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Primary renal mature cystic teratoma in an infant: A rare case presenting with an acute intestinal obstruction

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ABSTRACT

Primary renal teratomas are uncommon and preoperative diagnosis is difficult. Additional pathologies may be more difficult to diagnosis and may increase morbidity. We are reporting a case of large primary renal cystic teratoma which was presented with acute intestinal obstruction due to congenital band at ileum. Patient was successfully managed by dividing the congenital band and excision of teratoma in toto. Pathologic examination showed evidence for a primary renal mature teratoma. Serum alpha-fetoprotein is a reliable method of assessing recurrence.

Key Words: Teratoma; kidney; acute abdomen.

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Introduction

Teratoma is a germ cell tumor which is derived from totipotential cells originating from more than one or more of the primordial germ cells [1]. Most of the teratomas are usually located near midline organs of body. The most common site to be involved is gonads (testes and ovaries) followed by extragonadal such as intracranial, cervical, mediastinal, retroperitoneal, and sacrococcygeal regions [2]. Kidney is one of the least common locations for teratomas and other germ cell tumors [3]. They are usually asymptomatic and usually grow to large mass before get noticed. Ultrasonography (USG) and computed tomography (CT-scan) are important tools in making diagnosis. Malignant transformation can occur in these teratomas although incidence is very less [4]. Surgical resection remains the mainstay of therapy for mature teratomas. We describe a case of primary renal cystic teratoma in a 5 months old female presenting to pediatric emergency with acute intestinal obstruction..

Case report

A 5 months old female infant presented to pediatric emergency with multiple episodes of bilious vomiting and abdominal distension. On examination child was severely dehydrated. Per abdomen lump was palpable in right flank

with no associated organomegaly. After initial resuscitation, x-ray abdomen (erect) was done which showed multiple air fluid levels**.** Abdominal USG showed a large cystic mass in right side (hydronephrosis?) and dilated gut loops. Left kidney was normal in echo texture and morphology. Routine investigation were as Hb was 12.3g, TLC 10,000 /mm^3, Renal function tests were as blood urea 28 mg/dl, serum electrolytes Na^+ 138mEq/L, K^+ 4.5mEq/L and serum creatinine 0.7 mg/dl. Nasogastric tube inserted to decompress abdomen which also drained bilious aspirates. Intravenous fluids and broad spectrum antibiotics were continued. As general condition of patient was rapidly worsening, so we could not shift the patient for CT scan. Exploratory laparotomy was planned with provisional diagnosis of acute intestinal obstruction with primary aim to relieve the obstruction. During laparotomy, a congenital band compressing distal ileum was found which was divided to relieve the intestinal obstruction. On further exploration we failed to locate right kidney. But there was a large cystic tumor measuring 18×15×5 cm situated retroperitoneal at place of right kidney [**Fig. 1 and 2**].

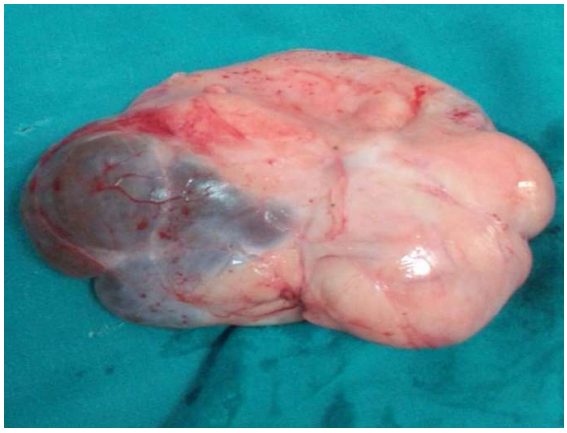

Fig. 1. Gross appearance of tumor showing reniform cystic and solid areas after excision.

