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CASE REPORT

Malignant gastric teratoma: report of two cases from a single center

Manoj Saha

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Abstract Gastric teratomas (GT) are rare, accounting for less than 1% of all teratomas. GT usually presents with an abdominal mass, but rarely does it present with gastrointestinal bleeding also. Gastric teratomas are rarely malignant and only two cases are reported in the literature. We treated two cases of malignant GT (endodermal sinus tumor) in infants. Both the patients were male. Surgical excision was done in both the cases. One patient received adjuvant chemotherapy. Both the patients are surviving without recurrence after 5 and 6 years of surgical excision, respectively. Prognosis of GT is excellent even when it is malignant.

Keywords Children · Teratoma · Gastric teratoma · Malignant gastric teratoma · Endodermal

Introduction

Teratomas are embryonic neoplasms, which arise from totipotent cells and contain elements from all three germ layers. Teratomas can occur at various locations such as brain, nose, tongue, neck, mediastinum, retroperitoneum, attached to coccyx, and organs like heart, liver, and stomach. Sacrococcygeal teratoma is the single most common teratoma found in newborn babies. Gastric teratoma is a rare tumor and accounts for less than 1% of all teratomas in infants and children [1]. To date, less than 100 cases of gastric teratomas have been reported in the literature [2, 3]. Gastric teratomas are usually benign, although malignancy was reported in two cases [3, 4]. Gastric teratoma most commonly presents with abdominal mass or abdominal distension [5]. Gastric teratoma has also been reported as a cause of gastrointestinal (GI) bleeding on few occasions [1]. We report two cases of malignant gastric teratomas in infants. First case presented with an abdominal mass and the second one with GI bleeding and severe anemia.

M. Saha
Department of Pediatric Surgery, Gauhati Medical College, Guwahati, India

M. Saha (✉)
D-1, Ashadeep Tower, A.K. Azad Road, Rehabari, Guwahati, Assam 781008, India
e-mail: drmanojsaha@gmail.com; drmanojsaha@yahoo.com

Case reports

Case no. 1

One 45-day-old male child, presented with abdominal distension for last 10 days. There was no history of vomiting and no bowel and bladder symptom. On examination, his general condition was good, but he was irritable. There was a large intra-abdominal mass, predominantly on the left and extending to the right significantly. The mass was firm-to-hard in consistency. The abdominal skiagram showed a large soft tissue mass with areas of calcification. Ultrasonography detected an intra-abdominal mass of variable echogenicity. Axial CT scan (Fig. 1) was done and it showed a large soft tissue mass with septation and calcification in the retrogastric area. His hemoglobin was 10/dl and serum alpha-fetoprotein (AFP) was abnormally raised. Chest X-ray was normal. He was operated on 06.05.2004. There was a mass arising from posterior wall of the stomach, near the greater curvature (Fig. 2a, b). The mass was adherent to the pancreas. The mass was dissected from the pancreas and excised along with a part of the

