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Case Report

Malignant retroperitoneal teratoma in a young girl: a rare case report

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ABSTRACT

Of all primary retroperitoneal teratomas, less than four percent occur in children and 90% are benign. Here we report a case of malignant retroperitoneal teratoma (dermoid) in a 15 year old girl who presented to our hospital - Acharya Vinoba Bhave Rural Hospital (AVBRH). She presented with a tender, large, irregular mass with variegated consistency in the entire left side of abdomen crossing midline. Ultrasound of abdomen suggested a complex intra-abdominal mass with septations and lobulations. It was not feasible to use other imaging modalities for evaluation due to poor socio-economic status and illiteracy. Patient underwent exploratory laparotomy with tumor resection along with left kidney and part of the descending colon which was densely adhered to tumor. Histopathological examination of tumor was suggestive of immature teratoma. Post operative recovery was uneventful and patient was discharged from the institution. Tissue adherence which can be observed in both benign and malignant form of teratomas, requires extended surgery for removal of adhered organ for the completeness of surgery and good prognosis.

Keywords: Teratoma, Retroperitoneal, Malignant

INTRODUCTION

Commonly germ cell tumors arise in the gonads.[1-3] Teratomas belong to a class of tumors known as nonseminomatous germ cell tumor (N.S.G.C.T.).[4] Retro-peritoneum is rare site of dermoid tumors (3.5 to 4% of all germ cell tumors in children)[5] in which 90% of these tumors are benign. Here we are reporting case of malignant retroperitoneal dermoid who underwent successful surgical excision.

CASE REPORT

A 15 year old girl presented with a slowly growing lump in the left side of her abdomen childhood. She had dull aching intermittent pain since one month. There were no associated symptoms in spite of the size of the lump.

Clinical examination revealed a large mass of variegated (soft and firm) consistency occupying the entire left side of the abdomen with tenderness and visible peristalsis. The patient was evaluated with Ultrasound (US) which showed a complex mass in the abdomen with solid and cystic areas with septations and lobulations. X-ray and other biochemical investigations were within normal range. It was not possible to evaluate the patient with more advanced imaging studies due to poor socioeconomic status and illiteracy. Ultrasound guided Fine Needle Aspiration Cytology (FNAC) was done which suggested a benign cystic teratoma.

Exploratory laparotomy revealed a large retroperitoneal mass of size 32x15x15cms with solid and cystic components. Tumor was located posterior to descending colon and mesocolon. Stomach and pancreas were pushed upwards and anteriorly where as left kidney was grossly

displaced to the iliac fossa with stretching of the renal vein and artery. Mesocolon, renal vein and kidney were firmly adherent to the solid part of the tumor. Tumor was freed from the retroperitoneum, colon and spleen. Superiorly it was separated from body of pancreas and aorta and inferior vena cava medially. It was not possible to separate the kidney and mesocolon from the tumor so an extended approach for removal of teratoma with kidney and part of descending colon was done as intra operative frozen section biopsy was in favour of malignant component.

On gross examination cystic portion of tumor was filled with the sebum and greasy material along with the hair within it. There was no bone or cartilage formation in the solid part of the tumor. Histological examination there was immature neuroepithelial tissue with areas of calcification, hemorrhage and necrosis. Formation of few glandular elements with secretins and immature cartilage was seen. Pseudostraitified columnar epithelium with abundant mucinous stroma with suspicious acini-insular pattern was seen suggestive of immature teratoma grade-III.

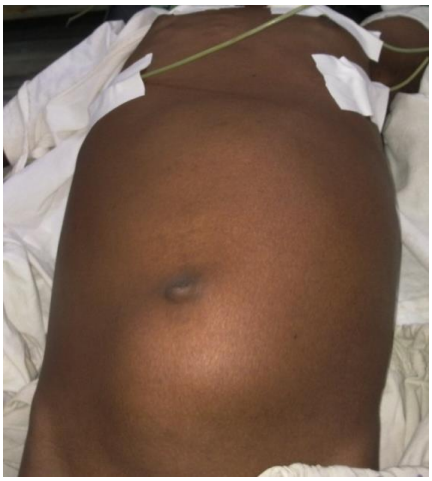

Figure 1: Pre - operative photograph of patient.

