Gastric teratoma in children: Our experience

Praveen Mathura, Rahul Gupta, Girish Prabhakar, Lila Dhar Agrawal, Randhir Singh Rao, Ram Babu Goyal

Paediatric Surgery, Sir Padampat Mother & Child Health Institute, Sawai Man Singh Medical College, Jaipur, Rajasthan, India
Paediatric Surgery, Sardar Patel Medical College, Bikaner, Rajasthan, India
GastroIntestinal Surgery, Sawai Man Singh Medical College, Jaipur, Rajasthan, India

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Summary Background/Introduction: Gastric teratoma is an extremely rare tumor, almost exclusively benign, accounting for < 1% of all teratomas in infants and children. Purpose/Aim: This work aimed to study the clinical presentation, investigation modalities, intraoperative findings, histopathological types, and surgical outcome of gastric teratoma in children. Methods: A retrospective study was performed from 1993 to 2013 in two pediatric institutes. Results: There were eight patients with gastric teratoma and all of them were male. Seven (87.5%) patients presented in infancy, out of which four (50%) patients were neonates, while one (12.5%) patient was a toddler. The manifestations were as follows: a palpable abdominal mass in seven (87.5%), abdominal distension in five (67.5%), anemia in four (50%), respiratory distress in three (37.5%), gastric outlet obstruction with recurrent vomiting in three (37.5%), and abdominal pain, anorexia, and melena in one (12.5%) each. Ultrasonography showed a solid cystic mass with mixed echogenicity in all the patients, while calcification was seen in seven (87.5%) cases. Computed tomography, which was performed in four (50%) patients, clinched the preoperative diagnosis. A growth was present on the posteroinferior wall near the greater curvature in seven (87.5%) cases, while in one (12.5%) patient, it was arising from the lesser curvature. Excision of the tumor was performed in all patients. Histopathology was mature type in six (75%) cases, and immature Grade 1 and 2 in one (12.5%) each. Complications were seen in five (62.5%) cases, and one (12.5%) mortality was observed in our series. All other patients were doing well in the postoperative period.

Conflicts of interest: The authors declare no conflicts of interest regarding the publication of this paper.

* Corresponding author. 202 A, A3 block, Kamal Apartment 2, Banipark, Jaipur, Rajasthan, India.
E-mail addresses: meetsurgeon007@yahoo.co.in, meetsurgeon007@gmail.com (R. Gupta).
These authors contributed equally to this work.

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1. Introduction

Gastric teratoma is an extremely rare tumor, almost exclusively benign, accounting for <1% of all teratomas in infants and children. These tumors usually occur in infants (94%), especially in the neonatal period. However, there have been reports of this tumor occurring in older children. Approximately 120 cases have been reported in the English literature to date.

Gastric teratomas form a distinct subset of teratomas as follows: (1) unusual male predominance (>95%), compared to female preponderance (65–70%) at other sites (e.g., sacrococcygeal teratoma); (2) not associated with any congenital anomalies in contrast to 10–15% incidence at other sites; (3) not associated with any syndrome, except for a solitary case report of gastric teratoma with the Beckwith–Wiedemann syndrome and peritoneal gliomatosis; (4) not associated with the dorsal body axis and embryonic body wall; and (5) almost always benign in nature (with the exception of 3 cases reported in the literature), as compared to 10–39% incidence of malignancy at other sites such as the sacrococcygeal region, mediastinum, and gonads. Owing to the extreme rarity and unique characteristics of this entity, we present, herein, a series of gastric teratomas presented to our institutes and share our experience along with a review of literature.

We aimed to study the clinical presentation, investigation modalities, intraoperative findings, histopathological types, and surgical outcome of gastric teratomas in children.

2. Methods

We present a retrospective study of pediatric patients with gastric teratomas. All clinical, operative, and postoperative records of patients admitted to two pediatric institutes over a period of 21 years from 1993 to 2013 were reviewed. The inclusion criterion was children diagnosed with gastric teratoma.

