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EXTRA-GONADAL ABDOMINAL GERM CELL TUMOR IN CHILDREN: PERIOPERATIVE CHALLENGES

Oncology

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ABSTRACT

Background: Primary extra-gonadal germ cell tumors (GCTs) are rare and account for less than 4% of all GCTs. Prognosis is usually excellent after complete excision but absence of clearly defined capsules and surgical guidelines with large size of tumor made the excision difficult. Various perioperative problems reported in literature during and after excision of retroperitoneal GCTs. Here, we present our experience with non-gonadal abdominal GCTs and highlighted the perioperative problems and morbidity associated with management of these tumors.

Method: From July 2012 to June 2019, all patients with primary abdominal GCTs were retrospectively reviewed. Demographic features; clinical, laboratory, radiographic, and intraoperative findings; perioperative complications; and pathology results were assessed.

Result: In last seven years, 17 patients with abdominal GCTs were managed. Diagnosis were teratoma in 13 [Mature- 8; Immature- 5] and yolk sac tumor (YST) in 4 patients. 6 patients were female and age ranged from 2 months to 6 years (median 14 months). Presentations were abdominal distention and palpable lump, Fever, pain, respiratory distress, vomiting, early satiety and sub-acute bowel obstruction. Serum alfa-protein was raised in all patients. Complete excision was achieved in 15 patients. 5 patients were managed by chemotherapy with surgery while in 10 patients, only surgery was performed. Perioperative problems include excessive blood loss (2), excision of adjacent organ (2), incomplete resection (2), hypothermia (1), respiratory problems (3), chemo-related toxicity (1) and adhesive bowel obstructions (6). Re-intervention needed in 5 patients [Re-exploration for adhesive bowel obstruction-4; and radiological drainage of retroperitoneal collection-1] within 3 month of first surgery. One patient was died while 3 patients were lost in follow-up. Follow-up ranged from 6 – 60 months.

Conclusion: The management of retroperitoneal/abdominal GCT is challenge. Various surgical difficulties and morbidity associated with resection of these tumors depend on its nature, size and site of tumor and on age of the patient.

KEYWORDS

Extra-gonadal; Germ Cell Tumor, Teratoma; Yolk Sac Tumor; Complications.

Primary extra-gonadal germ cell tumors (GCTs) are rare and account for less than 4% of all GCTs and up to 11% of all retroperitoneal neoplasms **[1,2,3].** The overall incidence of GCT is 0.9/100,000. In neonates, mature and immature teratomas predominate. In the first years of life, the overall incidence of GCT decreases for both sexes. Among toddlers, the relative proportion of malignant entities, especially yolk sac tumors (YST) increases. The incidence of gonadal tumors, mainly seminomas and dysgerminomas, increases with the onset of puberty **[1,2,3].** Origin of abdominal GCTs near close proximity of vital organs and major vessels and its tendency to grow between organs made the excision difficult **[1].** Prognosis is usually excellent after complete excision but absence of clearly defined capsules and surgical guidelines with large size of tumor leads to various perioperative problems and post-surgical complications **[1,2,3].**

Here, we present our experience with non-gonadal abdominal GCTs and highlighted the perioperative problems and morbidity associated with management of these tumors.

MATERIAL & METHOD:

From July 2012 to June 2019, all patients having primary abdominal GCTs with adequate records and follow-up period up to 6 months were retrospectively reviewed and included in study. Demographic features; clinical, laboratory, radiographic, and intraoperative findings; perioperative complications; and follow-up results, etc. were assessed.

RESULT:

From July 2012 to June 2019, total 17 patients with abdominal GCTs were included in the study. Diagnosis were teratoma in 13 [Mature- 8; Immature- 5] and yolk sac tumor (YST) in 4 patients. Patients with immature teratoma, one had grade-III, three grades-II and one had grade-I immaturity. 6 patients were female [M: F= 2:1] and age ranged from 2 months to 6 years (median 14 months). Presentations were abdominal distention and palpable lump, Fever, pain, respiratory distress, vomiting, early satiety and sub-acute bowel obstruction **[Table: 1]**. Serum alfa feto-protein (AFP) was raised in all patients and more than 10 fold rise observed in 8 patients while raised serum beta-HCG was observed in only 6 patients . Core biopsy was performed in 6 patients. Complete excision was achieved in 15 patients. 5 patients were managed by chemotherapy and surgery while in 10 patients, only surgery was performed. Neoadjuvant chemotherapy was given in 3 patients with YST (PEB-regime). Perioperative problems include excessive blood loss (2), excision of adjacent organ (2), incomplete resection (2), hypothermia (1), respiratory problems (3), chemo-related toxicity (1) and adhesive bowel obstructions (6) [**Table: 2].** Total 19 perioperative complications encountered in 8 patients (47%). Re-intervention needed in 5 patients. 4 patients needed laparotomy and adhesiolysis for adhesive bowel obstruction within 3 month of surgery while one patient with incomplete resection of grade-III immature teratoma receiving chemotherapy (PE-regime), developed retroperitoneal collections and needed guided percutaneous drainage. One patient was died due to aspiration during feeding in post-operative period while 3 patients were lost in follow-up. Follow-up ranged from 6 – 60 months.