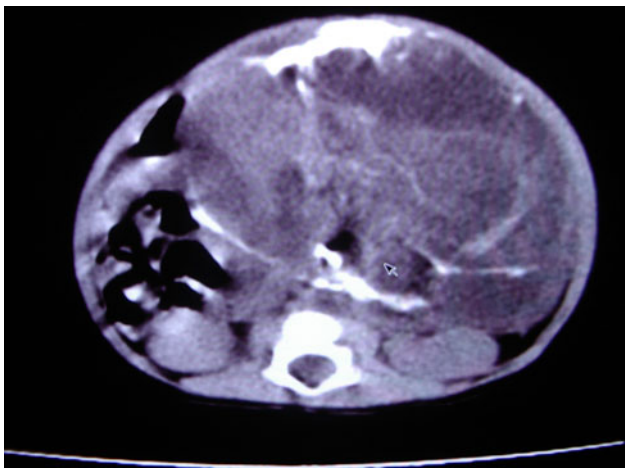

Fig. 1 Axial CT scan showing the tumor

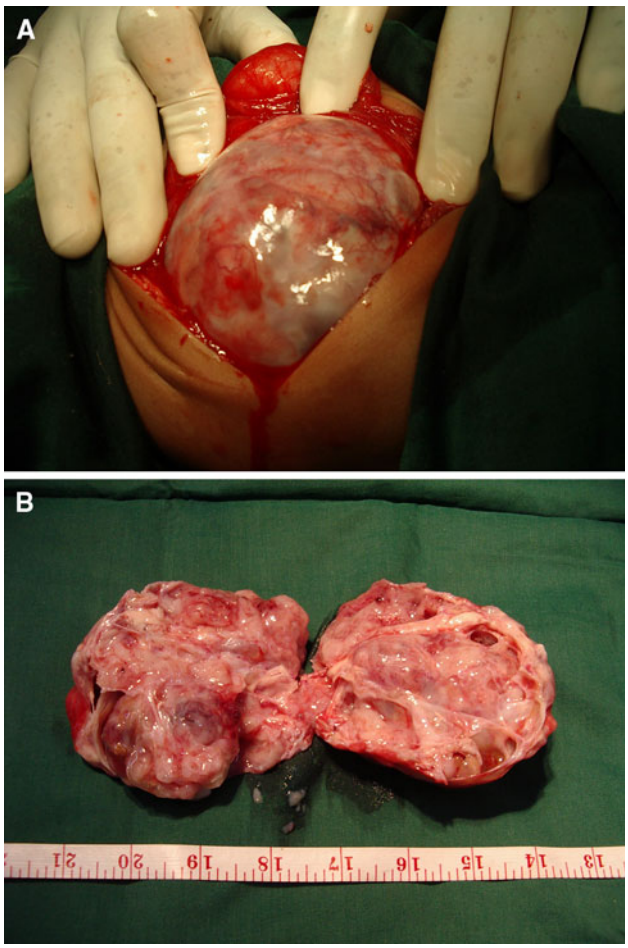

Fig. 2 **a** Tumor mass during operation. **b** Cut section of the tumor specimen

stomach. The stomach was repaired in two layers. The postoperative recovery was uneventful. The patient was discharged on the sixth postoperative day. Histopathology of the tumor showed malignant teratoma in the form of

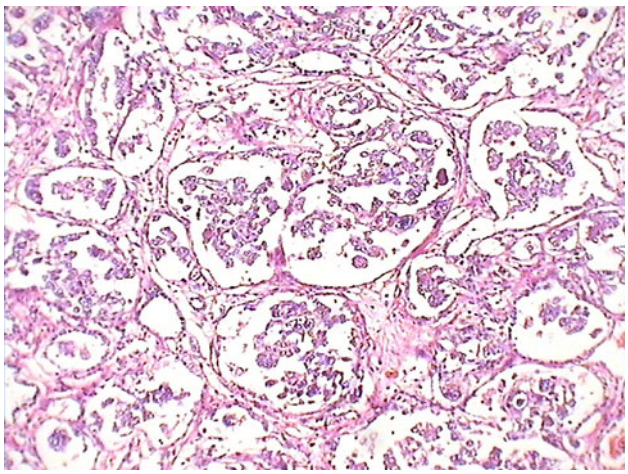

Fig. 3 Histology showing endodermal sinus tumor

endodermal sinus tumor (Fig. 3). In view of elevated AFP level and malignancy on histology, we advised adjuvant chemotherapy but the parents did not agree. The patient turned up after a period of 2 years. He was disease-free and the abdominal ultrasonography and serum AFP level were normal. At present, he is doing well without recurrence.

Case no. 2

A 1-year-old male child presented with passage of black stool for 2 weeks and two episodes of hematemesis at weekly intervals. He was otherwise well since birth. There was no history suggestive of portal hypertension. He was admitted with severe anemia (hemoglobin 4 gm/dl). Other than that, there was no significant finding on clinical examination. There was no clinical sign of portal hypertension. Ultrasonography of the abdomen was normal. Upper GI endoscopy was done and showed sessile gastric polyp at the fundus. Chest skiagram was normal. Anemia was corrected by preoperative blood transfusion. Laparotomy was done on 01 Oct 2005. There was a cauliflower-like growth arising from posterior wall of the stomach occupying fundus and part of the body of the stomach (Fig. 4a, b). The mass was protruding out of the stomach wall and was adherent to the pancreas. The mass was resected with adequate margin distally. Proximally adequate margin could not be achieved as the mass was encroaching near the gastroesophageal junction. The postoperative period was uneventful. He was discharged on the seventh postoperative day. Histopathology of the tumor showed yolk sac tumor (Fig. 5a, b). Perioperative AFP level was 2,200.0 IU/ml (normal 0–10 IU/ml). He received adjuvant chemotherapy with PEB (cis-Platinum, Etoposide, and Bleomycin) regimen. His AFP level came down to normal after three cycles of chemotherapy, and after that he received two more additional cycles. He was followed up with abdominal

