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CASE REPORT

Immature gastric teratoma in an infant: a case report and review of the literatures

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Key Clinical Message

Immature gastric teratoma is an uncommon germ cell tumor of the stomach. We report a rare case of immature gastric teratoma in an infant with down syndrome with clinically presenting with hematemesis and severe anemia. Complete surgical resection remains the cornerstone of treatment.

Keywords

Down syndrome, extramedullary hematopoiesis, immature teratoma, stomach

Introduction

Teratoma is defined as germ cell tumor composed of tissues derived from ectoderm, endoderm, and mesoderm and has been described in various locations, including the gonad, intracranium, anterior mediastinum, retroperitoneum, and sacrococcygeal region. The alimentary tract accounts for <1% of all teratomas [1]. Gastric teratoma was first reported by Eusterman and Sentry in 1922 [2]. Gastric teratomas are considered to be benign nature; however, immature teratomas appear to be more aggressive and have malignant nature. To date, 30 cases of immature teratoma have been reported in the literature [1, 3–15]. Herein, we report on the clinicopathologic findings of an infant with down syndrome who presented with hematemesis and severe anemia. Finally, he was diagnosed with immature gastric teratoma.

Case Report

An 8-month-old patient presented with hematemesis and anemia of one-month duration. He had an underlying disease of down syndrome and congenital hypothyroidism. Physical examination of abdomen showed an enlarged, intra-abdominal mass, predominantly in the left upper quadrant of abdomen. Relevant laboratory data included a hemoglobin of 2.4 g/dL, hematocrit 9.8%, and white blood cell count of 14,220 cells/mm^3. Anti-HIV was nonreactive by enzyme-linked immunosorbent assay (ELISA). Plain abdominal radiograph showed a soft tissue density in the left upper quadrant of abdomen. Endoscopic gastroduodenoscope was performed and revealed a large intragastric soft tissue mass with ulcer (Fig. 1A and B). An incisional punch biopsy of the mass was performed. He underwent a single-phase venous scan computed tomography (CT) of the whole abdomen that revealed an endophytic heterogeneous hyperattenuating soft tissue mass measuring 7.4 × 6.9 × 4.9 cm, locating in the stomach and protruding from the lesser curvature. Small areas of punctuated calcification and a small focal area of macroscopic fat were also observed. The incisional biopsy showed immature myeloid cells infiltration, compatible with granulocytic sarcoma. The bone marrow

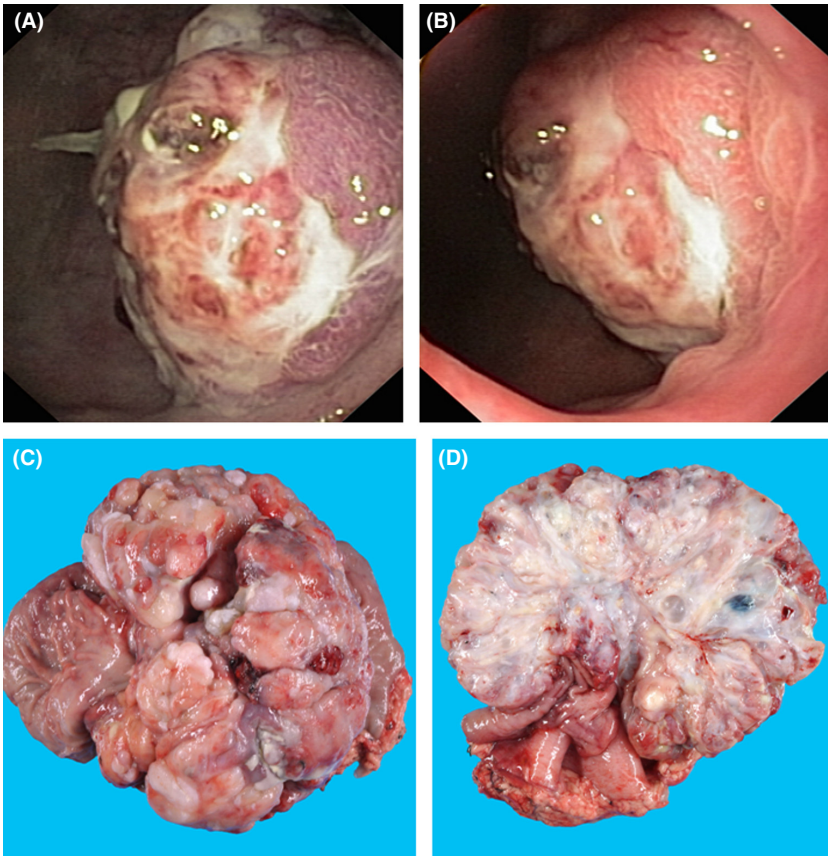

Figure 1. The gastroscope shows an endophytic soft tissue mass locating within the stomach (A, B). The gross section shows an endophytic well-circumscribed rubbery firm red-brown mass measuring 13 × 11 × 6.5 cm, originating from the lesser curvature (C). The cut surfaces of mass revealed a solid-cystic and gelatinous appearance with focal cartilaginous and pigmented areas (D).

