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Case Report

Midline Primary Retroperitoneal Mature Teratoma in an Infant

Ambar Prakash[1], Ravi Ranjan[1], Shekhar Chaudhary[1], Praveen Jhanwar[2]*

[1]Junior Resident, [2]Associate Professor

Department of General Surgery, Pacific Medical College and Hospital, Udaipur, Rajasthan, India

*Corresponding author Email: jhabbuu@gmail.com

ABSTRACT

Teratomas are tumors that arise from germ cells and can be present in gonads or other regions of the body and consist of tissues of all three germ cell layers. Primary teratomas, which have not spread from any gonadal organs, in the midline retroperitoneal region are particularly rare. They account for 1-11% of all retroperitoneal neoplasms. We report a case of a Midline Primary Retroperitoneal Mature Teratoma in an 11-month-old child. The characteristic imaging findings revealed a combination of different tissues in the tumor. Successful surgery was performed to excise the tumor completely, and the child is doing well. Careful clinical examination, timely imaging, and skillful surgical management are crucial to tackling the tumor.

KEYWORDS: Midline retroperitoneal mass, Mature teratoma, Germ cell tumor, Infant, Mattox maneuver.

INTRODUCTION

Germ cell tumors can be classified as seminomatous and non-seminomatous tumors. Teratomas belong to the non-seminomatous group, but based on their origin, they can be gonadal or extra-gonadal teratomas. Extra-gonadal teratomas are less common and mostly secondary by nature, meaning they usually develop due to spread from a primary gonadal tumor[1]. Retroperitoneal tumors in childhood usually include Wilms' tumors and Neuroblastomas, and due to Teratomas being primarily gonadal, they can be missed during diagnoses. Primary midline retroperitoneal teratomas are very rare and are mostly incidentally detected. These are radiologically identified on Computed Tomography (CT) scan by the typical presence of fat, fluid, and calcific attenuation. Complete surgical excision is the best mode of treatment with an excellent prognosis[1]. Here, we are presenting a case of primary mature retroperitoneal teratoma in a female infant with special reference to its midline position.

CASE HISTORY

An 11-month-old female child presented to pediatric out-patient clinic with complaints of fever and common cold for the last five days. Clinical examination was unremarkable except for rhinorrhea and fever (99.2F). Symptomatic treatment was advised with follow-up after a week. After two days, she was again brought with complaints of excessive crying and abdominal fullness since the night before. Further examination and deep abdominal palpation revealed an immobile, non-tender, hard lump in the left lumbar region. She was referred to the general surgery department for further evaluation & management. Abdominal sonography was advised, and it showed a retroperitoneal solid cystic mass lesion with internal calcification & fluid-fat echogenicity. The differential possibility of retroperitoneal teratoma and neuroblastoma was maintained. Further evaluation with contrast enhanced CT scan of the abdomen-pelvis was planned for a detailed outline, internal content, and extension of the lesion. It

demonstrated a fairly defined heterogeneously enhancing midline retroperitoneal mass lesion with approximate dimensions of 5.2 x 6.7 x 8.8 cm, extending from bilateral posterior-medial infra-diaphragmatic surface up to L2 vertebral body level (Fig A, B & C). There were internal fat and calcific components predominantly on the left half and cystic components on the right half of the lesion. The major vessels portal vein, inferior vena cava (IVC), and abdominal aorta were displaced anterolaterally towards the right. The tumor was seen abutting major abdominal visceral organs. On the basis of age, location, and imaging findings, a diagnosis of retroperitoneal teratoma, the mature type, was made. Tumor marker revealed raised Alpha-Feto protein (AFP), and 24-hour urinary Vanillylmandelic acid (VMA) was normal.

