnosing teratomas. CT/magnetic resonance imaging scans can better distinguish between fat (adipose tissue) and bone (calcified) masses (6),(7),(8). Plain abdominal film shows a calcification. The diagnosis of adrenal teratoma relies predominantly on an imaging examination because the findings from laboratory examinations will often be normal.

A typical mature teratoma often contains components from each of the three germ layers, including lipid, epithelium, bone, cartilage, hair, fat, muscle and nerve tissue.

Due to fat contents and homogenous density mass diagnosis of adrenal teratoma can easily made on radiological finding. Biopsy usually not needed and patient can directly undergo excision.

The majority of mature teratomas in the retroperitoneum are benign neoplasms, and 26% are malignant. The diagnostic criteria in the present case are completely in accordance with the benign mature teratoma. The malignancy rate of 6.8% documented in children

(9),(10). Surgical excision is the method of choice for the diagnosis as well as treatment of the mature teratoma (10). We did open transperitoneal adrenalectomy. Prognosis is usually excellent after complete resection of mature teratoma. Follow up is only required when specimen biopsy suggest any immature component.

Conclusion

To conclude the diagnosis of mature teratoma can be established by its characteristic appearance on radiologic evaluation. The definitive treatment for these neoplasms is surgical resection.

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