Case Report

Immature Gastric Teratoma of Childhood: A Case Report and Review of The Literature

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INRODUCTION

Teratomas are infrequent germ cell tumors of childhood. They may arise from any organ but the majority are found in the ovary, testis, sacrococcygeal region and mediastinum. Gastric teratoma is rare, accounting for less than 1% of all teratomas (1–3). Immature teratomas are pathologically distinct from the benign and malignant teratomas with their embryonic-appearing neuroglial and neuroepithelial elements. AFP and βHCG may not increase if there are no malignant germ cell components (4,5). The diagnostic utility of these oncofetoproteins is less in young children because of the elevated physiologic serum levels (6,7). We report a 5-month-old male with immature gastric teratoma who had a complete surgical resection and adjuvant chemotherapy. The need for adjuvant chemotherapy in this case is discussed.

CASE REPORT

A 5-month-old male was admitted to our center with abdominal distention. He was the product of an uneventful pregnancy, labor and vaginal delivery. The boy was healthy but his mother had noticed abdominal distention for 2 months. Physical examination revealed a large, irregular mass at the left hypochondrium and epigastric region. There were no other abnormalities. Complete blood count showed hemoglobin 11.7 g/dl, white blood cell 14200/μl and platelets 476,000/μl. Serum biochemical profiles were normal. Ultrasound and computed tomographic (CT) scanning showed a multiloculated tumor (11 × 11 × 9 cm) in the upper left abdominal cavity and epigastric region with cystic areas and calcification (Fig. 1). Serum AFP was 189 ng/ml (normal range for this age, 46.5 ± 19.0 ng/ml) (8) and other tumor markers were as follows: βHCG 0.5 mIU/ml (normal level <5.3 mIU/ml), neuron specific enolase 9 μg/l (normal level <12.5 (μg/l) and urine vanillylmandelic acid 0.2 mg/24 h (normal level <2 mg/24 h), all within normal limits. Thoracic CT showed no lung metastases. Laparotomy was performed 15 days after CT scan. An extragastric tumor (15 × 15 cm) arising from the lesser curvature of the stomach was completely excised with a small part of the lesser curvature, and the defect in the stomach was repaired. Macroscopically all tumor tissue was removed in one piece and enlarged lymph nodes were not observed and no lymph nodes were resected. Histologic examination revealed grade 2 immature teratoma with tumor cell free margins. Although, preoperative measurement of AFP was elevated, there was no microfocus of yolk sac tumor on the histologic slides. AFP level was 59 ng/ml on the fifth postoperative day. The postoperative period was uneventful but as a result of a fluctuation in serum AFP level (106 ng/ml on the 15th postoperative day) and grade II immature histology, we administered two courses of adjuvant chemotherapy every 3 weeks with modified BEP to avoid pulmonary toxicity (cisplatin: 20 mg/m²/d, 1–5 days and VP-16: 100 mg/m²/d, 1–5 days; bleomycin was omitted). Thus his chemotherapy was completed and follow-up with AFP level and imaging was planned. He was lost to follow-up for 45 days, and on readmission to our Center, we were informed that he had two more courses of chemotherapy every 3 weeks in another Pediatric Oncology Unit with cisplatinum, VP-16 and bleomycin (cisplatin: 20 mg/m²/d, 1–5 days and VP-16: 100 mg/m²/d, 1–5 days, bleomycin: 15 mg/m²/d, 1 day). On the 90th postoperative day after the fourth chemotherapy course serum AFP level was 10.4 ng/ml and remained in normal range with no recurrence of teratoma in the postoperative 15 months. During modified BEP administration and in the follow-up period no severe myelotoxicity or nephrotoxicity, mucositis, ototoxicity, pulmonary toxicity or nutrition disorder was observed. After the first modified BEP course the patient had red blood cell transfusion. Absolute neutrophile count decreased to 500/μL but he experienced no febrile
episode. Platelet count decreased to 123,000/µL and he never required platelet transfusion. We are not aware of the myelotoxicity experienced during BEP administration in the other center.

**DISCUSSION**

Gastric teratomas are rare in childhood and are found mostly in male infants. There are only nine reported female infants in the English literature (2,3,9–11). Gastric teratomas most commonly arise from the greater curvature and posterior wall of the stomach (2,9,12,13). Utsch et al (14) reported a 5-month-old male infant who presented with gastric teratoma originating from the lesser curvature and Moriuchi et al (15) reported a 3-month-old male infant showing endogastric tumor located on the lesser curvature. To our knowledge, this is the third report of immature gastric teratoma in a boy involving the lesser curvature. Typical presenting symptoms are abdominal mass, distention and vomiting, but tumors with intramural extension causing gastrointestinal bleeding and gastric perforation have also been reported (1,3,14,16,17). The presented tumor had no endogastric growth pattern.