Surgically, the child underwent a laparotomy; a superiorly based L-shaped incision was given. Performing the Mattox maneuver, the left abdominal wall flap was raised and stitched to the lower chest wall, and the left colon mobilized and rotated medially. The spleen, splenic artery, and tail of the pancreas were rotated medially. The left kidney was mobilized with Gerota's fascia. The renal vessels were identified and preserved. The gonadal vein on the left was ligated and divided. The left kidney, and the renal vein, now reflected medially to reach the aorta. The tumor was visualized, and part of the tumor was retro-aortic. The tumor was mobilized superiorly from the pancreas and medially from under the aorta and IVC in order to be removed enbloc without capsule breach and spillage (Figure D, E and F). There was no invasion of tumor into major vessels seen; no palpable regional nodes and all visceral organs in the vicinity were preserved.

Postoperative recovery was uneventful and histopathological examination of mass reported a mature teratoma (Fig G). The child was followed-up for 6 months and is doing well.

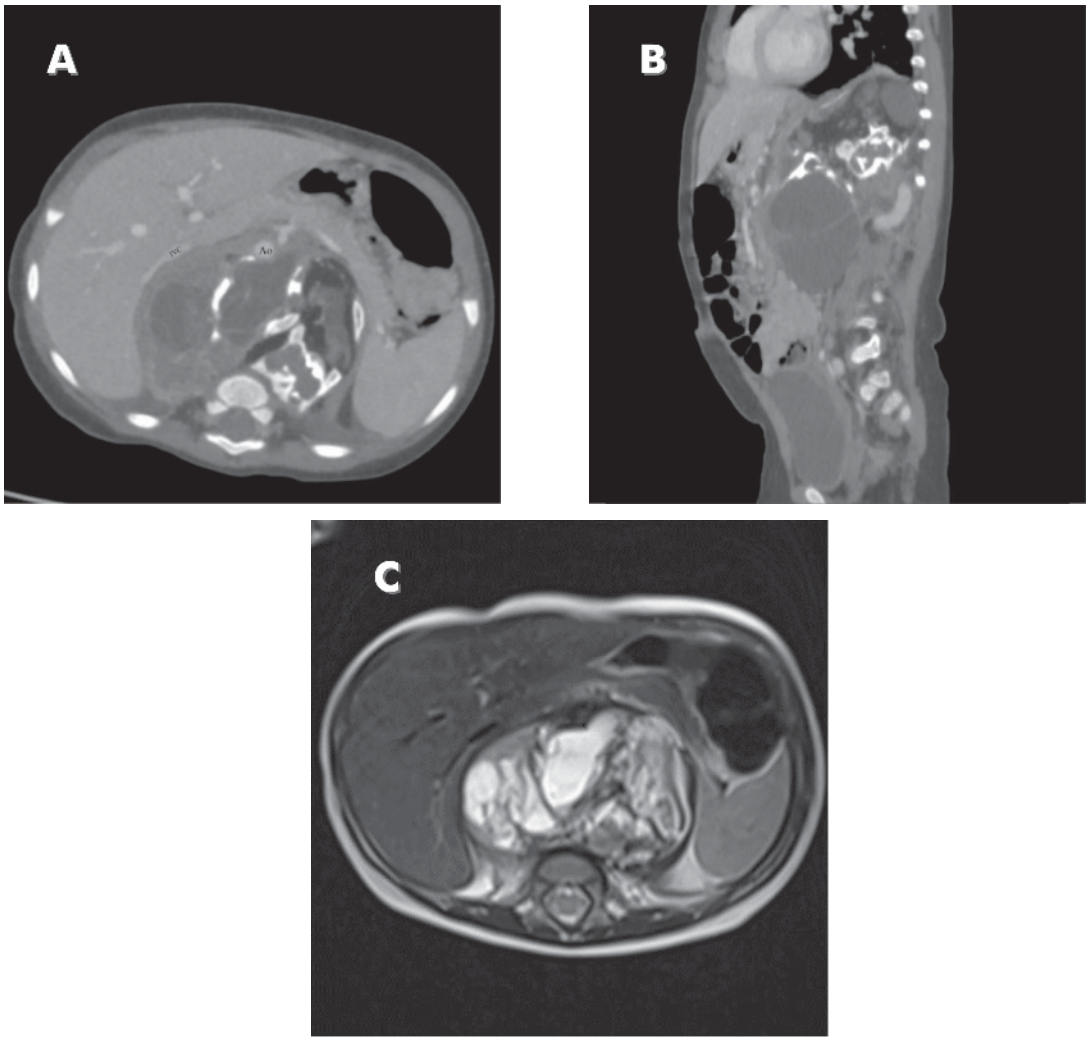


Figure (A): Axial Contrast CT scan shows fairly defined heterogeneously enhancing retroperitoneal mass. There is internal fat, fluid, and calcification. It is seen displacing aorta (Ao) and IVC anteriorly.

Figure (B): CT Sagittal section and

Figure (C): T2W MRI section showing fat, fluid, and calcification.

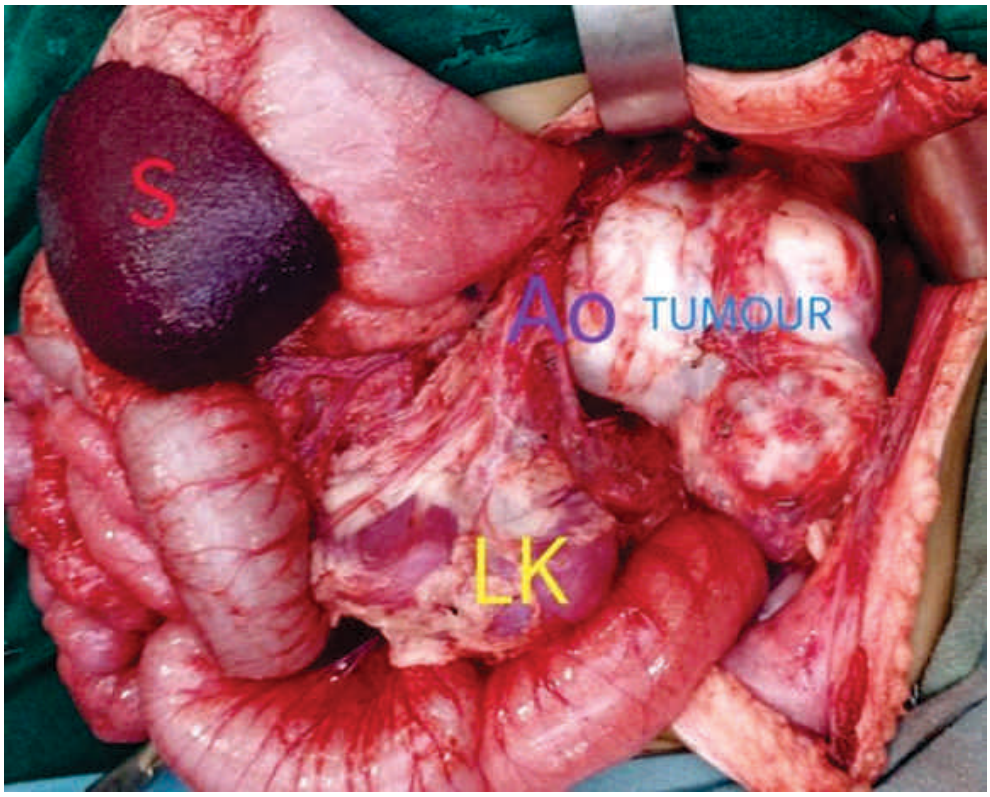


Figure (D): Intra operative–tumor location in relationship to the major vessels and visceral organ. (Ao- aorta, LK- left kidney, S- spleen)

