

Kidney teratoma in children: A rare entity; A case report and literature review

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Abstract

Teratomas are neoplasms that arise from pluripotent cells and can differentiate along one or more embryonic germ lines. Renal teratoma is a rare condition. Teratomas commonly arise in the gonads, sacrococcygeal region, pineal gland, and retroperitoneum. They present mainly as an abdominal mass with few other symptoms. Majority of the tumors are benign, situated on the left side and para renal, occasional lesions are bilateral. If diagnosed early, they are amenable to curative excision.

Renal teratomas are rare and most have been dismissed as cases of teratoid nephroblastoma or retroperitoneal teratomas secondarily invading the kidney. The differentiation between these two neoplasms in the kidney is often problematic.

We present a case of intrarenal mature teratoma in a five-month-old baby boy, discuss its pathology and review literature.

Keywords: kidney, teratoma, histology, mature, children

Introduction

Teratomas are neoplasms that arise from pluripotent cells and are composed of several types of tissues representing one or more of germinal layers, including ectoderm, endoderm and mesoderm. (1) Most common sites of teratoma are sacrococcygeal, ovary, testicle, pineal gland, and retroperitoneum. Primary renal teratoma is extremely rare. We here describe a case of primary renal mature teratoma in a five-months-old boy.

Case report

A 05-month-old boy was brought by his parents with abdominal swelling of 1 month and abdominal distension. On palpation, a firm mass occupying all of the left of the abdomen was palpable. Renal and liver functions were normal. A chest x-ray showed no abnormality. Abdominal ultrasonography demonstrated internal cystic and solid changes in the left kidney with no abnormalities found in the left kidney. Computed tomography scan of the abdomen revealed a 72x78x85 mm measuring left renal. No tumor in other sites of the body was identified. Preoperative chemotherapy was initiated with Vincristine and Dactinomycin for suspecting a nephroblastoma. A left transversal under umbilical incision was made to find a large encapsulated mass in left pelvic region (Figure1). It was roughly oval in shape. After uretero nephrectomy, the specimen (Figure2) was sent for histopathological examination. The patient had a good recovery after operation. Grossly it was a circumscribed lobulated gray and white solid and cystic mass measuring 11x9x5 cm and weighing 800 g (Figure3). On sectioning a variegated mass with areas of cyst and solid focus was identified. A thin compressed rim of renal tissue was present at periphery of the mass. The tumor consists of skin and its accessories, bowel tissues, lymphoid tissues as well as nerves and bronchia tissues. Thus, our pathological finding was in accordance with mature solid

cystic renal teratoma (Figure 4).

Discussion

Teratomas are rare neoplasms with tissue derivatives of all three germ layers. (2) They most commonly arise in ovaries and testis, but have also been reported in the anterior mediastinum, retroperitoneum, sacro-coccygeal region, brain, gastrointestinal tract and rarely in the kidney. Teratomas are thought to have been present since birth, or even before birth, and are therefore considered as congenital tumors. (3) Renal teratomas include both mature and immature types. Retroperitoneal teratomas exhibit a bimodal presentation, with peaks in the first six months of life and early adulthood. (4) Literature does not reveal a side or gender predilection and almost equal incidence in males and females have been reported. (5)

The clinical symptoms are an abdominal mass, abdominal pain, abdominal discomfort, pyelonephritis, and constipation. (6) Occasionally, the tumor is present antenatally and diagnosed at birth, these neonatal teratomas have a higher incidence of malignancy than those in older children. (7) The diagnostic algorithm was palpation of a solid flank mass, in our case it was a pelvic mass, plain X-ray to demonstrate calcification or formed bony components like teeth and phalanges which are pathognomonic. Ultrasound was sufficient to define the relationships of the tumor for planning surgery. CT scan was used to define the extent of the disease in lesions occupying both sides of the retroperitoneum and those tumors where calcification is not seen on plain X-ray.