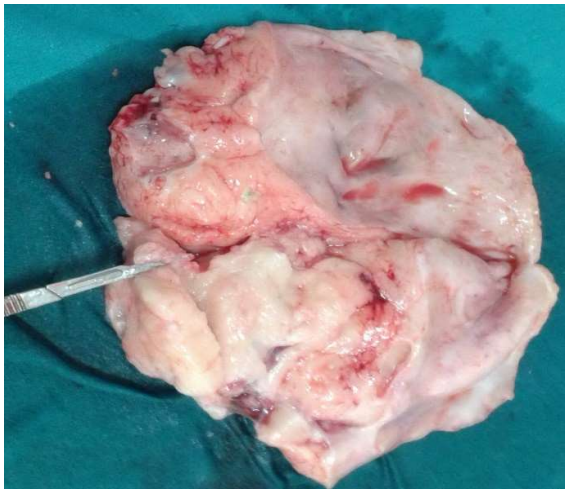

Fig. 2. Morphological details of cut section of tumor.

Feeding vessels of tumor were ligated and it was successfully excised in toto. Postoperative period was uneventful and patient was started on oral feeds on 3rd postoperative day. She was discharged on 8th postoperative day. Histopathologic examination confirmed the diagnosis of a primary renal cystic teratoma without malignant components. Microscopically cystic spaces lined by keratinizing stratified squamous epithelium with skin adnexae were identified. The solid areas showed large foci of cartilage, mucinous columnar epithelium and bone formation [**Fig. 3-6**].

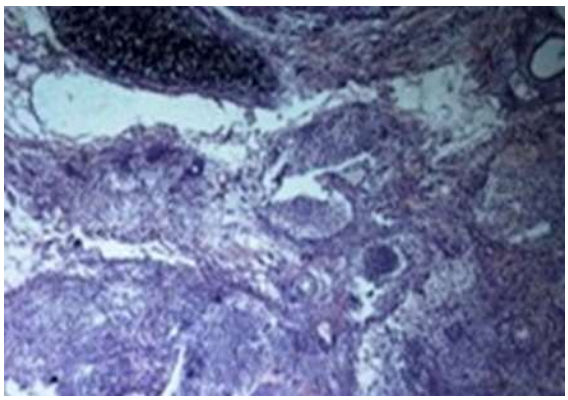

Fig. 3. Teratomatous components of bone, cartilage and columnar epithelium (H&E ×100).

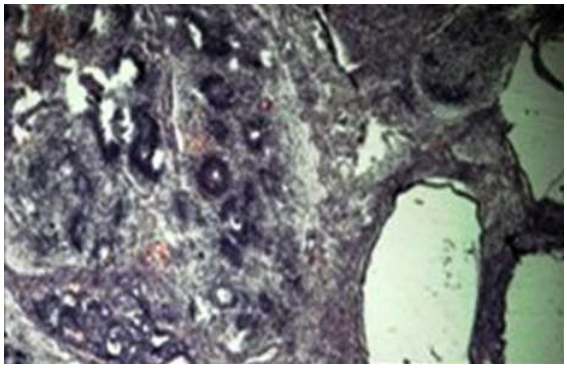

Fig. 4. Single layer cuboidal epithelium, hair follicles, shafts and sebaceous glands (H&E ×100).

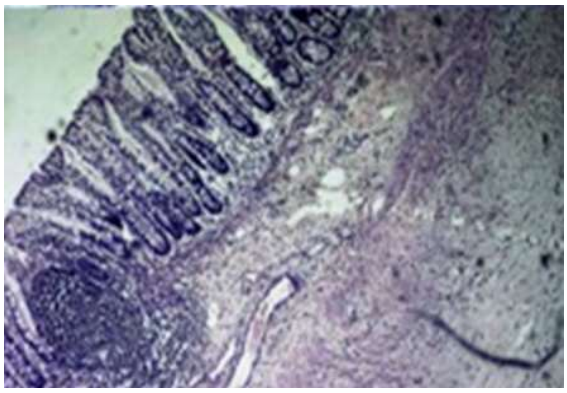

Fig. 5. Mucinous columnar intestinal epithelium with lymphoid follicle (H&E ×100).

