

Retroperitoneal Teratoma in Infancy: Report of an Unusual Entity

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Abstract

Primary retroperitoneal teratoma in infancy is extremely rare. In children, it accounts for 3.5-4% of all germ cell tumours and 1-11% of primary retroperitoneal neoplasms. Clinical presentation is often asymptomatic or may present as a palpable mass or abdominal distension. The imaging findings of teratoma are characteristic. Early diagnosis is important for prompt surgical resection. We present a case of huge retroperitoneal teratoma in a 5-month-old child presenting with abdominal swelling where computed tomography findings are distinctive. Biopsy led to the definitive diagnosis of mature cystic teratoma of retroperitoneum.

Keywords: Teratoma; Retroperitoneum; Germ cell tumours; Infant; Computed tomography; Fat

Introduction

Germ cell tumors are congenital tumors containing representations of all the three germ layers and are mostly seen in gonads. Extragonadal involvement includes the following sites in decreasing order of frequency: mediastinum, sacrococcygeal region, retroperitoneum, and pineal gland [1]. Primary retroperitoneal teratomas are very rare in infants and are generally seen following germ cell tumors of gonads [2]. Most of the cases are incidentally detected or present with nonspecific complaints. The main stay of definitive diagnosis and treatment of mature teratoma is surgical excision. Prognosis is generally good following complete surgical excision with an overall five-year survival rate of nearly 100% [3]. Here, we have described as case of retroperitoneal teratoma in an infant with special reference to imaging findings.

Case History

A 5 month old male child presented with progressive abdominal distension more in the left side for the past 3 months. There was no history of fever, bowel or urinary complaints or weight loss. He had no significant past surgical or medical history. He was first order child with normal birth history and no history of consanguineous marriage of the parents. No antenatal ultrasound was done. Vitals were within normal range on physical examination. The abdomen was distended. A large firm mass was palpable the left hypochondrium and lumbar region measuring about 11 × 10 cm in largest dimensions. The rest of the examination was unremarkable. The routine blood examination was found to be normal. Ultrasonography of abdomen revealed a large heterogeneous solid-cystic mass with areas of echogenicity corresponding to fatty components and calcification. Computed tomography (CT) scan of the abdomen and pelvis was performed with GE bright speed 16 slice machine to look for extent of the lesion. CT demonstrated an extremely large heterogeneous left-sided retroperitoneal mass containing areas of fat attenuation, calcification and cystic components (Figure 1). It measures about 11.5 × 9.5 × 11

cm in anterior posterior, width, and craniocaudal maximum dimensions respectively. There was displacement of aorta, inferior vena cava and bowel loops to the right side and the left kidney was displaced caudally to the left iliac fossa. There was no significant enlarged intra-abdominal or pelvic lymph node. USG-guided biopsy of the mass lesion was done, and histopathology confirmed the diagnosis (Figure 2).

Complete excision of tumor was done and at 1-year follow-up there is no sign of recurrence.

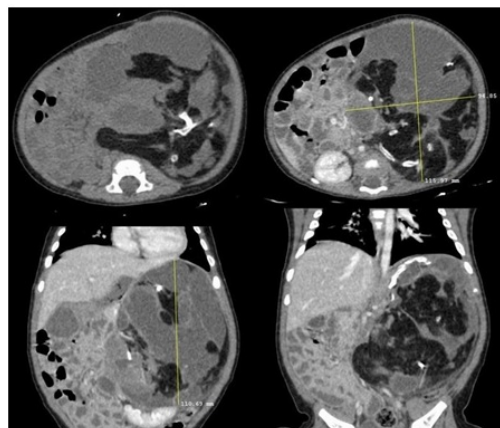


Figure 1: Computed tomography (CT) scan of the abdomen and pelvis showing a large heterogeneous left-sided retroperitoneal mass containing areas of fat attenuation, calcification and cystic components. The mass is noted to displace aorta, inferior vena cava and bowel loops to the right side and the left kidney caudally to the left iliac fossa.

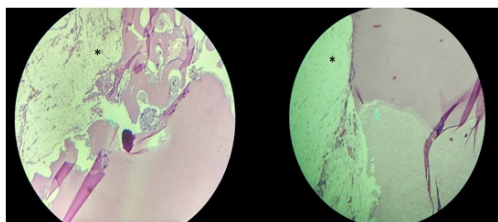


Figure 2: Photomicrograph showing mature odontoblast (arrow), present in, mature dentin, adipose tissue (*).

