

Original article:

Retroperitoneal teratoma – a case series

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Abstract:

Introduction: Extra gonadal primary teratoma is usually encountered in infants and children. The involvement of extra-gonadal sites in decreasing order of frequency is mediastinum, sacrococcygeal region, retro peritoneum, pineal gland .

An incidence of teratoma is reported to be 6-11% of primary retroperitoneal tumors, with 60% occurring in children less than 15 years old, 70% of which occurs in infant.

Aim : To study rare entity of retroperitoneal teratoma cases in children less than 15 years

Material & Methods : Prospective study conducted at our centre for identifying cases of retroperitoneal teratoma and management of such patients 5 cases presented of study from June 2012 to May 2016.

Results : Complete excision of tumor offers the best chance of cure. Malignancy is uncommon in retro-peritoneal teratoma hence non mutilating excision is possible and should be attempted . Prognosis is generally good and curative if the tumor is completely removed. Most important prognostic factor is complete removal, however as it is possible that histologically mature tumor may take a malignant clinical course.

Conclusion : Retroperitoneal teratoma comprise 3.5-4% of all germ cell tumors in children and 1-11% of primary retro peritoneal neoplasm . Antenatally Patients usually present with abdominal distension or a palpable mass. An accurate diagnosis of a teratoma cannot be made on clinical basis. Radiological features include presence of calcification, teeth and fat, however calcification cannot be considered an indicator of a benign tumor since 12.5% of calcified tumor are malignant.

Key words : Retroperitoneal Teratoma, Benign, Extragonadal

INTRODUCTION

Germ cell tumor is congenital tumor containing derivatives of all the three germ layers. Germ cell tumors arise due to aberrant migration of germ cells from yolk sac during fetal development. They are frequently seen in gonads. Extra gonadal primary teratoma is usually encountered in infants and children. The involvement of extra-gonadal sites in decreasing order of frequency is mediastinum, sacrococcygeal region, retro peritoneum, pineal gland [1, 2]. They present mainly as an abdominal mass with few other symptoms. Majority of them are benign, situated on the left side and para renal in location; occasional lesions are bilateral. If diagnosed early, they are amenable to curative excision. [2]

AIM

To study rare entity of retroperitoneal teratoma cases in children less than 15 years

MATERIALS AND METHODS

Diagnosed patient retroperitoneal teratoma admitted to the department of Pediatric Surgery, Era's Lucknow Medical college & hospital, Lucknow, India between June 2012 and May 2016 were included sequentially in the study. Preoperative evaluation included plain X-ray, intravenous urography, abdominal ultrasound and contrast enhanced CT scan to delineate the nature and extent of the tumor. Hematological investigations including serum alpha-fetoprotein assay (AFP) were done to obtain preoperative values. Surgery was done through a transperitoneal approach in all cases. The excised

specimens were subjected to detailed histological examination to detect malignant elements.

The patients were reviewed at three weeks when serum AFP levels were estimated.

RESULT

Case no.	Age/sex	Tumor	Imaging	Treatment
1	9months/F	Left pararenal	Plain X-ray, IVU, CT scan	Excision/benign
2	14 years/F	Right pararenal	Plain X-ray, IVU, CT scan	Excision/benign
3	6 years/M	Bilateral	Plain X-ray, CT scan	Excision/benign
4	9 years/F	Left pararenal	Plain X-ray, CT scan	Excision/benign
5	2 years/F	Left pararenal	Plain X-ray, CT scan	Excision/benign