Charts were reviewed according to age, sex, chief complaints, associated signs and symptoms, associated anomalies, preoperative diagnosis/method of diagnosis, location of tumor, type of growth (endogastric/exogastric), surgical procedure, histopathology report, and postoperative complications, including recurrence if any. All possible radiologic investigations, including abdominal radiographs, ultrasonography, and computed tomography (CT) scans were carefully reviewed in order to identify calcification and growth characteristics. The patients were followed up for 5 years, while the last patient is being under supervision for 2 years.

3. Results

The results of the study are shown in Table 1. There were eight patients with gastric teratomas and all of them were male. Their ages ranged from 1 day to 3.5 years. Seven (87.5%) patients presented in infancy, out of which four (50%) were neonates, while one (12.5%) was a toddler. The manifestations were as follows: a palpable abdominal mass in seven (87.5%), abdominal distension in five (67.5%), anemia in one (12.5%) each. Ultrasonography showed a solid cystic with mixed echogenicity mass in all the patients, while calcification was seen in seven (87.5%) cases. CT, which was performed in four (50%) patients, provided classical findings of teratomas, seen as the presence of a well-encapsulated, multiseptate, enhancing, solid cystic mass with areas of calcification (Figure 1) and clinched the preoperative diagnosis.

Operative intervention was carried out in all the patients. A growth was present on the posteroinferior wall near the greater curvature in seven (87.5%) cases (Figures 2–4), while in one (12.5%) patient, a growth was arising from the lesser curvature. Tumor was purely endogastric in six (75%) cases, while it was predominantly exogastric with minimal endogastric component in two (25%) patients. Excision of the tumor along with a small fringe of the gastric wall and primary gastric closure was performed in all patients. Histopathology was mature type in six (75%) cases, and immature Grades 1 and 2 in one (12.5%) each. Complications were seen in five (62.5%) cases, and there was one (12.5%) mortality in our series. All other patients were doing well in the postoperative period. Recurrence was not seen in any of our cases.

4. Discussion

The word teratoma is derived from the Greek word “teraton,” meaning “monster.” Teratomas are relatively common embryonic neoplasms arising from totipotent cells and contain elements from all the three germ layers. In infancy and early childhood, the most common site of teratomas is the extragonadal region, which includes sacrococcygeal, mediastinal, presacral and rarely intracranial, retroperitoneal, and cervical regions, whereas after childhood, they are more usually located in the gonads.

Gastric teratomas, a unique subgroup of teratomas, present especially in neonates and infants. It was first reported in 1922 by Eustermann and Sentry. The exact cause of gastric teratomas is not known, and, similar to other...
Table 1  Clinical features, investigative modalities, intraoperative findings, and surgical outcome in our series.

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Chief complaints</th>
<th>Associated signs &amp; symptoms</th>
<th>Pallor (Hb%)</th>
<th>USG showing mixed echogenic solid cystic mass</th>
<th>Calcification</th>
<th>CT showing encapsulated multiseptate heterogeneous solid cystic mass</th>
<th>Preoperative diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3.5 y</td>
<td>M</td>
<td>Palpable abdominal mass</td>
<td>Abdominal pain Abdominal distension Vomiting Anorexia</td>
<td>Marked (7.5 g%)</td>
<td>Present Extension—epigastrium to pelvis</td>
<td>Present</td>
<td>Present, Contrast enhancement</td>
<td>Present</td>
</tr>
<tr>
<td>2</td>
<td>1 d</td>
<td>M</td>
<td>Palpable abdominal mass Respiratory distress</td>
<td>Nil</td>
<td>Nil (13.6 g%)</td>
<td>Present Central necrosis Extension—epigastrium to hypogastrium</td>
<td>Present</td>
<td>Present Large intratumoral vessels Displacement of adjacent organs Not performed (nonavailability)</td>
<td>Present</td>
</tr>
<tr>
<td>3</td>
<td>4 mo</td>
<td>M</td>
<td>Palpable abdominal mass</td>
<td>Abdominal distension Melena</td>
<td>Moderate (8.5 g%)</td>
<td>Present Extension—epigastrium to pelvis</td>
<td>Present</td>
<td>Not performed (nonavailability)</td>
<td>Absent</td>
</tr>
<tr>
<td>4</td>
<td>5 mo</td>
<td>M</td>
<td>Abdominal distension Palpable abdominal mass</td>
<td>Vomiting</td>
<td>Mild (9.5 g%)</td>
<td>Present Extension—epigastrium to hypogastrium</td>
<td>Present</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>5</td>
<td>4 d</td>
<td>M</td>
<td>Palpable abdominal mass Respiratory distress</td>
<td>Born premature at 37 wk</td>
<td>Nil</td>
<td>Present Extension—epigastrium to pelvis</td>
<td>Present</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>6</td>
<td>2 d</td>
<td>M</td>
<td>Palpable abdominal mass Respiratory distress Abdominal distension</td>
<td>Nil</td>
<td>Nil (14.6 g%)</td>
<td>Present Extension—epigastrium to hypogastrium</td>
<td>Present</td>
<td>Not performed</td>
<td>Absent</td>
</tr>
<tr>
<td>7</td>
<td>15 d</td>
<td>M</td>
<td>Abdominal distension</td>
<td>Born premature at 34 wk</td>
<td>Nil</td>
<td>Present Multicystic mass</td>
<td>Absent</td>
<td>Not performed</td>
<td>Absent</td>
</tr>
<tr>
<td>8</td>
<td>2 mo</td>
<td>M</td>
<td>Abdominal distension Palpable abdominal mass</td>
<td>Vomiting</td>
<td>Mild (10.5 g%)</td>
<td>Present Extension—epigastrium to pelvis</td>
<td>Present</td>
<td>Not performed (nonavailability)</td>
<td>Absent</td>
</tr>
</tbody>
</table>