Table-1: Presentations at admission (n=17) –

Symptoms	No. of Patient (%)
Abdominal distention and palpable lump	14 (82.35)
Fever	4 (23.53)
Pain / Heaviness /Backache	8 (47.050
Respiratory distress	3 (17.65)
Vomiting,	5 (29.41)
Early satiety	4 (23.53)
Sub-acute bowel obstruction	4 (23.53)

Table-2: Peri-operative Complications (n=8) –

Excessive blood loss	2
Excision of adjacent organ	2

Incomplete resection	2
Hypothermia	1
Respiratory problems	3
chemo-related toxicity	1
Adhesive bowel obstructions	6
Wound infection	2

DISCUSSION:

A bimodal peak of incidence is seen with retroperitoneal GCTs, first at six months of life and then in adolescence age **[1].** The most common primary sites of GCTs are ovary (26%), coccyx (24%), testis (18%) and brain (18%) in children younger than 15 years of age. In infancy, coccygeal tumors are most prevalent. Girls have a higher overall incidence of GCTs, but boys are more at risk of malignant GCTs. Other extra-gonadal sites are the mediastinum (4%), retroperitoneum (4%) and the vagina (2%). Malignant germ-cell tumors (GCT) account for 2.9% of all malignant tumors of children younger than 15 years of age **[2,3].** In these patients, malignant non-seminomatous GCTs may be diagnosed clinically due to the increased serum or cerebrospinal fluid levels of the tumor markers AFT and/or beta-HCG **[1,2,3].** In this cohort, AFT was raised in all patients.

GCT is characterized by a high heterogeneity of their histological differentiation and histogenic origin is still debatable, but they show a similar histological pattern independent of their primary site or sex **[1-4]**. Extra-gonadal GCTs may reach a large size with no or relatively few symptoms. The most common presenting symptoms for patients with endodermal primitive tumors are rapidly enlarging pelvic mass and pain **[5,6].** Although the imaging characteristics can be suggestive of teratoma, they may be confused with other retroperitoneal tumors like sarcoma, neuroblastoma, adrenal tumors, and lymphomas. Malignant GCTs are rare and most of the time in young patients, it is yolk sac tumor (endodermal sinus tumor) **[1-6].** We encountered 4 patients with YST. These tumors are highly sensitive to chemo-radiation. Doubt of other malignancy on radiology leaded for core biopsy. Neoadjuvant chemotherapy (PEB-regime; Cisplatinum, Etoposide and Bleomycin) was given in all these patients followed by surgery and adjuvant therapy. One child with incomplete resection and grade-III immature teratoma was also treated with three cycle of PE-regime (Cisplatinum and etoposide) **[1,7].**

Most of the patients in this cohort are below 2 year of age and leading complaints were abdominal distension and pain/heaviness. Respiratory distress due to diaphragmatic splinting and chest infection was observed in 3 patients. Large size of abdominal masses in small sized patients displaced and compressed all vital structure. It resulted in early satiety, recurrent vomiting and constipation. Lack of well-defined tumor capsule and nature of GCTs to grow between the organs planes and sometime invasion and encasement of adjacent organs or vessels resulted in difficult surgery. Excessive bleeding in two patients occurred due to injury to inferior vena cava and its tributaries and required 2 unit of blood transfusion. One patient developed hypothermia. Tumor resection is considered complete, if it is performed as "en-bloc" resection of the tumor including the adjacent organ of origin. There is no role of debulking surgery in pediatric GCTs. Surgery of metastases is not indicated **[7]** unless they show an insufficient response to chemotherapy **[1,7].** Excision of part of small bowel and stomach was required in one patient while splenectomy was required during excision of tumor in one patient in our series.

Excision of large tumor resulted in wide raw/rough areas in abdominal cavity. It might favor bowel adhesions. We encountered 6 patients with adhesive bowel obstruction in postoperative period whom 4 patient required re-exploration within 3 months of follow-up. Jones NM, *et al* [**8]** and Qureshi SS, *et al* **[3]** reported intestinal obstruction in their series, which required laparotomy. 4 patient required postoperative ventilator support for 12- 36 hours due to pre-existing chest infections. One patient was died on 5th postoperative day due to aspiration pneumonitis followed by respiratory failure during feeding. In our series majority of the patients presented with an abdominal mass and few unusual presentations included hypertension with cardiac failure and abdominal wall abscess. De Backer, *et al* **[9],** Jones NM*, et al* **[8]** and Qureshi SS*, et al* **[3]**; all are agreed that excision of retroperitoneal GCTs is a distinct surgical challenge while Luo, *et al* and Chaudhry, *et al* did not encounter any major challenges in the resection of these tumors **[10,11].** Perioperative complications include esophagogastric, choledochal and vena cava tear, cyst rupture, excessive bleeding, bowel obstruction, cardiac dysfunctions, chylous leak, infections etc. Earlier literature showed surgical mortality of 54%, however, with improvement in anesthesia and surgical techniques; surgical mortality is very rare **[12].** De Backer, *et al* reported a single mortality owing to persistent bleeding in their series but there was no procedure-related mortality in our series.

In conclusion, management of retroperitoneal/abdominal GCT is challenge. Total 19 perioperative complications encountered in 8 patients (47%) in our series. Various surgical difficulties and morbidity associated with resection of these tumors which depend on tumor nature, size and site of tumor and on age of the patient. These difficulties may be compounded by blood loss, incomplete removal of tumor and occasionally excision of adjacent organs.

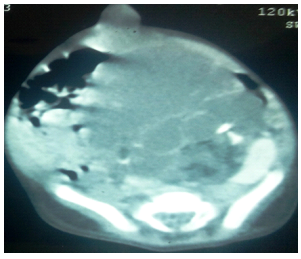

Figure-1: CT-picture of Large Immature Teratoma.

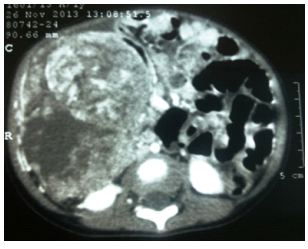

Figure-2: CT picture of right sided Yolk Sac Tumor.

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