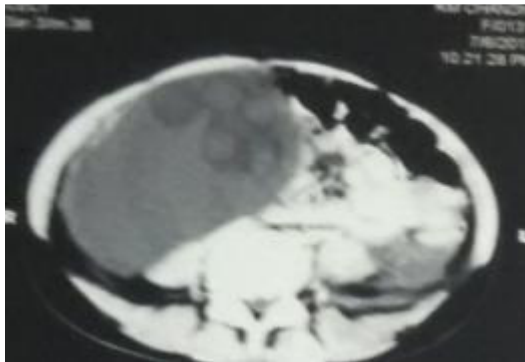


Fig. 1: CT scan showing the lesion on right side of abdomen



Fig. 2: Picture of the excised specimen



Fig. 3: Operative appearance of the lesion

There were 5 patients aged 9 months to 14 years with a female preponderance (Table 1). The mode of presentation was abdominal enlargement with a palpable abdominal mass in all 5 cases. The general condition of all the patient was good despite the presence of a large intra-abdominal tumor. Preoperative serum alpha-fetoprotein was elevated in all the 5 children evaluated. Imaging studies should the presence of a variegated tumor with solid and cystic areas. Majority of the tumors were left-sided and closely-approximated to the kidney with the renal vessels stretched over the surface of the tumor. Two patient had tumor involving both sides of midline. All patients were operated through a supraumbilical transverse transperitoneal approach. The tumor was excised without difficulty; the renal vessels had to be carefully mobilized off the surface of the lesion, allowing preservation of the kidneys in all children, and in one instance a small rent in the left renal vein was repaired. All the specimens were examined in theatre; among the contents were teeth, hair, bone and brown or pale fluid. Histological examination of the lesions showed that all were benign mature tumors. Postoperative recovery was uneventful in all patients. AFP levels had returned to normal levels in all 5 patients at 3 weeks followup.

DISCUSSION

Retroperitoneal teratomas comprise 3.5 – 4% of all germ cell tumors in children^[1,2] Patients present with abdominal distension or a palpable mass. Occasionally, the tumor is

present antenatally and diagnosed at birth,^[3,4] these neonatal teratomas have a higher incidence of malignancy than those in older children^[3,4]. Majority of the tumors in this study (4/5) were left-sided with a female preponderance. All the patients in the present study had a mature benign tumor. In the present series the diagnostic algorithm was palpation of a solid flank mass, plain X-ray to demonstrate calcification or formed bony components like teeth and phalanges (which are pathognomonic). If these were present on X ray, ultrasound was sufficient to define the relationships of the tumor for planning surgery. If formed bony components were not visualised, CT scan was used to define the extent of the disease. Schey and Vesley^[7] have recommended only a plain abdominal X-ray and excision of the tumor if the characteristic calcification is demonstrated. Lack and Travis^[8] have also reported that the presence of bones or teeth on an X-ray was the most helpful in establishing a preoperative diagnosis. The authors suggest that CT scan is useful to delineate the extent of the disease in lesions occupying both sides of the retroperitoneum and those tumors where calcification is not seen on plain X-ray. However, it was seen in some of the patients under evaluation that the degree of tumor adherence to the adjacent structures suggested by CT was more than that found on exploration. CT findings should not therefore prevent surgical exploration of the tumor; even bilateral lesions are amenable to complete removal. Hayasaka and Yamada^[9] have reported internal homogeneity, fat density,

cyst formation and calcification to be important predictors of a benign retroperitoneal tumor on CT. Serum alpha-fetoprotein was elevated preoperatively in six of the patients, it returned to normal after operation and formed a useful marker of monitoring recurrence. All reports agree that complete excision of the teratoma offers the best chance of cure.

Malignancy is uncommon in retroperitoneal teratomas except endodermal sinus tumors and hence non-mutilating excision is possible and should be attempted even in lesions involving both sides of the abdomen. The most important aspect of the excision is to remember the close relationship of these tumors with the kidneys. The renal vessels are invariably stretched out over the

lesion, with care however, they can be separated from the teratoma. Removal of the kidney was not necessitated in any of the patients.

CONCLUSION

Retroperitoneal teratomas are uncommon tumors in children and majority of the lesions are benign. X-ray findings of calcification/ bone/ teeth are pathognomonic. Ultrasonography and CT scan are useful to delineate the extent of the tumor. Despite extensive local spread, the lesions are amenable to curative surgical excision. Recurrence can be monitored with tumor markers like serum alpha-fetoprotein.

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