Hb = hemoglobin.
<table>
<thead>
<tr>
<th>Location of tumor</th>
<th>Growth characteristics</th>
<th>Size (cm)</th>
<th>Treatment: excision along with small fringe of gastric wall &amp; primary closure</th>
<th>Cut surface of tumor</th>
<th>Histopathology</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Exogastric</td>
<td>$11 \times 15 \times 10$</td>
<td>Yes</td>
<td>Not performed</td>
<td>Mature</td>
<td>Seroma</td>
</tr>
<tr>
<td>Lesser curvature</td>
<td>Exogastric</td>
<td>$9.2 \times 8.2 \times 7.3$</td>
<td>Yes</td>
<td>Variegated Grayish white to brown Solid areas filled with brown-colored fluid</td>
<td>Grade 2 immature Absence of microfocus of yolk sac tumor</td>
<td>Nil</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Predominantly exogastric Minimal endogastric component</td>
<td>$10 \times 8 \times 7$</td>
<td>Yes</td>
<td>Variegated Grayish white to brown</td>
<td>Mature</td>
<td>Nil</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Exogastric</td>
<td>$9.5 \times 7.5 \times 7$</td>
<td>Yes</td>
<td>Not performed</td>
<td>Mature</td>
<td>Nil</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Exogastric</td>
<td>$10.5 \times 7.5 \times 6.5$</td>
<td>Yes</td>
<td>Not performed</td>
<td>Mature</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Exogastric</td>
<td>$7 \times 6.5 \times 5.5$</td>
<td>Yes</td>
<td>Not performed</td>
<td>Mature</td>
<td>Wound infection</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Predominantly exogastric Minimal endogastric component</td>
<td>$14 \times 9 \times 6$</td>
<td>Yes</td>
<td>Exogastric—large cystic areas, endogastric—mainly solid (grayish white areas, foci of cartilage &amp; bone)</td>
<td>Grade 1 immature (neuroepithelial elements)</td>
<td>Mortality (sclerema, &amp; succumbed to septicemia on the 5th postoperative day) Seroma</td>
</tr>
<tr>
<td>Greater curvature posteroinferior wall</td>
<td>Exogastric</td>
<td>$7.5 \times 6 \times 5.5$</td>
<td>Yes</td>
<td>Not performed</td>
<td>Mature</td>
<td>Seroma</td>
</tr>
</tbody>
</table>
extragonadal teratomas, it has been proposed to originate from migrated totipotential germ cells.\textsuperscript{15}