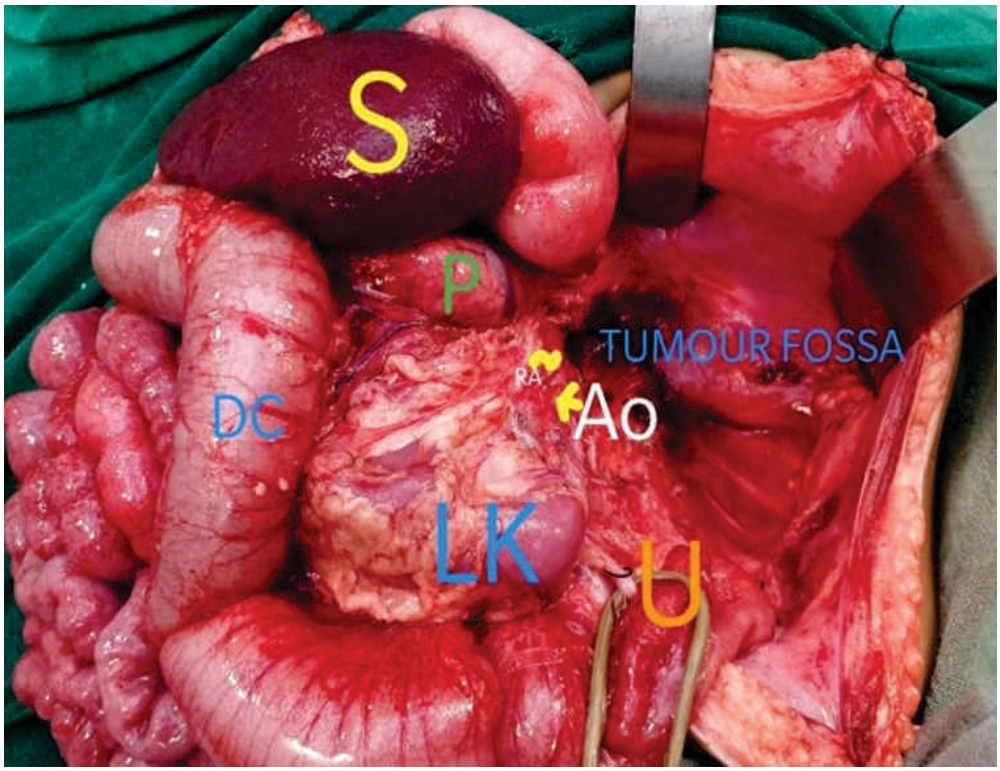


Figure (E): Intra-operative after removal of tumor mass