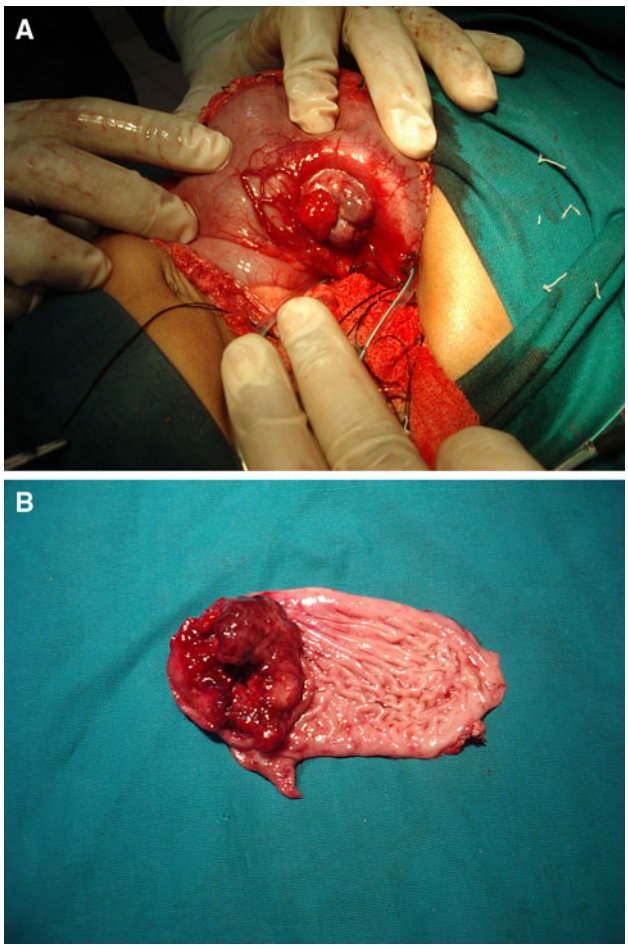

Fig. 4 **a** Tumor arising from the posterior wall of the stomach. **b** Resected tumor specimen with part of the stomach

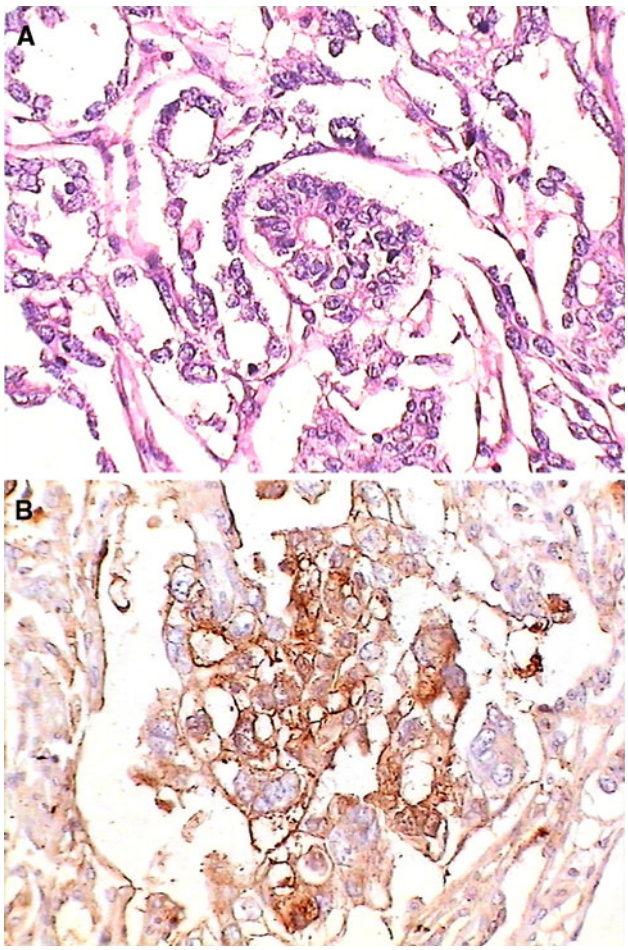

Fig. 5 **a** High power microscopy showing yolk sac tumor with perivascular Schiller Duval bodied (*center*). **b** Special stain showing AFP positive tumor cells

ultrasonography and serum AFP levels at 6-month intervals initially for 2 years and then yearly. Now he is doing well without recurrence.

