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Excision of retroperitoneal germ cell tumor in children: A distinct surgical challenge

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ABSTRACT

Purpose: The excision of retroperitoneal germ cell tumor (GCT) is a surgical challenge compounded by the absence of definite surgical guidelines. The aim of this study is to present the surgical difficulties and morbidity associated with resection of these tumors.
Methods: Fifteen patients (7 males and 8 females) undergoing excision of retroperitoneal GCT between February 2008 and February 2016 were evaluated.
Results: Except for an adolescent, the majority of patients were infants (median age: 4 months). The surgical excision entailed extensive vascular dissection in all patients with associated significant blood loss in two, adjacent organ removal in five, and vessel repair in one patient. The resection was complete in all except two patients. Both the patients with incomplete resections had immature teratoma and received postoperative chemotherapy. At a median follow-up of 53 months, 13 patients are alive and disease free, one patient is alive with stable disease, and one patient had died owing to respiratory complications. There were no local recurrences in the patients with complete excision.
Conclusion: Although the outcomes are excellent after surgery, resection of retroperitoneal GCT is a distinct surgical challenge. The surgical difficulties emanate from the need for extensive vascular dissection and risk to adjacent structures.
Level IV evidence: Therapeutic study.

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The most common site of extragonadal germ cell tumors (GCTs) in children is the sacrococcygeal region while they are rare in the retroperitoneum [1]. Primary retroperitoneal GCTs account for less than 4% of all GCTs and up to 11% of all retroperitoneal neoplasm's [2]. The large size at presentation and the close relation to the kidneys and major blood vessel and their branches in the retroperitoneum make resection of these tumors a surgical challenge. Furthermore, the rarity of these neoplasms has led to publications of case reports or small case series of patients treated over long periods [3–8]. Consequently, definite surgical guidelines are wanting and the morbidity associated with resection of these tumors is underreported. With a special reference to the surgical challenges and postoperative morbidity, we reviewed our experience with the management of retroperitoneal GCTs in children.

1. Material and methods

All patients with primary retroperitoneal GCT treated between February 2008 and February 2016 were included in this study. Demographic details, clinical presentation, preoperative imaging and investigations, surgical procedure and postoperative course and follow-up of the patients were collected from the prospectively maintained database. A comprehensive clinical evaluation preceded imaging with computerized tomography (CT) scan of the abdomen and thorax. The extent of disease and relation to the kidney and major blood vessels of the retroperitoneum were evaluated in all patients [Fig. 1]. The hematological investigations included serum alpha-fetoprotein (AFP), beta human chorionic gonadotropin (HCG) and lactate dehydrogenase (LDH). The diagnosis of retroperitoneal GCT was established in the presence of characteristic imaging features, which included the presence of calcification, fat density, cystic areas, etc. Only when the imaging features were uncharacteristic and the tumor markers negative, an image-guided core biopsy was performed. All patients were discussed in multidisciplinary tumor board meeting for treatment planning.

1.1. Surgery

The tumors were resected using a transverse abdominal incision at the transpyloric plane. The retroperitoneum was exposed by reflecting

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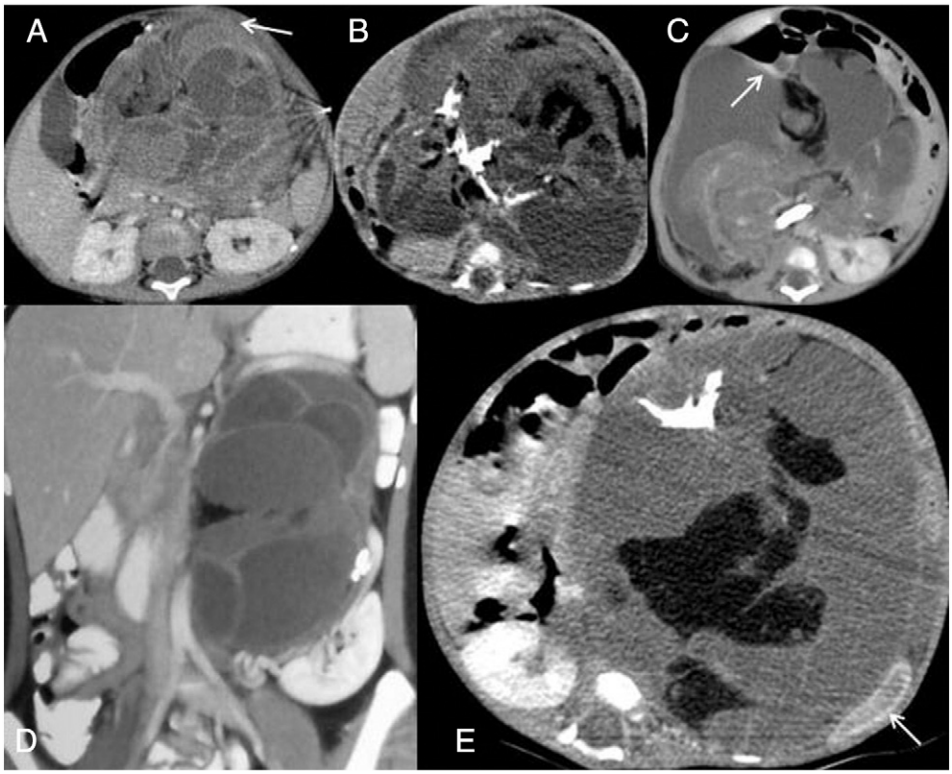