biopsy showed active trilineage hematopoiesis without evidence of malignancy. He received two cycles of chemotherapy including cytosine arabinosine 2.5 mg/kg/day for 7 days and idarubicin 0.25 mg/kg/day for 3 days. The gastric mass was progressively enlarged. Abdominal CT revealed a huge heterogenous enlarge gastric mass measuring 11.8 × 10.5 × 4.7 cm. (Fig. 2). The mass increased in size with internal calcification and

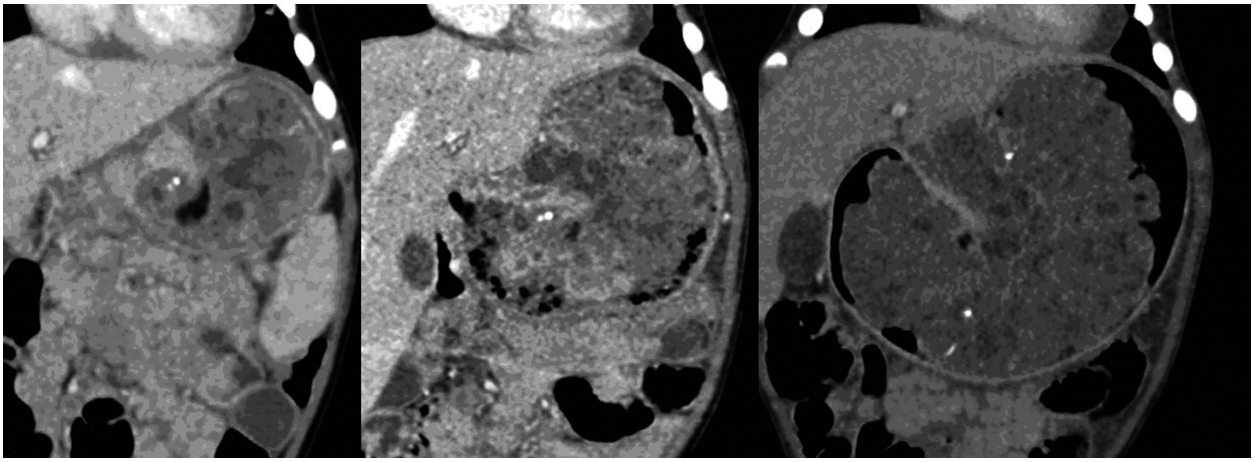

Figure 2. Coronal CT scan of the whole abdomen before, during, and after chemotherapy shows markedly increased size of the mass within 2 months with increased internal calcification and fat components.

intratumoral fat component. In addition to an increase in size, he developed upper gastrointestinal bleeding which required blood transfusion. Subsequently, he underwent near-total gastrectomy. Laboratory investigation on admission showed increased serum alpha-fetoprotein (50 ng/mL; reference 0–7.02 ng/mL). The final pathologic diagnosis was immature teratoma, grade I. The AFP level returned to a normal range after complete surgical resection. At the 2 years of follow-up, he remains well and exhibits no evidence of recurrence and systemic metastasis. He has been advised routinely follow-up.

Pathological findings

The resected stomach measuring 15 × 11 × 7 cm showed an endophytic well-circumscribed rubbery firm red-brown mass measuring 13 × 11 × 6.5 cm, originating from the lesser curvature (Fig. 1C). The cut surfaces of mass revealed a solid-cystic appearance with focal cartilaginous, gelatinous, and pigmented areas (Fig. 1D). The histopathology revealed various types of tissues including skin, respiratory epithelium, adipose, cartilage, bone, muscle, brain, uvea, choroid plexus, and focal immature germ cell component including neural tube and immature cartilage. Focal extramedullary hematopoiesis was observed (Fig. 3). The tumor invaded mucosa and submucosa without involvement of muscularis propria. Angiolymphatic invasion was not detected. The tumor was completely excised. The final pathologic diagnosis was immature gastric teratoma, grade I Table 1.