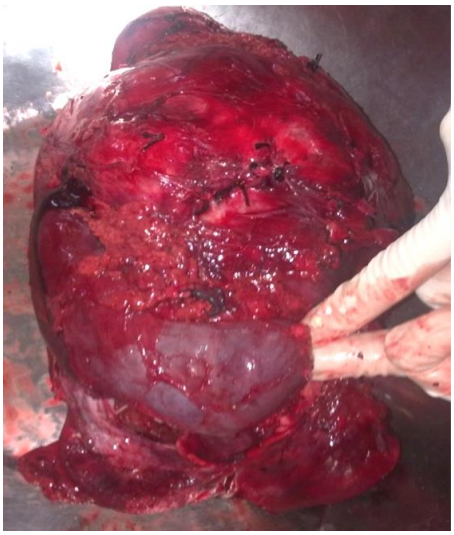

Figure 2: Excised retroperitoneal dermoid with excised left kidney and part of descending colon.

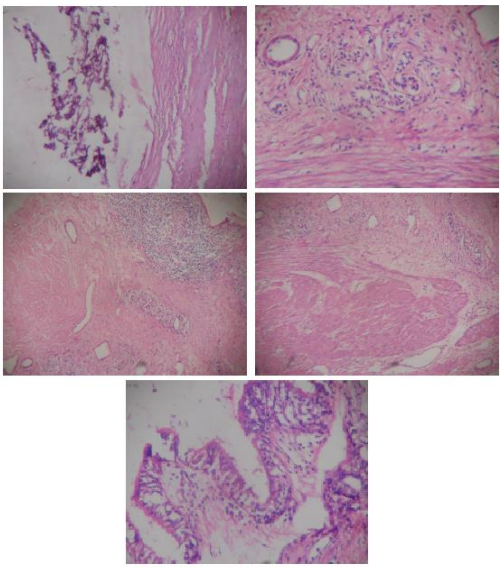

Figure 3: Histopathology slides of excised tumour.

DISCUSSION

Teratomas are the tumors that arise from the pleuripotent embryonic cells[7] of the germinal layers and will have elements of Ectoderm, Mesoderm and Endoderm.[8] Teratomas can occur anywhere in the body or in any organs but mostly found in the paraxial and midline locations.[8,9]

Retroperitoneal teratomas are the least and gonadal teratomas are highest in their occurrence; other sites being sacrococcigeal, near pineal body, near thyroid and anterior mediastinum.[8] Only 3.5 to 4% of all teratomas occur in the children. Among retroperitoneal teratomas, over one-half occur in the pediatric age group, and about 90% are benign.[7] Malignancy is uncommon in retroperitoneal teratomas except tumor which presents antenatally and diagnosed at birth. The chance of malignant transformation in extra gonadal teratomas occurring in infancy and childhood is 10% whereas 26% in the adults.[8,10-12]

Patients with teratomas are usually asymptomatic due to enough retroperitoneal space[13] and may become symptomatic due compression on surrounding structures[8] like colon, kidney, pancreas stomach or the vessels. Incidence of retroperitoneal teratomas is more on left and twice in female than male.[8]

In about 53 to 62% of teratomas, calcification is seen at the rim of the tumor.[7,14] Calcification, bone formation, and teeth on US or X-ray are manifestations of benign teratoma.[7,14] However calcification cannot be considered indication of benign tumor since 12.5% of calcified tumors are malignant.[7,14]

Patient can be evaluated by X-ray of the abdomen and contrast CT.[5,15] Some authors recommend angiography, inferior venacavography and needle biopsy for accurate

diagnosis. Alfa-feto protein (AFP) can be used as biomarker for diagnosis as well as for recurrence. Main aim of evaluation of the benign teratoma is to diagnose malignant change which is reported in about 0.25 to 0.8% cases.[5] Prognosis is good if the cyst wall is not penetrated.

Macroscopically teratomas are of two types: - A) Cystic usually benign and B) Solid teratomas are generally malignant and formed of various tissues like fibrous, fatty, bones and cartilage and also may have immature embryonic tissue.[14] Complete surgical excision of the tumor tissue is most important factor for cure of the patient.[14-17]

Tissue adherence which can be observed in both benign and malignant form of teratomas, requires extended surgery for removal of adhered organ for the completeness of surgery and good prognosis.[1]

Differential Diagnosis of Retroperitoneal Tumours: Sarcomas comprise a third of retroperitoneal tumours. Other retroperitoneal neoplasms include lymphomas and epithelial tumours or might represent metastatic disease from known or unknown primary sites. The most common benign pathologies encountered in the retroperitoneum include benign neurogenic tumours, paragangliomas, fibromatosis, renal angiomyolipomas and benign retroperitoneal lipomas.[18]

CONCLUSION

Retroperitoneal teratomas are rare and 90% of the tumors are benign.[7] Calcification, bone formation and teeth in retroperitoneal region are pathognomonic of the condition.[7,14] Possibility of malignancy can not be ruled out even in infancy. Complete surgical removal is essential for good outcome.[1] Recurrence can be monitored with tumor markers like serum alpha-fetoprotein.[5]

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