Original Article

Primary retroperitoneal teratomas in children: A single institution experience

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ABSTRACT

Objective: This study aims to highlight the clinical features, investigations and treatment outcome of retroperitoneal teratomas (RPT) in children. **Materials and Methods:** A total of eight patients (six males and two females, age range between 6 months–10 years) of RPT admitted in the department of Paediatric Surgery, PGIMS, Rohtak, between 1996–2008, were studied. The patients were investigated with hematology, x-ray, ultrasound, and computerised tomography (CT) of abdomen and serum alpha-fetoprotein levels in pre and postoperative period. All patients underwent complete surgical resection. In one patient, the tumour had malignant component (yolk sac) and was given postoperative chemotherapy. Postoperative follow-up included serum alpha-fetoprotein in addition to clinical examination and radiological assessment to detect recurrences. **Results:** The tumours were located on both sides in almost equal proportion (four on right, three on left, and one bilateral]. All tumours could be excised completely preserving the kidneys in all patients. But in one patient injury to inferior vena cava (IVC) occurred which was repaired successfully. Majority (7 out of 8) were histological benign, and in one yolk sac tumour was malignant component which needed chemotherapy. All children were on follow-up and one patient with malignancy lost to follow-up after three cycles of chemotherapy. In rest there was no tumour recurrence. **Conclusion:** RPT are rare paediatric neoplasms. As majority are benign, a complete excision preserving the kidneys, is usually curative. Serum alpha-fetoprotein is a reliable method of assessing recurrence. Malignancy in the tumour may warrant further chemotherapy.

Key words: Extragonadal germ cell tumours, retroperitonium, teratomas

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INTRODUCTION

Retroperitoneal teratomas (RPT) are the third most common primary retroperitoneal tumour in the paediatric population after neuroblastroma and Wilms tumour.[1] They occur outside the pelvis and represent only 5% of all childhood teratomas.[2] Due to its enormous size and close approximation with the intraabdominal structures, these tumours pose a challenge to the surgeon, at the same time giving a good result after complete excision. We report our experience with RPT at a single institution during a 12-year experience.

MATERIALS AND METHODS

A retrospective review was carried out on the records of the patients diagnosed with retroperitoneal teratoma at Pt. B. D. Sharma PGIMS, Rohtak, between January 1996–January 2008. The clinical history, examination findings, investigations, and treatment details of the patients were obtained from hospital records. The records of these patients with paediatric RPT were reviewed for age of presentation, sex, mode of presentation, site of teratoma, method of diagnosis, treatment, and outcome. Preoperative evaluation included plain x-ray, abdominal ultrasonography, and contrast enhanced CT-scan. Haematological investigations and serum alpha-fetoprotein assay (AFP) were done to obtain preoperative values and the surgery was done through a transperitioneal approach in all cases. The excised specimen were analysed by histological examination to detect malignant elements. The patients were reviewed in follow-up after three weeks, when serum AFP was estimated and it returned to normal levels by this time in all, except in the patient with malignancy. Thereafter, they were followed up in outdoor with monthly estimation of serum AFP levels (for one year), to detect the recurrence and then three monthly till last follow-up.

RESULTS

Over a period of 12 years, eight patients of paediatric

RPT were treated at our hospital. The ages of these patients ranged between 6 months to 10 years, with a male preponderance [Table 1]. The mode of presentation was abdominal distension with palpable abdominal mass. In spite of the large intraabdominal tumour, all the patients had a very good general condition. Four patients had elevated preoperative serum alpha-fetoprotein. X-ray of the abdomen showed ground glass appearance with calcification [Figure 1]. Ultrasound of abdomen showed a large, complex, densely echogenic mass, with no ascites. CT scan picture showed solid and cystic mass containing fluid component, adipose tissue, and calcification [Figures 2 and 3].

All tumours were approached through a supraumbilcal transverse transperitoneal method. There was no definite preponderance to any side (three on left, four on right, and one midline). All the cases were in close relation to the kidney with stretched renal vessels over the surface of tumour. In one patient, the tumour displaced IVC and it got injured but was repaired successfully. Peroperatively, on gross examination, the tumour contained either teeth, hair, bone, and brown/pale fluid or fetus *in fetu* [Figures 4 and 5]. Postoperative period was uneventful. Histologically, all the tumours except one proved to be benign mature teratomas. In one patient, the histopathological examination of specimens showed yolk sac tumour and the patient was given three courses of BEP regime (bleomycin, etoposide, cisplatin). However, this patient was lost to follow-up afterwards. Rest all seven patients were followed upto a maximum of six years and follow-up investigations included AFP levels monthly upto one year. These levels returned to normal in all the benign tumours.