It commonly presents in a neonatal age group, which was seen in 50% of our cases. Late presentation beyond infancy was seen in one of our patients. It has also been reported in the adult age group.\textsuperscript{3,16} The most common presentation is a palpable abdominal mass (75%) in the epigastrium and the left side of the abdomen with abdominal distension, as per the literature.\textsuperscript{17} The mass is usually bosselated, firm in consistency, and mobile in both directions, and moves with

**Figure 1** Abdominal CECT scan (with intravenous contrast) of a 3.5-year-old child. Images of (A) coronal and (B,C) transverse sections showing an extremely large, well-encapsulated, multiseptate, heterogeneous (solid cystic) enhancing mass of $11 \times 15 \times 10$ cm$^3$ in size with intratumoral calcifications arising from the left half and occupying most of the abdominal cavity. The mass is displacing the bowel loops inferolaterally and extending into the pelvis. Low density indicates cystic tissue and high density indicates calcifications in the tumor.

**Figure 2** Intraoperative photograph of a 3.5-year-old child showing a (A) massive multicystic mass. (B) The mass is seen to be arising from the posteroinferior wall of the stomach along its greater curvature.
Figure 3  Intraoperative photographs of (A) a 5-month-old child showing preoperative marking of the lump, (B) involvement of the posteroinferior wall of the stomach along its greater curvature, (C) involvement of the lesser sac, and (D) excised specimen of gastric teratoma in toto.

Figure 4  Intraoperative photographs of a (A) 2-month-old child showing massive abdominal distension, stretched abdominal wall with visible veins, and bilateral hydrocele; (B and C) involvement of the posteroinferior wall of the stomach along its greater curvature, and (D) excised specimen of gastric teratoma in toto.
respiration.2,16,17 The reported incidence of the presence of only a distended abdomen is ~56%.17 The other presenting features are as follows: hematemesis and melena due to gastrointestinal bleeding in patients in whom the tumor grows intramurally and results in ulceration of the overlying mucosa; feeding problems and respiratory distress due to splinting of the diaphragm as a result of its anatomical location and an enormous size of the tumor; vomiting due to gastric outlet obstruction; fever; anorexia and constipation; weakness, anemia, and weight loss; and sometimes abdominal pain in the older age group.1,16 Spontaneous rupture and perforation of large gastric teratomas have also been reported.18 Large tumors may cause obstetric complications (polyhydramnios, premature labor, or dystocia) in the perinatal period because of gastric obstruction produced by the mass, as seen in one of our cases.

In most of the cases, the preoperative diagnosis of gastric teratoma is difficult. Plain films usually reveal a soft tissue mass with associated irregular areas of calcifications. The presence of irregular areas of calcification on abdominal radiographs or on abdominal CT may give some clues to the diagnosis.17,18 Calcification is present in 35–60% of cases, according to various series, but in our study, it was seen in 87.5% of children.17,18 Most mature cystic teratomas can be diagnosed by ultrasonography. It demonstrates a large midline heterogeneous mass with mixed echogenicity. The other common appearance is a cystic lesion with a densely echogenic tubercle (Rokitansky nodule) projecting into the cyst lumen.19 Origin of tumor is usually not appreciated. Mild ascitis is usually present. Prenatal detection by ultrasonography has also been reported in the literature.20 The diagnosis of mature cystic teratomas is usually appreciable on CT, with findings of a well-defined upper abdominal mass with separate cystic and solid components in varying proportions, discrete areas with densities similar to that of fat, and coarse globular calcifications within solid components, areas of necrosis, and intratumoral vessels. A floating mass of hair can sometimes be identified at the fat–aqueous fluid interface.21 Tumor arises most commonly from the posterior wall of the stomach near the greater curvature, as seen in 87.5% of our cases. The other sites where it has been seen are the antrum, prepyloric region, anterior wall, fundus, lesser curvature, cardiac orifice, and very rarely the entire stomach.1–21 Some of these tumors are pedunculated.22,23 The mass is usually large and extends into the retrogastric tissues. The left kidney, spleen, visceral surface of the liver, and bowel loops are pushed by the tumor.

Exogastric in up to 70% of cases, while it is endogastric in 30% of cases.22,23 Contrast enhanced computed tomography (CECT) is the modality of choice, as it detects the organ of origin, its relation with other organs and major blood vessels, tumor extent, and the presence of calcifications.

