Case Report

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A Neonate with Retroperitoneal Mature Cystic Teratoma – A Case Report

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Abstract

Retroperitoneal teratoma is a rare occurrence in neonates. They are usually asymptomatic and the only finding may be abdominal distension. We report an unusual case of a large retroperitoneal teratoma in a neonate who presented with abdominal distension. Imaging done showed a cystic lesion with calcifications, internal septations and fat dense areas. It was successfully managed by surgical excision, findings of which were consistent with that of a mature cystic teratoma. Despite being rare, we must consider retroperitoneal teratomas as a differential diagnosis of an abdominal mass in a newborn. Early detection and complete surgical excision may be life-saving.

Introduction

Teratomas are the most common germ cell tumours. Retroperitoneal teratomas are uncommon neoplasm accounting for 3.5 to 4 % of all germ cell tumours in children.[1] Most of these tumours are benign, but malignancy may be encountered especially in neonates compared to other paediatric age groups.[2] They usually present in the pararenal area and more common on the left side, often presenting as abdominal distension or a palpable mass.[1] Most cases are detected incidentally. Once detected, early management may help in preventing morbidity and mortality. The definitive treatment is surgical removal and prognosis is generally good following complete surgical excision.[2] Here, we describe an interesting case of retroperitoneal teratoma in a neonate.

Case Report

A 28 days old male neonate born by full-term normal vaginal delivery to a non-consanguineously married couple with a birth weight of 3.6 kg, was brought with the complaints of abdominal distension since 14 days of life. Mother had an uneventful pregnancy and antenatal ultrasonography reports were not available. On general physical examination, the child had mild pallor with no icterus, cyanosis or lymphadenopathy and weighed 4.6 kg. Vitals were within normal limits. Abdominal examination revealed a single non-mobile, non-pulsatile, non-compressible mass measuring approximately 15 cm x 15 cm with firm consistency with a dull note on percussion. The mass extended superiorly from left hypochondrium to left iliac fossa craniocaudal and from left lateral wall of the abdomen, crossing the midline to the right midclavicular line horizontally. There was no hepatosplenomegaly and bowel sound was normal. Bilateral testes were palpable. Other systemic examination was unremarkable. Initial haematological investigations revealed haemoglobin of 9.3 g / dL (10.7-17.1 g / dL) and a white blood count of 23800 mm^3 (4000 -19500 mm^3). Serum beta-HCG was 0.107 mIU / mL (Normal < 50 mIU / mL), and serum alpha-fetoprotein was 1184 IU / ml (Normal 260-5215 IU / mL). Ultrasonography of

abdomen was done at other centre, however its report was not available.

CECT of the abdomen revealed a large well defined peripherally enhancing cystic lesion with few irregular dense calcifications, enhancing thick internal septations and fat dense area in left lumbar, hypogastrium and umbilical regions. The mass was retroperitoneal measuring 8.2 cm x 9.9 cm x 10.2 cm extending anteriorly up to the abdominal wall, posteriorly compressing left kidney, pancreas, abutting abdominal aorta, inferior vena cava, and bilateral renal arteries (Figure 1). It was compressing small bowel loops with fat planes maintained on all sides.

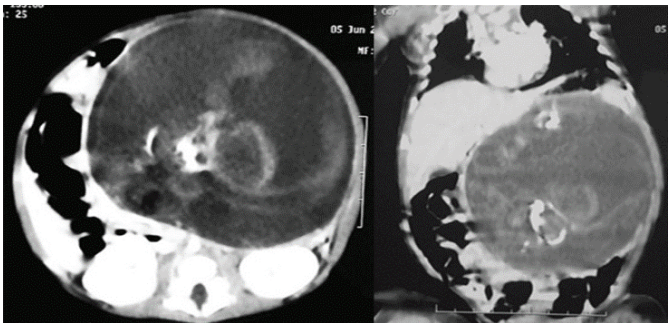
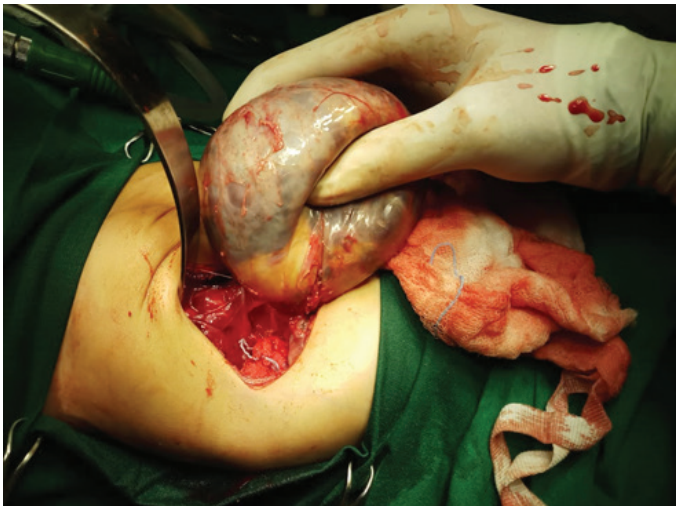