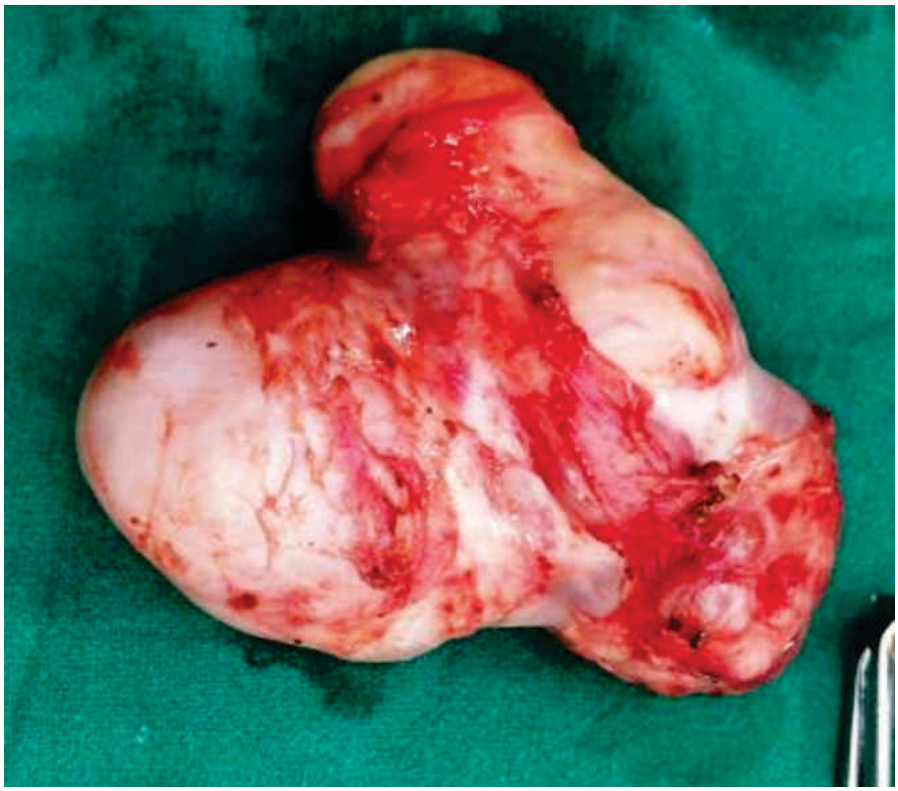
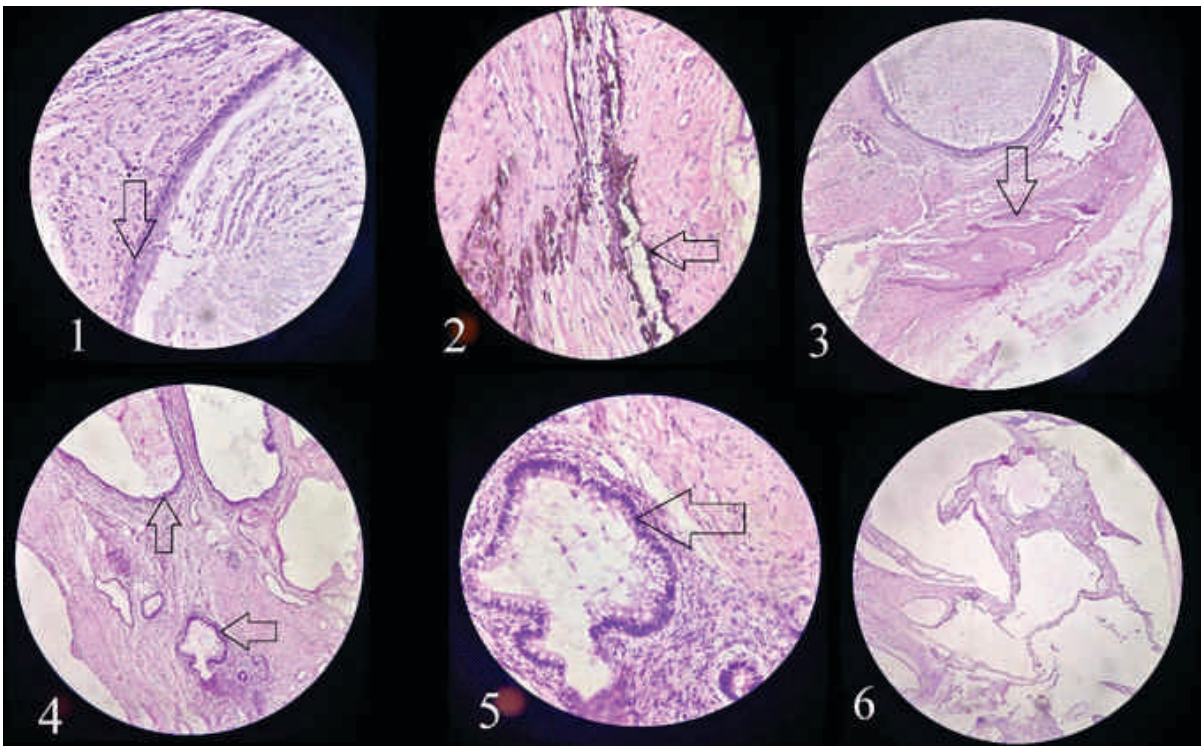