Age-related serum alpha-fetoprotein levels are abnormally raised when there is presence of intestinal tissue in mature teratoma, in case of immature teratoma or its malignant counterpart. Baseline alpha-fetoprotein and beta-human choriionic gonadotropin levels help in postoperative follow-up to detect recurrence or presence of residual tumor and malignant transformation of the tumor.14–18

The differential diagnosis of a gastric teratoma in a neonate (with regard to the calcified mass in the left upper quadrant) would include mesoblastic nephroma, congenital neuroblastoma, infantile hemangioepithelioma, hepatoblastoma, Wilms’ tumor, and retroperitoneal teratoma.17,24,25 Gastric teratomas constitute 1.6% of all abdominal teratomas.25 Other differential diagnoses of pediatric abdominal masses include pancreatic cyst, omental cyst, mesenteric lymphangioma, duplication cyst, splenic cyst, rhabdomyosarcoma, and liposarcoma.17,24,26 Other benign gastric tumors in children include hyperplastic and adenomatous polyps, leiomyomas, and lipomas.1

Treatment of choice for gastric teratomas is total excision along with a small fringe of the involved gastric wall and primary closure (gastroplasty). If the tumor grows intramurally, then a greater area of the stomach may be involved. Partial, subtotal, and total gastrectomies have been performed as per the extent of stomach involvement.1,21

Pathological examination reveals mature adipose tissue, cartilage, smooth muscle, gastric mucosa, brain, choroid plexus, and skin. Bone, bone marrow, and teeth are pathognomonic of teratoma, but less frequently seen in gastrointestinal teratomas. Gastric teratomas, like other teratomas, are classified into three types according to the histological composition.12,14,28,29 (1) Mature variety, which forms bulk of the reported cases, consists of well-differentiated tissue, i.e., mature neuroglial tissue, along with other derivatives of all germinal layers. These types of teratomas are considered benign tumors and are given Grade 0. (2) Immature gastric teratomas, with ~20 of cases reported in the literature, consist of immature/embryonic-appearing neuroglial and neuroepithelial tissues (varying degrees of immature fetal tissues). Immature teratomas are graded from 1 to 3, according to the amount of immature tissue contents (mainly neural elements) and the degree of mitotic activity according to the Norris grading system. In Grade 1, the immature neuroectodermal tissue is confined to one site in a slide, whereas in Grades 2 and 3, the immature tissue is found in less than four fields per slide and more than four fields per slide, respectively. Malignant potential is present in immature variety, and therefore histopathological examination is meticulously carried out to detect the presence of microfocus of yolk sac tumor, and in its absence, only close monitoring is carried out.12,14,28,29 (3) Malignant type contains at least one of the malignant germ cell elements. The term “malignant teratoma” is restricted to the yolk sac tumor and choriocarcinoma.

Complete surgical excision is sufficient for mature and Grade 1 immature teratomas. Optimal treatment by complete resection followed by close observation and follow-up, withholding chemotherapy until there is an evidence of disease recurrence, is recommended for Grade 2 and 3 immature teratomas. This principle is generally followed for all extragonadal immature teratomas.29,30 In a case where the alpha-fetoprotein level starts rising after a few months of complete excision of the teratoma, chemotherapy is administered.29,30
In our patient with Grade 2 immature teratoma, no recurrence was seen postoperatively. Complete surgical excision is curative even in malignant cases.

The prognosis following complete surgical excision of a gastric teratoma, both mature and immature types, has been shown to be excellent. It holds good, even if there is extension to adjacent organs. Excision of tumor offers recurrence-free survival without requiring chemotherapy or radiotherapy. One case of recurrence has been reported in the literature.

5. Conclusion

Gastric teratoma is a very rare tumor occurring in neonatal period and infancy, and is extremely rare beyond early childhood. It is almost always benign and has predilection for male sex. It commonly presents as a palpable abdominal mass. Evaluation by CT helps exclude other palpable masses encountered in infancy; it can demonstrate the gastric origin and may clinch the preoperative diagnosis. Complete surgical excision is curative in all cases, with close observation and follow-up being recommended in Grade 2 and 3 immature teratomas. Postoperatively, the prognosis is excellent.

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References