Figure 1. CECT Abdomen showing a large, well defined peripherally enhancing cystic lesion with few irregularly dense calcification, thick internal septations and fat dense areas.

The baby was taken up for surgery. Exploratory laparotomy was performed with a left upper transverse incision. Intraoperatively, the tumour was superiorly related to the stomach and left lobe of the liver on the right side and spleen and dome of the diaphragm on the left side, inferiorly extending till the pelvic brim and mesentery was pushed to the right side of the abdomen (Figure 2). There was no encasing of the vessels or evidence of infiltration. Other intra-abdominal organs were normal.



Tumour measuring approximately 15 cm x 15 cm x 15 cm was excised completely and sent for histopathological examination. The tumour weighed 350 grams. On gross examination, the outer surface of the cyst was smooth with few congested areas (Figure 3A). The inner surface showed an attachment of a firm mass covered with hair, soft cystic mass and a pale brown mass. Cut section of the hair covered mass showed mucoid haemorrhagic areas along with hard bony areas while similar section of the soft mass showed light yellow mucoid areas. Cut section of the pale brown mass showed fatty areas and was gritty (Figure 3B). Histopathological examination of the cystic lesion revealed various proportions of squamous epithelium, mucous glands, cartilage, bone, glial tissue, hematopoietic elements, adipocytes, adnexal structures, melanin pigment, and tooth thereby confirming the diagnosis.

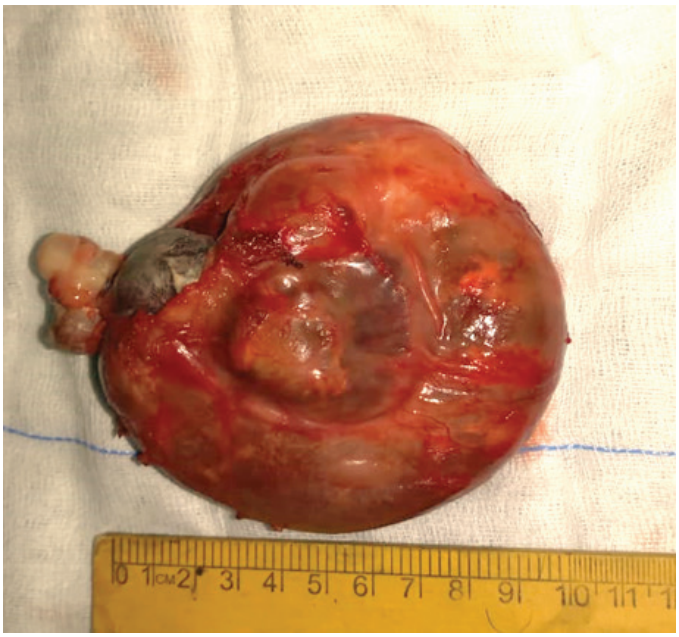


Figure 3 A. Gross image of the resected tumour showing smooth surface with few congested areas