Figure (F): Gross surgical specimen after complete excision. (P- Pancreas, DC- Descending colon, U – Ureter, RA- Renal artery)



Histopathological Examination: Figure (G.): (1) - Squamous epithelial lining (Ectodermal component), (2) - Melanin pigment, (3) - Cartilage (Mesodermal component), (4 & 5) - Mucous filled columnar epithelial lining (Endodermal component) and (6) - Multiple tiny cysts (Cystic Teratoma)

DISCUSSION

Teratomas arise from abnormal development of pluripotent cells: germ cells and embryonal cells, which greatly influence the age of presentation and location. An association with modifications in chromosome 12, leading to progression and malignancy of these germ cell tumors, has been noted by Schmoll, although there is still more clarity needed regarding the specific mechanism of progression[4]. Teratomas of germ cell origin can be congenital or acquired and are usually gonadal. In contrast, teratomas of embryonic cell sources, which are always congenital, are usually found in extragonadal (15%) locations including the mediastinum, sacrococcygeal region, retroperitoneum, and pineal gland[2]. Very rarely they may be found in the lumbar region[3]. Retroperitoneal teratomas comprise only 3.5 – 4% of all germ cell tumors in children. Primary retroperitoneal teratomas are extremely unusual neoplasms accounting for approximately 1–11% of all primary retroperitoneal neoplasms and typically occur in neonates, infants, and children[4].

Teratomas are also classified according to contents, epithelial lining, and degree of maturation. Based on their composition, teratomas can be classified into solid, cystic, or mixed[5]. Solid teratomas contain only parenchymal tissues. Cystic teratomas consist of only sacs of fluid, semi-fluid, or fat, while mixed teratomas, as the name suggests, constitute both solid and cystic components, as in our case. In the indexed case, a cystic component was found more on the right side and a solid component on the left side.

Clinical presentations are either asymptomatic or may present with abdominal/flank or back pain which is nonspecific in nature, abdominal swelling and a palpable mass, or obstructive gastrointestinal and genitourinary symptoms depending upon the location and size of the mass. Literature does not reveal a side or gender predilection, and almost an equal incidence in males and females has been reported[6]. In the indexed case also, the baby presented with nonspecific complaints, and then a suspicion of retroperitoneal mass was made during the clinical examination.

X-ray findings of calcification/ bone/ teeth are pathognomonic[7]. Other various imaging modalities, including ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI), are used for the diagnosis as well as to delineate the extension of the tumor mass and its relation to vital organs and major vessels[8]. Teratomas may show a spectrum of elevated serum tumor markers such as alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and CA 19-9[9]. Diagnosis of these tumors is based on a combination of a high index of clinical suspicion and laboratory and radiological investigations, though histopathology is the gold standard.

Complete excision of the teratoma offers the best chance of cure. Tapper and Lack reported that complete removal in the initial surgery was the most important prognostic factor[10]. For mature teratomas, only complete surgical removal will suffice. However, complete resection plus adjuvant therapy, such as chemotherapy, radiotherapy, or concurrent chemo-radiotherapy, may be needed in the case of immature teratomas. Nevertheless, during surgery, the surgeon should be careful to dissect the tumor from renal and other major vessels, which are usually stretched out over the lesion. After complete removal, the prognosis of a mature teratoma is generally good, with a five-year survival rate of nearly 100%.

CONCLUSION

A primary retroperitoneal teratoma is a rare entity but always consider other differential diagnoses of retroperitoneal masses in children. The success of treatment lies in a high index of suspicion, prompt diagnosis, and complete surgical excision. The surgical incision should be planned preoperatively by looking at the tumor location and extent on imaging. The retroperitoneal teratoma, situated in the midline, required not only medial visceral rotation but also lifting major vessels to avoid catastrophic hemorrhage.

CONFLICTS OF INTEREST: None

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