In mature teratomas skin with dermal appendages, bronchial structures with bronchial glands and cartilage, neuroglial tissue, and teeth are commonly present and regarded as evidence of organogenesis. In the other hand immature teratoma contained neuroepithelial components with an embryonic appearance and ependymal rosette-like. (8)

For a tumor to be termed a renal teratoma, Beckwith suggested it should meet the following minimal criteria: (A) Primary tumor should be unequivocally of intra renal origin, meaning the entire lesion should be contained within the renal

capsule and there are no teratomas in remote sites which may have metastasized to the kidney. (B) Tumor should exhibit unequivocal heterotopic organogenesis, with clearly recognizable evidence of an attempt to form organs other than kidney. (2)

The differential diagnosis of intrarenal teratoma include Wilms' tumor. (9) Both of these tumors originate from the mesodermal metanephrogenic blastema, and in histological examinations they are similar. Wilms' tumor can contain a variety of heterologous elements with histologic findings of blastemal, stromal, and epithelial cell types. (6) Therefore, a differential diagnosis between intrarenal teratoma and teratoid nephroblastoma is difficult even when making a pathological diagnosis because it can only be made based on a detailed analysis of the tumor after resection. It is therefore highly possible that anticancer drug treatment is required for Wilms' tumor. (6) The second differential diagnosis is congenital mesoblastic nephroma. (10)

Sonographically, classic congenital mesoblastic nephroma may appear as a hypoechogenic tumor with an echogenic rim, but it sometimes may also appear as a heterogenous solid mass like teratoma. A cut section of congenital mesoblastic nephroma reveals an unencapsulated mass having a whorled pattern. As a result, a differential diagnosis between intrarenal teratoma and congenital mesoblastic nephroma may thus be found to be macroscopically possible. Moreover, the histologic features of congenital mesoblastic nephroma mainly consist of elongated spindle-shaped cells arranged in interweaving bundles with renal glomeruli and tubules. Classical congenital mesoblastic nephroma has an excellent prognosis in patients younger than the age of 3 months. A radical resection of the tumor is the therapy of choice, and it is usually curative.

Other differential diagnosis is cystic neuroblastomas. (11) This neoplasm is characterized by its cystic appearance with no calcification inside and just a small portion of solid tissue. (11)

An accurate histologic diagnosis is very important. Complete excision of the tumor mass is recommended and anticancer drug treatment is unnecessary.

Mature teratomas are usually benign, but they have the potential for malignant transformation. All patients with the diagnosis of benign teratoma should undergo regular long-term follow-up examinations. (12)

Conclusion

The purpose of this review was to stress on the fact that though primary renal teratomas are extremely rare, this entity must be taken into consideration in the differential diagnosis of any renal mass in childhood.



Fig 1: Intraoperative encapsulated mass in left pelvic region solid mass in left



Fig 2: Specimen after uretero nephrectomy



Fig 3: Cut section shows encapsulated variegated mass with cystic, necrotic and solid focus. A small rim of compressed renal tissue is indicated by the arrow.

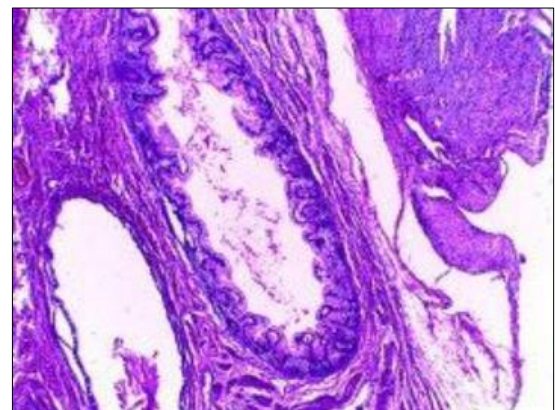


Fig 4: Teratoma components of keratinizing stratified squamous epithelium with skin adnexa. Original magnification $\times 100$ (H&E).

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