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A Rare Case Of Primary Adrenal Teratoma

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Abstract: Teratomas are tumours of the germ cells, primarily of the gonadal origin. Mediastinal, sacro-coccygeal, and pineal areas are additional typical extragonadal locations.The primary adrenal teratoma is a rare lesion which is often misdiagnosed. We reported a case of Primary Adrenal Teratoma in a 3yr old female child.

Keywords: Teratoma, Retroperitoneal, Germ cell tumour. Adrenal

Introduction

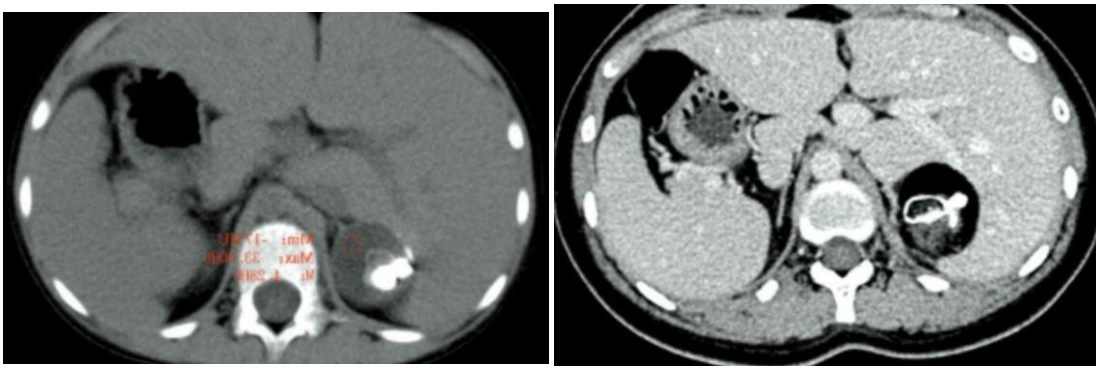
Teratomas are congenital tumours thought to arise from pluripotent embryonal cells and contained tissues derived from two or more than two germ layers. Teratomas are most commonly found in paraxial and midline locations, but they can occur in almost any region of the body. Primary adrenal retroperitoneal teratomas are uncommon, accounting for only less than 4% of all primary teratomas

Case History

3yr old female child Was brought in by her mother who incidentally detected a mass per abdomen

Imaging Investigations

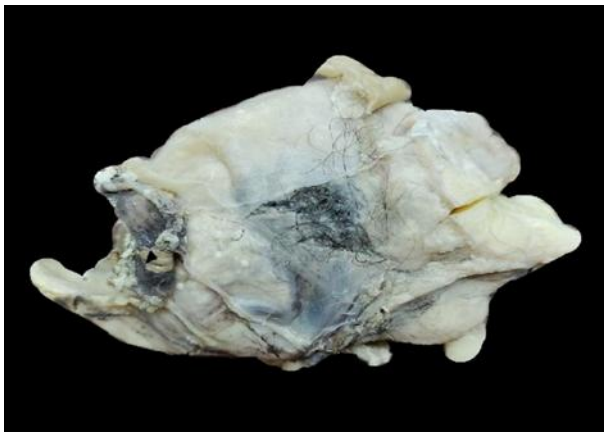
A cystic mass in the the (R) Retroperitoneal region, arising from Right Suprarenal gland-Mixed density lesion with fat, bone and other soft tissue densities along with calcifications



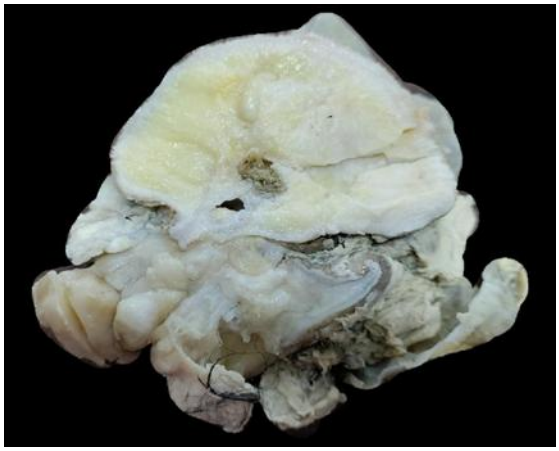
Per Operative Fingings

Lesion arising from Right Suprarenal gland with no fixity to kidney, extending on both sides of IVC.