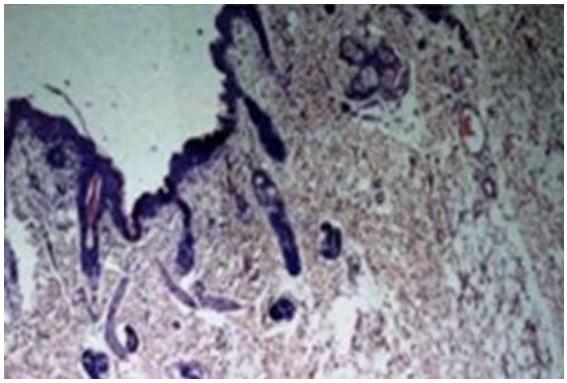

Fig. 6. Teratomatous components of keratinizing stratified squamous epithelium with skin adnexae (H&E ×100).

On follow-up USG abdomen, tumor markers alpha-fetoprotein (AFP) and beta HCG were in normal range.

Discussion

Primary renal teratoma is an extremely rare entity with less than 30 cases reported until now [5-8]. The first reported case of teratoma of kidney was in 1934, when Mc Curdy described this pathology in a seven-week-old child with Prune-Belly syndrome [9]. The patients' age ranged from 6 weeks to 71 years [5,6] and the clinical symptoms are an abdominal mass, abdominal pain, anorexia, vomiting, constipation, hematuria and pyelonephritis [10,11]. Occasionally, the tumor can be diagnosed antenatally and at birth, these patients have a higher incidence of malignancy than those in older children [12]. Additionally, retroperitoneal teratomas produce symptoms due to increase of size and compression of surrounding organs. In the current case, she presented to pediatric emergency with multiple episodes of bilious vomiting and abdominal distension. In operation, the cause of intestinal obstruction was found as congenital bands and, the association of renal teratoma and congenital bands was first presented in our case.

Diagnosis of a primary renal teratoma is often made on basis of radio-imaging techniques. Primary renal teratoma may be predominantly cystic or completely solid in appearance. X-ray findings of calcification, bone or teeth are pathognomonic for teratoma [13]. Ultrasonography is a very important investigation in making early diagnosis due to its availability and easiness to perform in the evaluation of any pediatric abdominal mass [14]. In current case, abdominal USG showed a large cystic mass in right side and dilated gut loops. But at times USG can fail to correctly identify the lesion as happened in our case. Both CT and MRI have an integral role in the characterization of primary renal teratoma and in evaluation of their extent and involvement

of adjacent structures, and therefore in treatment planning. CT scan is ideal for the assessment of retroperitoneal teratomas because it provides discrete sectional images of the organs and retroperitoneal compartments [14]. At CT scan, a mature retroperitoneal teratoma will be seen as a complex mass containing a well-circumscribed fluid component, adipose tissue, and calcification [15]. As general condition of our patient was rapidly worsening, so we could not shift the patient for CT scan. Among hematological investigations, serum alpha feto-protein level is a good indicator for diagnosis and assessing the recurrence of tumor [14]. In our case, on follow-up tumor markers alpha-fetoprotein (AFP) and beta HCG were in normal range.

Associated congenital anomalies such as horseshoe-kidney, duplicated collecting system, etc. are occasionally seen [5,6]. Differential diagnosis includes other cystic lesions such as multicystic renal dysplasia, hydronephrosis and infected renal cysts. Intrarenal teratoma must be differentiated from the rare teratoid variant of Wilms' tumor. It can contain a variety of heterologous elements with histologic findings of blastemal, stromal, and epithelial cell types [11].

Surgical resection remains the treatment of choice for mature teratomas and is also essential for definitive diagnosis [16]. Total excision of the tumor was done successfully in the present case. After excision Benign teratomas have a good prognosis, yielding a 5-year survival rate of 100% [17]. However close follow up is required to detect early recurrence and malignant transformation.

In conclusion, although primary renal teratomas are rare and preoperative diagnosis is difficult, an accurate histopathologic diagnosis is very important. Additional pathologies may be more difficult to diagnosis and may increase morbidity. Complete excision of the tumor mass is recommended. Mature teratomas are usually benign, but they have the potential for malignant transformation. Hence, all patients should undergo regular long-term follow-up study.

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