Teratomas are embryonal neoplasms composed of derivatives of all three germinal cell layers (3,14). They can be classified into three types according to their histologic composition. Mature teratoma consists of well-differentiated tissue, immature teratoma has varying degrees of immature fetal tissues and the malignant type contains at least one of the malignant germ cell elements (7). 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 (18).

As a general approach, complete surgical excision is sufficient with grade 1 immature teratoma if AFP and βHCG values are within normal age-related ranges and there are no malignant germ cell elements (2,4,19,20). Incomplete tumor resection, coccygeal or ovarian site and immaturity are known risk factors for both malignancy and relapse (19). Although adjuvant therapies (i.e., chemotherapy or radiotherapy) are not recommended with completely resected grade 2 and 3 immature teratomas, optimal treatment in these groups is still controversial (3,7,21,22). Chemotherapy is recommended for patients with grade 2 and 3 ovarian immature teratomas after surgical resection (7). Marina et al. (21) suggested that it seems safe to treat all patients with extragonadal immature teratomas by surgical excision followed by close observation, withholding chemotherapy until there is evidence of disease recurrence. On the other hand, there are limited reports regarding the relationship be-

**TABLE 1. Outcomes in immature gastric teratoma**

<table>
<thead>
<tr>
<th>Ref. no</th>
<th>Authors/year</th>
<th>Pt number</th>
<th>AFP levels</th>
<th>Histologic grade</th>
<th>Treatment</th>
<th>Follow-up duration</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1*</td>
<td>Wakhlu A, et al. 2002</td>
<td>1</td>
<td>?</td>
<td>?</td>
<td>Complete excision</td>
<td>Median 3 years†</td>
<td>No recurrence</td>
</tr>
<tr>
<td>3‡</td>
<td>Gupta DK, et al. 2000</td>
<td>2</td>
<td>100 and 1750 ng/ml</td>
<td>3</td>
<td>Complete excision</td>
<td>4 and 18 months</td>
<td>No recurrence</td>
</tr>
<tr>
<td>14</td>
<td>Utsch B, et al. 2001</td>
<td>1</td>
<td>697 ng/ml</td>
<td>2</td>
<td>Complete excision</td>
<td>16 months</td>
<td>No recurrence</td>
</tr>
<tr>
<td>17</td>
<td>Park WH, et al. 2002</td>
<td>1</td>
<td>33,456 ng/ml</td>
<td>2</td>
<td>Complete excision</td>
<td>30 months</td>
<td>No recurrence</td>
</tr>
</tbody>
</table>

*Ref 1: Total 7 gastric teratoma, only 1 immature teratoma; †median follow-up of 7 cases; ‡Ref 3: Total 10 gastric teratoma, only 2 immature teratoma, one patient with liver and transverse colon infiltration, second case with regional lymph node and omentum infiltration at diagnosis.
tween histologic grade and outcome in patients with gastric immature teratomas.

The term “malignant teratoma” should be restricted to embryonal carcinomas, yolk sac tumors or choriocarcinomas (4,14,22). Because clusters of yolk sac tumor may be very small or intimately associated with the immature neural tissue, they are easily overlooked (7). Elevated serum AFP levels may be the only alerting sign of the presence of malignant yolk sac component. Preoperative assessment of the tumor markers helps to individualize treatment planning (4,7). Evaluation of serum AFP levels in infants, especially in those younger than 8 months, is an enigma because of wide physiologic variation in infants, especially in those younger than 8 months, is also an enigma because of wide physiologic variation (7,8). MEDLINE from 1970 to 2003 contains case reports of immature gastric teratoma with good outcome after complete surgical excision in childhood (1,3,9,13,14,17,23–26) (Table 1). Although we could not obtain histologic grades and AFP levels for the majority of the cases, good outcomes with complete excision are reported by some authors in grade 2 and 3 immature gastric teratoma even with elevated AFP levels (3,14,17). The same therapeutic approach was suggested to be adequate even in two cases with abdominal metastasis (3). However, the follow-up of one of these patients was only 4 months. We were discouraged with unusual gastric localization and postoperative increase in serum AFP level and administered chemotherapy.

It seems that the grading system is more useful in evaluating immature teratomas in adolescents and adults. In childhood, immature teratomas have a better prognosis (7). Although the value of AFP cannot be denied in the diagnosis and follow-up of germ cell tumors, in immature gastric teratoma it does not seem predictive for malignant potential. More case reports with well-defined histologic grading and AFP values will clarify the controversy in the therapeutic approach.

In conclusion, elevated AFP levels may be the only clue of a histologically missed tiny focus of yolk sac component in a 1-kg tumor mass. In completely excised grade II immature gastric teratoma of infants with elevated AFP at presentation, no chemotherapy is indicated if postoperative AFP values show the expected decrease. Thus, a “wait and watch” strategy is justified for these tumors after surgery. We suggest close follow-up and adjuvant chemotherapy for patients with AFP elevation after surgery. Although tumor residue and metastasis, which can be responsible for the AFP elevation, are not detected in postoperative imaging studies, we prefer to be safe rather than sorry.

REFERENCES