Pediatr Surg Int (2000) 16: 329–332

ORIGINAL ARTICLE

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Gastric teratoma in children

Accepted: 8 November 1999

Abstract Gastric teratoma (GT) comprises less than 1% of all teratomas in children. Though GT in the presence of immature neuroepethelial elements is regarded as malignant, the prognosis is excellent after complete excision of the tumor. Because of its rarity the world literature lacks a large study. Clinical experience with ten cases of GT is presented and discussed. Only one patient was female; the mean age at presentation was 3.2 months. Two cases were immature grade III GT; one of these had infiltrated the left lobe of the liver and the transverse colon while the other had metastasized to the regional lymph nodes and omentum. All the patients underwent complete excision. There were no deaths, and after a mean follow-up period of 4.2 years, all the patients had no recurrence and were healthy. Both the mature and immature types of GT have an excellent prognosis after complete excision of the tumor. Even when the immature type infiltrates surrounding structures, complete excision offers recurrence-free survival without requiring chemo- or radiotherapy.

Key words Teratoma · Stomach · Child

Introduction

Teratomas are embryonal neoplasms that contain tissue from all three germ layers, namely, ectoderm, endoderm, and mesoderm and are derived from totipotent cells. The majority of the patients present during infancy and early childhood and the tumors may be benign or malignant. The nomenclature is somewhat misleading in that immature teratomas are frequently labeled as malignant, but in childhood they do not behave in a really malignant fashion unless they have foci of malignant germ cells or neural elements, a yolk-sac tumor, neuroblastoma, or medulloepithelioma is present [6]. These elements may be overlooked, as the clusters may be small, may be intermingled with immature tissue, or may not stain for alpha-fetoprotein (AFP). To add to this confusion, structures called fetal or embryonic intestine may occasionally be found in immature teratomas and may be related to elevated AFP levels. In an analysis of 85 benign and malignant teratomas in children, Grosfeld et al. reported that 21% of all teratomas in children were malignant [13].

Gastric teratoma (GT) is extremely rare, comprising less than 1% of all teratomas [4]. We culled the reported cases of GT in the world literature using the MEDLINE database from 1966 onward; a total of 102 cases were found. More than 90% of these were in males. Although the literature is replete with occasional case reports of malignant GT [3, 4, 19], there is only one case report of a GT undergoing malignant transformation [17].

The purpose of this review was to present the authors' clinical experience during the period 1987–1999 with ten cases, which to our knowledge is the largest series of GTs from a single center. The importance of immature elements and the response to surgical treatment alone even in cases with infiltration of surrounding structures and lymph-nodes metastases is discussed.

Materials and methods

Ten patients with GT diagnosed and treated at our institute were all born at full term; nine were males. The mean age at presentation was 3.2 months (range 10–180 days). Eight presented with an abdominal mass and two with abdominal distention and vomiting. Ultrasonography (US) revealed a mixed echogenic mass, and calcifications were noted in four cases. Computed tomography (CT) was done in five cases and revealed a solid and cystic multiloculated tumor of mixed echogenicity and calcifications. Serum AFP was determined by immunoenzymetric assay (AFP Serozyme, Biochem

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Immunosystems) and the results were compared with our laboratory nomogram. On comparison with age-related serum titers, AFP levels were grossly elevated in two cases that turned out to be grade III immature GTs, and the level in one case of a mature GT was just above 2 standard deviations.

Surgical exploration revealed the mass localized to the posterior wall of the stomach in six cases and arising from the posterior wall and extending to the lesser curvature and anterior wall as well in two. All except two tumors were well-encapsulated without any gross metastases, and complete excision with a rim of normal stomach completed the treatment. The mean follow-up period was 4.2 years. The details of this series are briefly summarized in Table 1. There were no deaths, and histologic examination showed mature teratomas in eight cases and immature grade III teratomas in two, which are discussed here briefly as case reports.