DISCUSSION

RPT comprises 3.5–4% of all germ cell tumours in children and 1–11% of primary retroperitoneal neoplasms.[2,3] Patients present with abdominal distension or a palpable mass. Occasionally, the tumour is present antenatally and diagnosed at birth, these neonatal teratomas have a higher incidence of malignancy than those in older children.[4] In the present study, the age of patients ranged from 6 months to 10 years (mean age, 5–25 years). Majority of these tumours are benign and pararenal in location with no specific side or gender predilection as reported in the literature.[2] Also, in the present study, there is no specific side predilection (three on left, four on right, and one bilateral); but there is definite male preponderance (6/8) which is in contrast to another study where there is female preponderance.[5] All the patients except one in the present study had a mature benign teratoma, the malignant tumour being a primary yolk sac tumour.

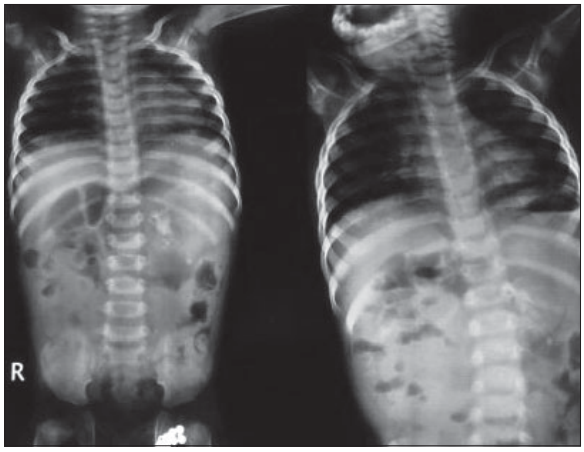


Figure 1: Abdominal radiograph of retroperitoneal teratoma showing calcification

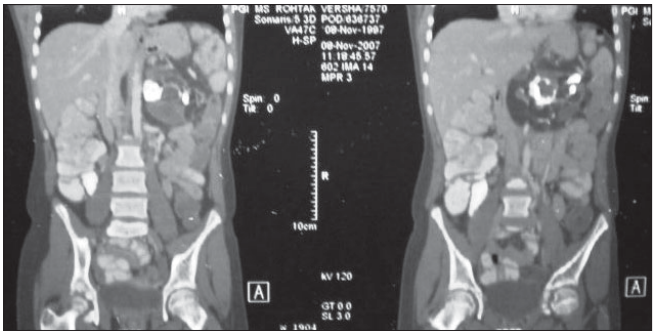


Figure 2: CT image showing retroperitoneal teratoma on left side with calcification

Table 1: Patients' characteristics and treatment outcome of primary retroperitoneal teratomas in children

Age	Sex	Tumour Site	Tumour size (cm)	Histo-pathology	Treatment	Outcome
6 months	M	Right pararenal	12 x 13	Benign	Resection	Cured
1.5 years	M	Left pararenal	14 x 18	Benign	Resection	Cured
10 years	F	Midline bilateral	18 x 15	Benign	Resection	Cured
1 year	M	Right pararenal	12 x 18	Benign	Resection	Cured
5.5 years	F	Right pararenal	25 x 25	Benign	Resection	Cured
8 months	M	Left pararenal	15 x 10	Benign	Resection	Cured
1 year	M	Left pararenal	15 x 19	Benign	Resection	Cured
2.5 years	M	Right pararenal	13 x 18	Malignant	Resection	Lost to follow-up

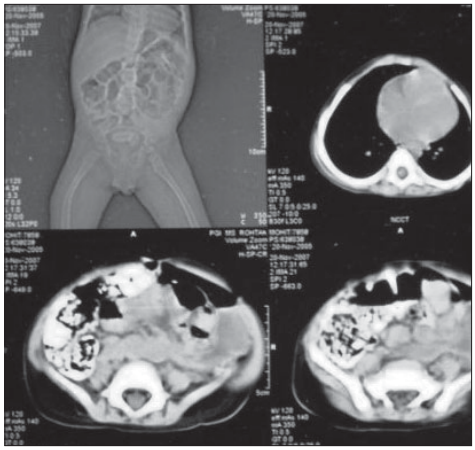


Figure 3: CT abdomen and pelvis showing retroperitoneal teratoma on the right side