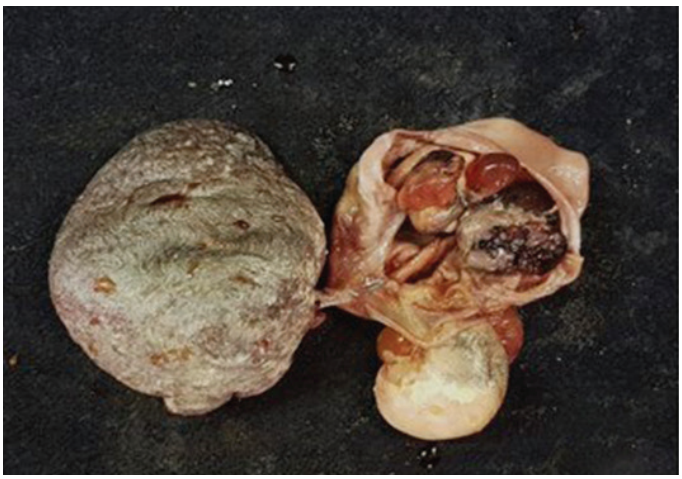


Figure 3 B. Cut-section of the tumour

Discussion

Germ cell tumours are neoplasm that contain derivatives of all three germ cell layers.[3] They are usually gonadal in origin. Extra- gonadal germ cell tumours are most commonly found in the sacrococcygeal region with retroperitoneal location being a rarity.[3] The patients with retroperitoneal teratomas have a female preponderance, but conversely, our patient was a male neonate with a teratoma which was located at the retroperitoneum.[4] They are usually secondary neoplasms and primary retroperitoneal neoplasms are extremely rare.[1] The anatomic variety seen in these tumours and their occurrence in midline and gonads is explained by the germ cells migratory capacity.[5] They may be solid, cystic or mixed.

Our patient presented with abdominal distension and was found to have a left- sided mass with no complications. Occasionally the tumour may present late, with infection and rupture leading to complications like abscess formation and peritonitis which may be life-threatening.[1] Therefore, early diagnosis becomes imperative and may be life-saving. Imaging plays a vital role in the diagnosis and management of teratomas.[6] Prenatal ultrasound may also provide valuable information for early diagnosis.[7] In our case, prenatal ultrasound was performed, but the final reports were not available. CECT of abdomen aids in determining the location, morphology, extent, relation to great vessels and other adjacent structures, and the benign nature of the tumour as suggested by homogeneity and fat density.[8] Following clinical examination and imaging, differential diagnoses which we considered were namely germ cell tumour, neuroblastoma and nephroblastoma. Our neonate did not have cutaneous manifestations, opsoclonus-myoclonus, periorbital ecchymosis or any hepatic involvement, which reduced the possibility of neuroblastoma. The tumour did not arise from the kidneys; hence, nephroblastoma was ruled out. Due to the presence of well- defined cystic lesion with calcification and fat densities, we considered the possibility of germ cell tumour.

Surgical excision is the mainstay treatment for teratomas and complete excision is adequate for benign mature teratomas.[9] On the other hand, malignant teratomas may require adjuvant therapy following surgical excision. Surgery may become challenging for large tumours related to great vessels with a risk of injury during dissection. Yang et al reported retroperitoneal teratoma resection to have a high perioperative complication rate.[9] Our neonate underwent complete excision of the tumour with no intraoperative or postoperative complications. Prognosis is excellent after complete surgical excision of mature cystic teratoma. Recurrence depends on the histological variety of the teratoma and is seldom seen in mature teratomas. However, some cases of recurrence have been reported following surgical excision.[4] Hence a long-term follow-up is recommended to look for any recurrence and initiate early intervention. Alpha-fetoprotein (AFP) is a tumour marker that may be elevated, and serum AFP levels measurement during the follow-up period can help detect recurrence. Raised serum AFP levels, especially in cases with immature teratomas have a higher risk of malignancy.[3] Our patient had no complications or recurrence at one-year follow-up.

Conclusions

Retroperitoneal teratoma is a rare entity in neonates. Early diagnosis and management can give a favourable outcome. They have distinct imaging findings like teeth, hair, calcifications; which are pathognomonic. Majority of them are benign. The lesions can be treated most effectively via surgical excision, and it is always preferable to excise the tumour as early as possible during the neonatal period. The prognosis is usually favourable and recurrence can be followed with tumours markers such as AFP.

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