Case 1

A 6-month-old male presented with an abdominal mass noticed by a pediatrician during a routine check-up. Abdominal examination revealed a firm, irregular mass at the left hypochondrium and epigastric region, the dullness of which to percussion was continuous with that of the liver. The liver was palpable up to 3 cm below the costal margin. US showed a large, multiloculated tumor of mixed echogenicity related to the stomach and liver with solid and cystic areas and calcifications. Serum AFP was 100 ng/ml. Laparotomy confirmed a large, multicystic tumor arising from the lesser curvature of the stomach and extending to the anterior wall, infiltrating the left lobe of the liver and transverse colon. Resection of the tumor along with a left hepatic lobectomy and resection and end-to-end anastomosis of the transverse colon was performed. Histologic examination of the tumor revealed a grade III immature teratoma. Serum AFP was absent 12 weeks postoperatively. At 18 months following the surgery, the child was asymptomatic and doing well.

Case 2

A 3-month-old male presented with abdominal distension and poor weight gain. Physical examination revealed a large mass in the umbilical and right lumbar regions. CT showed a large, multiloculated tumor with calcifications displacing the right kidney and bowel loops. Serum AFP was 1750 ng/ml. At laparotomy, a large tumor was arising from the posterior wall of the stomach. The regional lymph nodes were enlarged. Excision of the tumor (Fig. 1) and repair of the stomach was performed. Lymph nodes were also sent for histologic examination. The specimen was reported as a grade III immature GT with metastases to the lymph nodes. An omental biopsy also revealed metastases. Four months after surgical excision, there was no evidence of recurrence of the tumor. Serum AFP was undetectable.

Discussion

Teratomas are commonly observed in the sacrococcygeal region, gonads, mediastinum, and retroperitoneum. The etiology is not well-elucidated and may be variable at different sites. Totipotent cells are found in close proximity to the primitive streak and Hensen's node in early embryogenesis, and the most widely accepted theory is that these are responsible for teratomas [1]. The site of origin in one of the largest series of benign and malignant teratomas in children was sacrococcygeal in 65%, mediastinal in 10%, gonadal in 11%, and presacral in 4% of cases [13].

GT was first reported by Eusterman and Sentry in 1922 [9]. It constitutes less than 1% of all teratomas in children, and so far 102 cases have been reported in the world literature. GTs are found mostly in male infants [16], and in our series there was only one female. The mean age at presentation in this series was 3.2 months. Presentation is usually as an abdominal mass or distension with or without vomiting. The patient may present with gastrointestinal (GI) bleeding [5, 10, 14], as the intramural component may ulcerate and hemorrhage. Rarely, the child may present with respiratory distress [15].

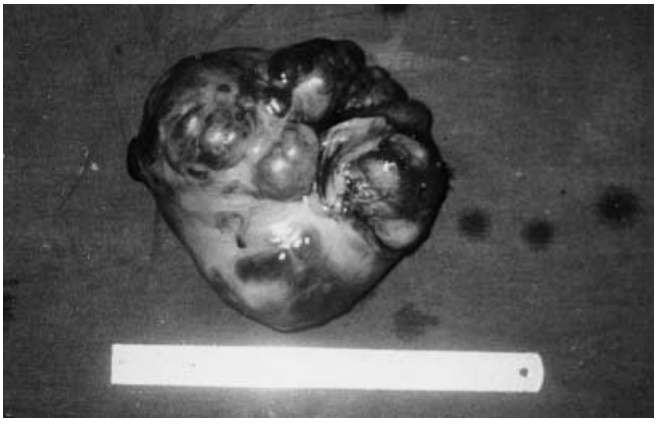

Fig. 1 Specimen of gastric teratoma arising from posterior wall of stomach after complete excision

Table 1 Summary of cases comprising this series (*Calc* calcification, *HPE* histopathological examination, *MT* mature teratoma, *IMT* immature teratoma, *PW* posterior wall of stomach, *LC* lesser curvature of stomach, *AFP* alpha-fetoprotein, *Gr* grade, *SD* standard deviation, *Mets* metastases)