Fig. 1. Computed tomography images showing retroperitoneal teratoma. (A) The mass is extending into the anterior abdominal wall (arrow). (B) Large lesion filling the abdominal cavity and displacing the vessels, which are not seen distinctly. (C) Inferior vena cava is displaced anteriorly (arrow). (D) Kidney is pushed downwards and laterally till the pelvic brim with stretching and encasement of the renal artery (not shown). (E) Predominantly left sided mass causing extensive displacement and distortion of the kidney (arrow) with reduced parenchymal enhancement as compared to the right kidney.

the colon and the spleen towards the midline. The tumor was dissected all around avoiding a rupture of the capsule. Displaced or encased blood vessels were meticulously dissected from the tumor, securing all the feeding or draining tributaries. All efforts were made to prevent removal of adjacent organs. Nevertheless, the exceptional presence of extensive vascular stretching or distortion compromising the vascularity of the affected organ necessitated an excision.

The entire excised specimen was evaluated for malignant components. In general, adjuvant therapy was not offered to patients with completely excised mature or immature teratoma, however, chemotherapy was offered when there was an unanticipated incomplete excision of immature teratoma. All patients were followed up with serum AFP estimation and abdominal ultrasound at regular intervals.

Table 1
Characteristics of all patients with retroperitoneal teratoma.

Patient	Age at surgery (months)	Sex	AFP (ng/ml)	Size (cm)	Biopsy	Treatment	Complications	Pathology	Follow-up (months)
1	4	F	483	20 × 15 × 10	YES	Complete resection	Vein injury, fever	MT	NED, 46
2	10	F	17.46	15 × 13 × 18	NO	Complete resection	Nephrectomy Prolong ICU stay	MT	NED,3
3	1	F	3178	20 × 16 × 18	NO	Partial resection, chemotherapy	None	IT Grade II	AWD, 32
4	2	M	4605[a]	15 × 12 × 3.5	YES	Partial resection, chemotherapy	Blood loss, Abdominal collection	IT Grade III	DOC, 5
5	19 years	F	1.84	16 × 8 × 13	NO	Complete resection	Blood loss	MT	NED, 60
6	28	M	3.65	13 × 10 × 6	YES	Complete resection	Stomach, skin resection, infection	MT	NED, 93
7	2	M	2826[a]	11 × 9 × 8	YES	Complete resection	Stomach resection	IT Grade III	NED, 101
8	3	M	45	17 × 10 × 8	Yes	Complete resection	Stomach resection	MT	NED,3
9	4	F	83. 5	8.5 × 9.5 × 8.5	No	Complete resection	Fever, intestinal obstruction	IT Grade NA	NED, 79
10	6 years	F	839[a]	10.4 × 9.1 × 10.5	NO	Complete resection	Nephrectomy	IT Grade II	NED,115
11	4	M	103	15 × 20	YES	Complete resection	None	IT Grade I	NED, 79
12	23	F	3.09	8.3 × 9.3 × 10.9	NO	Complete resection	None	MT	NED, 69
13	23	M	2.2	9.2 × 6.6 × 9.1	NO	Complete resection	None	MT	NED, 27
14	3	F	?	7.7 × 10 × 9	NO	Complete resection	Chyle leak	MT	NED, 11
15	9	M	11.74	13.5 × 11 × 8.5	NO	Complete resection	None	MT	NED, 15

AFP = alphafetoprotien, M = male, F = female, IT = immature teratoma, MT = mature teratoma, NA, not available, NED = no evidence of disease, AWD = alive with disease, DOC = died of other cause.
[a] Elevated for age.

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