Discussion

Teratomas belong to the nonseminomatous group of germ cell tumors [GCT] and it is the most common form of all GCTs. They are encapsulated neoplasm consisting of varying tissues showing varying degrees of differentiation and having origin from more than one germ cell layer (ectoderm, mesoderm, and endoderm). Generally, teratomas consist pluripotent cells originating from germ cells and embryonal cells. The teratomas of germ cell origin can be congenital or acquired and are gonadal in origin i.e. from testes in male and ovaries in female. Whereas teratomas of embryonic cell origin are usually congenital and tends to be centered in extragonadal locations, most commonly in intracranial, cervical, retroperitoneal, mediastinal, and sacrococcygeal regions [1]. Retroperitoneal teratomas in most of the cases are considered to be secondary neoplasms and are mostly seen in males [4]. Primary retroperitoneal teratomas are extremely rare neoplasms accounting for approximately 1-11% of all primary retroperitoneal neoplasms and are typically seen in neonates, infants, and children age groups [5]. In adults, these neoplasms are commonly seen in the third or fourth decades of lives [6].

Further, teratomas are classified according to their origin, content, and epithelial lining and degree of maturation. Based on their origin, teratomas can be gonadal or extragonadal teratomas. Of these two, gonadal teratomas are more common and are mostly primary by nature, mainly seen in adults, and occur in gonads. In contrast, extragonadal teratomas are less common and are mostly secondary by nature, mainly seen in infants and young children [7], and generally metastasize to sacrococcygeal, mediastinal, retroperitoneal, and pineal gland sites in that order from a primary gonadal tumor [3]. Based on their composition, teratomas can be classified into solid, cystic, or mixed teratomas. Solid teratomas contain only parenchyma tissues. Cystic teratomas are consisting of only sacs of fluid, semi fluid, or fat, while mixed teratomas as the name suggests constitutes both solid and cystic components [8]. Besides, based on the epithelial lining and dermal contents of tumor, teratomas can be divided into epidermis, dermoid, and teratoid teratomas.

Epidermoid and demoid cysts differ in the content of dermal elements. Both show stratified squamous epithelium linings. Epidermoid cysts lack dermal contents whereas dermoid teratomas have contents having dermal origin like hair, sweat, and sebaceous glands. Teratoid teratomas are separate entity. They show respiratory columnar epithelium lining and contain sebum [8]. Finally based on the degree of maturation of the tumor, teratomas can be classified into mature and immature teratomas. Mature teratomas are usually benign in nature, tends to be asymptomatic and more commonly seen in females. They are further divided into solid, cystic, or mixed. They are

characterized by different well differentiated parenchymal tissues. Conversely, immature teratomas are histologically solid in nature and contain undifferentiated parenchymal tissues and can be benign or possibly malignant, or frankly malignant. They are more commonly seen in males [8,9].

Clinical presentations are usually asymptomatic and often usually asymptomatic and often are detected incidentally. In others, they may present with abdominal pain/flank pain/back pain which are nonspecific in nature, abdominal swelling, palpable mass or obstructive gastrointestinal and genitourinary symptoms [6]. Rarely they may present with abscess formation due to secondary infections [10], or acute peritonitis due to rupture [11], or malignant transformations [12]. In some cases, the tumor may be present antenatally and detected at birth. However, these neonatal teratomas possesses higher incidence of malignancy in comparison to older children [13]. Routine blood investigations are not useful in arriving at a diagnosis.

Teratomas may show a spectrum of elevated serum tumor markers such as alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and CA 19-9. Imaging findings are usually diagnostic. In about 62% of cases, plain radiographs can identify calcified elements [1]. The x-ray showing presence of bone and teeth was most helpful in establishing a diagnosis as was reported by Lack and Travis [14]. Ultrasound helps in differentiating between cystic and solid elements in the tumor [6].

The role of CT scan is mainly to determine the retro-peritoneal extension and its relationship to major vessels. Important features suggesting benign nature on CT scan includes homogeneity, cyst formation, fat density, and calcification [15]. Fat fluid level and chemical shift between fat and fluid are pathognomonic, however fat-fluid level can be seen in well differentiated liposarcomas of retroperitoneum. Liposarcoma appear as lobulated or ill defined fatty mass with soft tissue components unlike teratoma which are encapsulated and contains dense calcification [16].

Magnetic resonance imaging scans have the advantage of better resolution of soft tissues, differentiate benign and malignant neoplastic features, and most importantly aid in superior tumor staging assessment [17]. Complete excision of the tumor is preferred for histological confirmation and cure. The surgeon should be careful to dissect the tumor from renal and other major vessels, which are usually stretched out over the lesion [3]. In the cases where there is histological evidence of an immature teratoma, adjuvant therapy, such as chemotherapy, radiotherapy, or concurrent chemo radiotherapy may be given, after complete resection of the primary tumor [2]. Prognosis is excellent after complete surgical excision with an overall five-year survival rate of nearly 100% [3].

Conclusion

Teratoma should be considered as a rare but important differential diagnosis of fat-containing retroperitoneal masses. Most of them are benign in nature. Plain radiographs findings of calcifications, bone, or teeth are pathognomonic. CT is needed to evaluate the extent and character of the lesion and differentiate from other fat-containing lesions of retroperitoneum. Surgical excision of mature teratoma remains the mainstay of definitive diagnosis and treatment with an excellent five-year survival rate of nearly 100%.

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