Patient no.	Age (days)	Cardinal sign	Calc	Site (predominant)	HPE	AFP (ng/ml)
1	58	Mass	–	PW	MT	358 (normal)
2	110	Mass	–	PW	MT	288 (>2SD)
3	10	Mass	+	LC	MT	2316 (normal)
4	112	Mass	–	PW	MT	84 (normal)
5	96	Distension	–	PW	MT	76 (normal)
6	92	Mass	–	LC	MT	84 (normal)
7	180	Mass	+	LC, liver, transverse colon	Gr III IMT	100 (>2SD)
8	94	Mass	+	PW	MT	114 (normal)
9	90	Distension	+	PW, Enlarged lymph nodes	Gr III IMT, Mets+	1750(>2SD)
10	96	Mass	–	PW	MT	122 (normal)

The mass is palpable in most cases and may produce intestinal obstruction, so that the child may present with dehydration as a result of vomiting. Large tumors may produce premature labor and dystocia [16]. GTs may also present after spontaneous rupture [2]. Roentgenography of the abdomen shows a large soft-tissue mass with irregular calcifications in 50% of cases [18]. The presence of teeth or bone is pathognomonic for teratomas, but such features are less frequently present in GT. The preoperative diagnosis is made by US, which demonstrates solid and cystic areas. Areas of calcification may also be apparent. CT or magnetic resonance imaging (MRI) confirms the diagnosis, delineates the mass, and clarifies its extension to surrounding structures.

AFP synthesis occurs in the fetal liver, yolk sac, and GI tract [12]. AFP is an excellent indicator of the presence of a malignant tumor. In a large series of different teratomas in children, Tsuchida et al. reported that AFP was positive in 25 of 33 patients with malignant and 9 of 45 with benign teratomas [22]. All the latter instances occurred in children less than 1 month of age. Although AFP may not be a good predictor of malignancy, it has a definite role in follow-up, as persistently raised or increasing titers may point to recurrence or residual tumor. The differential diagnosis includes neuroblastoma; abdominal roentgenography here may show calcifications, but CT or MRI will reveal the mass arising from the adrenal gland. Solid tumors like hepatoblastoma and mesoblastic nephroma also must be considered. If the tumor has both cystic and solid components, and if CT/MR imaging reveal fat and calcifications, it is likely to be a teratoma.

Grossly, GTs have both cystic and solid components. The tumor is commonly exogastric. A popular classification devides teratomas into mature forms, which comprise the majority of cases and contains mature tissues; immature forms, which contain immature elements such as yolk-sac tumor, germinoma, and embryonal carcinoma; and malignant forms, which have definite evidence of malignancy. The degree of immaturity was graded conventionally by Thurlback and Scully [21], and this scheme was modified by Dehner [8].With the presence of foci of endodermal-sinus tumor, ideally it should be diagnosed as a malignant mixed germ-cell tumor.

Complete surgical excision is curative, and no cases of recurrence have been reported so far. One of our patients had a large mass with extension into the left lobe of the liver and adherence to the transverse colon. A left hepatic lobectomy, resection of the tumor, and end-to-end anastomosis was performed followed by close follow-up. Giacomono and Zagaroni [11] excised a GT extramucosally, leaving behind the intact gastric mucosa, but the report of malignant transformation of a GT by Matsukama et al. [17] should prompt complete resection and regular follow-up after surgical treatment. Chemo- or radiotherapy is not recommended even in cases of immature teratoma.

Immature GTs have unique features and may contain neuroepithelium. Regular follow-up is needed. Unlike sacrococcygeal teratomas, which have a high incidence of recurrence and malignant transformation [20], GT has no propensity to become malignant and has an excellent prognosis. Coulson [7] reported peritoneal gliomatosis 10 months after excision of a GT while operating upon the same child for an inguinal hernia. However, without any treatment the child remained asymptomatic. There is no evidence that GT has a higher incidence in this country. The series reported here is the largest to date, which could be due to the fact that this institute is a tertiary referral center serving to a large population base.

In summary, GT is a very rare teratoma that commonly presents during infancy and early childhood with a very high male predominance. Although in the presence of immature neuroepithelial elements GT is labeled as malignant, the prognosis is excellent. Even when a malignant GT infiltrates the surrounding structures, complete excision offers recurrence-free survival.

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