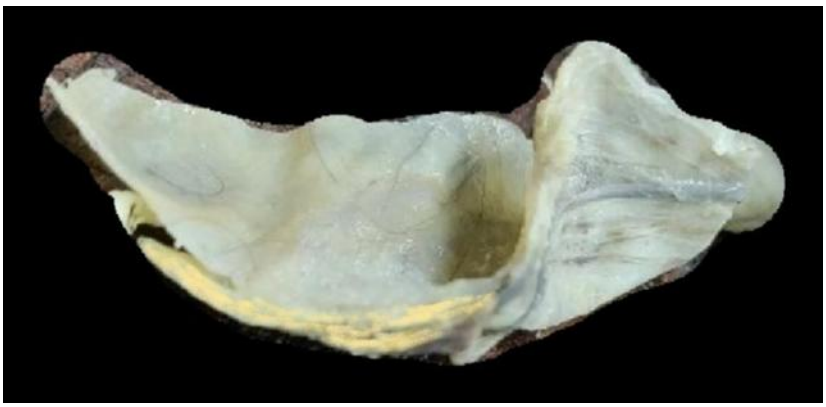
Gross Morphology



Received a nodulocystic mass with Ruptured capsule, weighing 100gms and measuring 12×8×5cm.



Cut section shows a cyst with solid areas showing (1)skin with hair follicle and (2) teeth (3) areas of cartilage (4) bone (5) fat (6) pultaceous material.



Thin cyst wall showing yellowish areas- adrenal tissue

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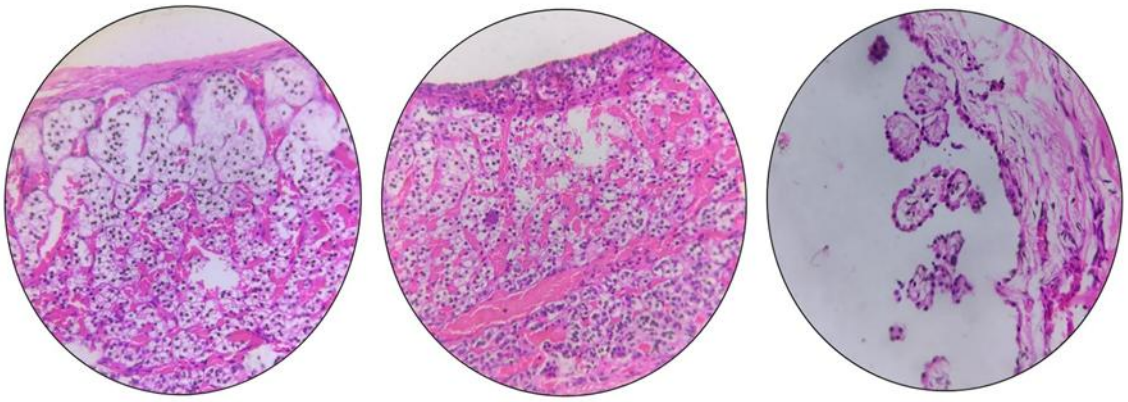
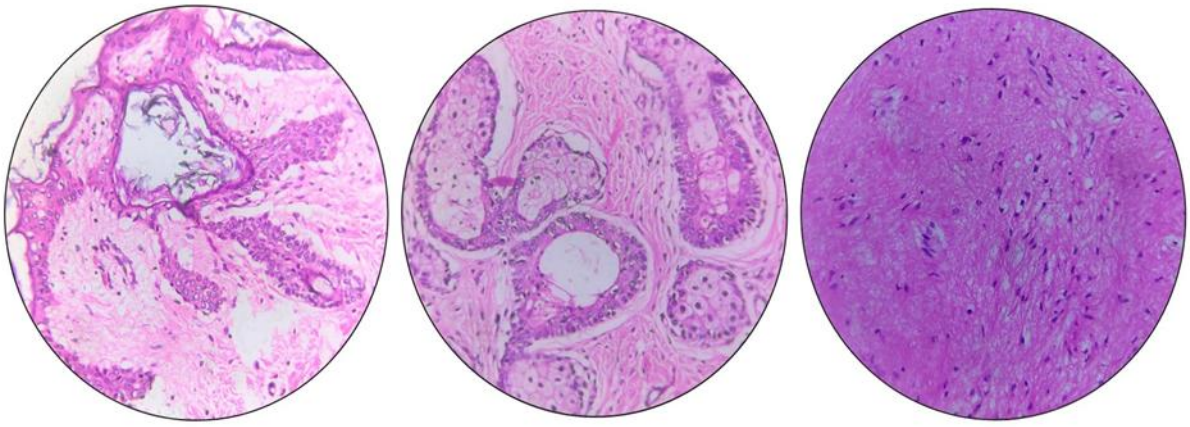
Histo Morphology