Discussion

Gastric teratoma is a rare tumor. The first case of GT was reported in 1922 by Eusteman [6]. Gastric teratomas occur most often in males. In 1979, Purvis et al. [7] reported the first case in a female infant. As of date, about 10 cases of gastric teratoma have been reported in females [4, 8]. In 1977, Moriuchi et al. [5], reviewed 44 cases of GT and found that 85% of the cases were below 1 year of age and all the cases were males. In 90% of the cases, the tumor arose from the greater curvature of the stomach and all the cases were benign on histopathology. Only two cases of malignant GT have been reported in the literature [4]. We report two more cases of malignant gastric teratomas after 5 and 6 years of follow-up, respectively. We believe that these two cases are new additions to the world literature.

Gastric teratomas usually occur below 1 year of age. However, there have been reports of this tumor in older children [6]. Akram et al. [9] has reported a case of immature GT diagnosed prenatal sonography in the third trimester of pregnancy. The baby was delivered by cesarean section at 37 weeks gestation. Surgical excision was done on the 10th day of life.

Teratomas are almost always benign. However, 1–2% of the ovarian teratoma and 2–3% of the testicular teratomas [10] undergo malignant transformation. Malignant transformation has also been reported in the mediastinal, gastric, brain, and sacrococcygeal teratomas [11].

Teratomas can be classified into three types according to their histology: mature, immature and malignant. Mature teratoma consists of well-differentiated tissue; immature teratoma has varying degree of immature fetal tissue; the malignant type consists of at least one of the malignant germ cell components [12]. Immature teratomas are also graded (from 1 to 3) by the amount of immature tissue contents, which are mainly neural elements, and by the

degree of mitotic activity [13]. The term 'malignant teratoma' is restricted to the endodermal sinus tumor (EST) or yolk sac tumor and choriocarcinoma.

Alpha-fetoprotein synthesis occurs in fetal liver, yolk sac, and gastrointestinal tract. Preoperatively, an abnormal elevated level of serum AFP can be obtained because of the presence of intestine in the teratoma or due to presence of yolk sac tumor in immature teratomas. Therefore, serum AFP level is very useful as it provides information regarding recurrence or presence of residual tumor [14]. Nevertheless, the initial raised level of AFP does not necessarily indicate malignancy. However, the elevated AFP level may be the only clue of a histologically missed tiny focus of yolk sac component in a 1-kg tumor mass [15]. On the other hand, elevated serum AFP below 8 months of age is also an enigma because of wide physiological variation in young infants [12].

Total excision and primary closure of the gastric wall is the treatment of choice for benign gastric teratomas. The extent of resection is dictated by the area of the stomach involved. Prognosis following surgical excision has been shown to be excellent [14]. In childhood, even the immature gastric teratomas have a better prognosis [12]. Good results have been shown after complete excision of childhood immature teratomas [2, 8, 16, 17]. Some authors have shown good results with only surgical excision in grade 2 and grade 3 immature teratomas, even when serum AFP level was raised [8, 16]. The same approach was shown to be adequate in two cases of immature GT with abdominal metastasis [8]. Marina et al. [18] suggested that it seemed safer to treat all patients with extragonadal immature teratomas by surgical excision followed by close observation, withholding chemotherapy until there was evidence of disease recurrence. Corapcioglu et al. [15] has suggested that after complete surgical excision of grade 2 immature teratoma with raised serum AFP level at presentation, no chemotherapy is needed if postoperative AFP level shows expected decrease. They treated a case of grade 2 immature teratoma with adjuvant chemotherapy in view of postoperative high serum AFP value and unusual gastric localization of the tumor. For malignant teratomas, chemotherapy is recommended following surgical excision [12]. Nevertheless, treatment schedule for malignant GT is not clearly defined. Out of our two cases, we treated one case with postoperative chemotherapy. The parents of the other patient (45-day-old boy) did not agree for chemotherapy. Now both the patients are tumor-free at 5 and 6 years after surgery, respectively. Refusal to accept chemotherapy by the parents of the 45-day-old baby has brought to light the fact that complete surgical excision only may be adequate for malignant GT in small infants. However, as tumor residue and metastasis, which can be responsible for elevated AFP, are undetected by postoperative imaging studies, it is better to administer adjuvant chemotherapy in cases of malignant GT, with dose adjustment for smaller infants. It is better to be safe than sorry.

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