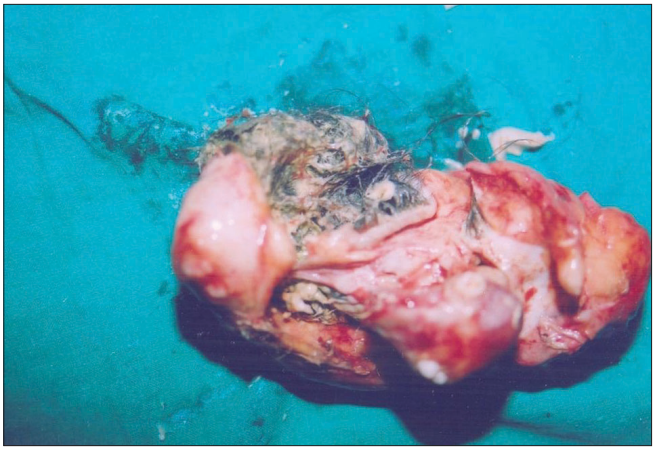


Figure 4: Postoperative gross specimen of retroperitoneal teratoma showing hair

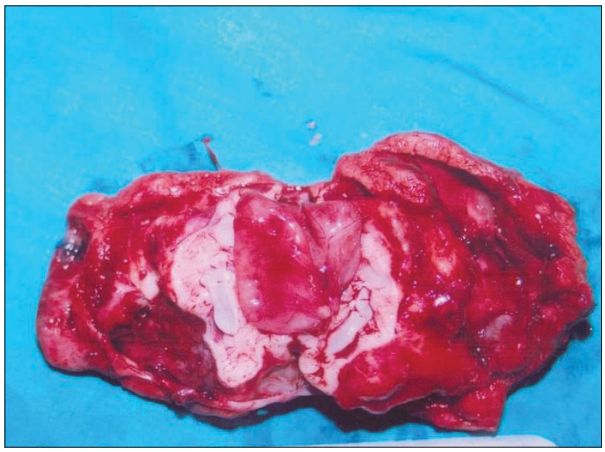


Figure 5: Postoperative gross specimen showing fetus *in fetu*

Ultrasound of the abdomen is usually the first imaging modality employed in the evaluation of any paediatric abdominal mass. However, in RPT, x-ray may demonstrate calcification or formed bony components such as teeth and phalanges (which are pathogenomic). Schey and Vesley have recommended only a plain abdominal X-ray and excision of tumour if the characteristic calcification is demonstrated.[6] Lack and Travis have also reported that the presence of bones or teeth on an x-ray was the most helpful in establishing a diagnosis.[7] However, from experience of this study, the authors argue that preoperative CT-scan is useful to delineate the extent of the disease in retroperitoneum and its relationship to major vessels. However, CT-scan can overestimate the degree of tumour adherence to adjacent structures than actually seen on exploration.[5] Therefore, CT-scan findings should not prevent surgical exploration of the tumour and even bilateral lesions are amenable to complete removal. Hayasaka and Yamada have reported internal homogeneity, fat density, cyst formation, and calcification to be important predictors of a benign retroperitoneal tumour on CT.[8] Some authors have even advocated angiography, inferior venacavography, and needle biopsy for the accurate diagnosis of these tumours, which were not done in the present study.[9] Among haematological investigations, serum alpha feto-protein level is a good indicator for diagnosis and assessing the recurrence of tumour. Serum AFP was elevated peroperatively in four (4/8) and it returned to normal in three (3/4) and formed a useful marker of monitoring recurrence.

Complete excision of the tumour offers the best chance of cure.[5] Malignancy is uncommon in RPT hence nonmutilating excision is possible and should be attempted even in lesions involving both sides of abdomen.[10] The most important aspect of the excision is to dissect the tumour from renal and other major vessels, which are invariable stretched out over the lesion. If inadvertently any major renal is injured, as IVC in one if the case in present case, can be repaired. Removal of the kidney was not necessitated in any of the patients in this study.

Prognosis is generally good and curative if the tumour is completely removed. One patient in present study showed yolk sac tumour on histopathology. This patient was given chemotherapy with bleomycin, etoposide and cisplatin (BEP regime). However the patient lost to follow-up after three cycles of chemotherapy. Rest all patients were doing well till last follow-up.

Unresectable lesions, immature teratomas and endodermal sinus tumours have a comparatively worse prognosis, and may warrant aggressive chemotherapy due to their metastatic nature.[11] Lastly the single most important factor in prognosis in RPT is complete removal which must be tried in every case of RPT irrespective if its size.

CONCLUSION

RPT are uncommon paediatric tumours with horrific intraoperative appearances. These tumours behave opposite to their appearances, with a generally benign nature and curative to complete surgical excision. Even though the anatomy of the region might be severely affected, a surgical cure is possible in the hands of the experienced surgeon. Modern imaging modalities have proven their value in delineating the extent of the disease as well as predicting the surgical prognosis. Malignancy in these tumours, even through rare; may warrant chemotherapy. Recurrences can be monitored with tumour markers (AFP) in these patients.

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