Cyst wall showing normal adrenal tissue

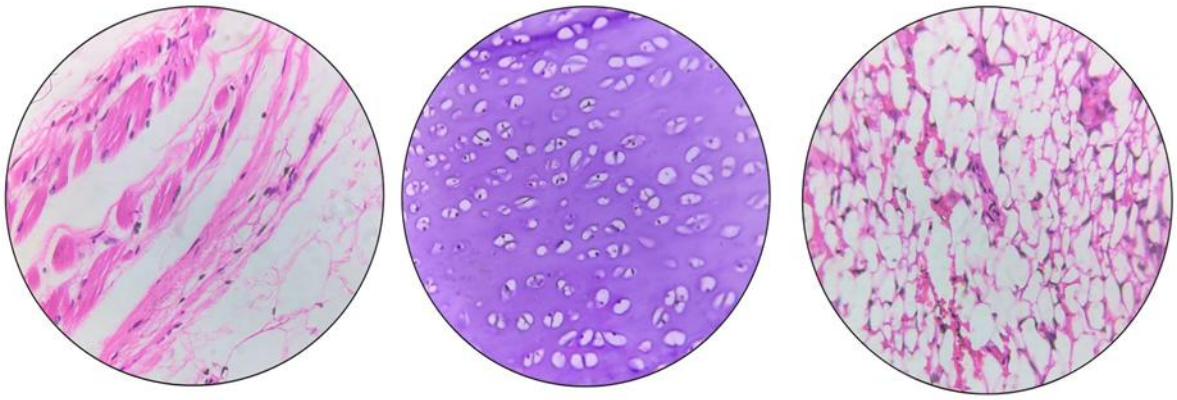
Choroid plexus tissue



Stratified squamous epithelium

Adnexal structures

Glial tissue



Smooth muscle fibres

Cartilage

Adipose tissue

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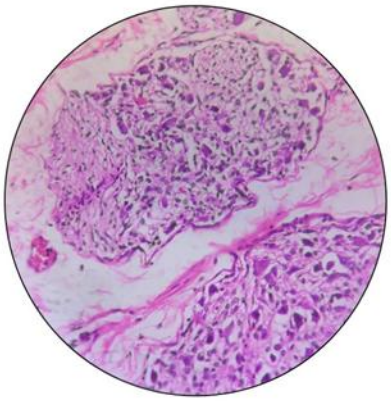
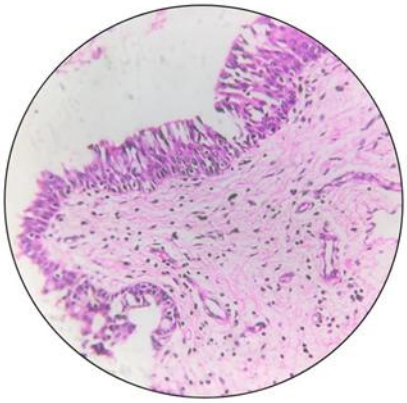
Bone marrow tissue showing maturing hematopoietic elements



Ganglion cells



Pseudostratified ciliated columnar epithelium

1. Section from nodular mass shows adrenal tissue with a neoplasm composed of varying elements derived from all three germ layers. Varying elements include cystic spaces liked by stratified squamous epithelium with luminal keratin flakes.
2. Subepithelium shows adnexal structures.
3. Also identified smooth muscle bundles, nerve bundles, adipocytes, cartilage and bony spicules showing mature hematopoietic elements.
4. Glial tissue surrounded by ependymal cells and choroid plexus tissue noted.
5. Also identified islands of ganglion cells.

Diagnosis

Mature Teratoma, arising from Right Suprarenal gland.

Discussion

Teratomas are encapsulated neoplasms composed of multiple parenchymal tissues (of varying degrees of differentiation) that are derived from more than one germ cell layer (ectoderm, mesoderm, and endoderm)

Adrenal teratomas only accounted for 0.13% of adrenal lesions. Primary adrenal teratomas occur as "incidentalomas" and lack a characteristic clinical presentation with normal adrenal-related hormones. They are also difficult to distinguish from other adrenal tumours without endocrine function.

Pediatric adrenal teratomas are uncommon, representing~5% of all pediatric teratomas

The majority of mature teratomas in the retroperitoneum are benign neoplasms, and 26% are malignant.

The differential diagnosis of retroperitoneal teratoma in infant includes nephroblastoma, neuroblastoma, ovarian tumors and lymphangioma . It can be difficult to distinguish between adrenal teratomas and myelolipomas, angiomyolipomas, or liposarcomas.

While majority of these tumours are benign, malignant transformation may happen more frequently in adults than in children, making it an important entity that needs the proper protocol for therapy. The prognosis is generally good and curative if the tumor is completely removed

At present, laparoscopic surgery remains the primary choice for the treatment of adrenal tumors.

Conclusions

Hence, to conclude primary adrenal teratomas are very rare and should be considered in the differential diagnosis of a retroperitoneal mass. The prognosis is excellent after complete resection; however, an adequate follow-up is advised in view of risk of malignant transformation.

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