Discussion

Teratoma originates from the precursor totipotential stem cells and is the most common germ cell tumor in children. It can be either gonadal or extragonadal tissue in origin. The extragonadol teratoma is usually found in younger children, where as the gonadal tumor is often diagnosed in the older ones [2]. The sites of extragonadol teratoma are sacrococcygeal (60–65%), mediastinal (5–10%), sacral (5%), and rarely intracranial, retroperitoneal, cervical, and alimentary [8]. Gastric teratoma is uncommon, contributes less than 1% among teratoma in pediatric patients [1]. Moreover, immature gastric teratoma is relatively rare. There are thirty reported cases in the literature [1, 3–15]. The age at presentation occurs principally during infants and young children. The reported ages of patients range from birth to children with 4-year-old with a mean age of 4.1-month-old [1, 3–15]. One-fifth of cases have been described at birth [1, 5, 6, 13]. Immature gastric teratoma occurs mostly in boys; only two cases have been reported in girls (6.7%) [1, 15]. The tumor size ranges from 4 to 23 cm with a mean and median size of 12 cm in greatest dimension [1, 3–15]. The most frequent clinical presenting symptoms are abdominal distension, palpable mass, and vomiting [1, 3–15]. Moreover, upper

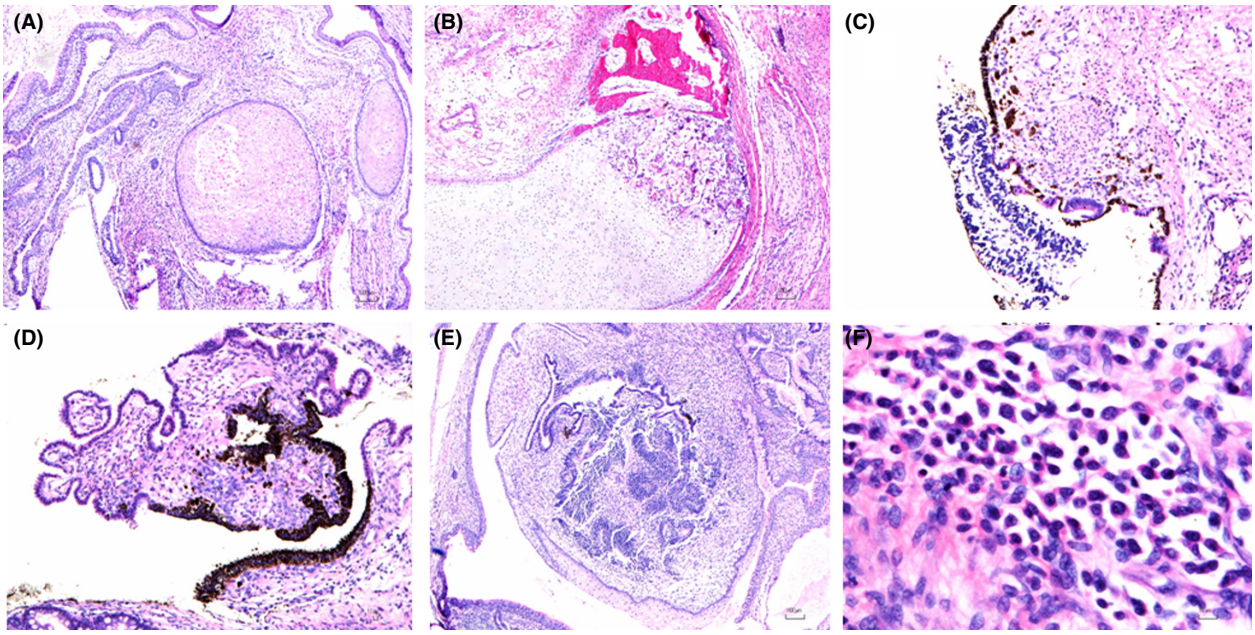

Figure 3. The histopathology shows various types of tissues including skin, respiratory epithelium (A), adipose, cartilage, bone (B), muscle, brain, uvea (C), choroid plexus (D), and focal immature germ cell component including neural tube (E) and immature cartilage. Focal extramedullary hematopoiesis is observed (F).

Table 1. Clinicopathological data for 31 pediatric patients with immature gastric teratoma.

Reference	Year	Age	Sex	AFP level (ng/mL)	Tumor size (cm)	Location	Histologic grade	Treatment	Follow-up duration (months)	Outcomes
Falik-Borenstein et al. [1[a]]	1991	Congenital	M	NA	9.5	NA	2	Complete excision	4	No recurrence
Muñoz et al. [1[a]]	1992	45 days	M	NA	12	Anterior gastric wall	1 at least	Complete excision	96	No recurrence
Gengler et al. [1[a]]	1995	1 month	F	NA	9	Posterior gastric wall	1 at least	Complete excision	NA	No recurrence
Sarin et al. [1[a]]	1997	45 days	M	NA	10	Greater curvature	1 at least	Complete excision	5	Recurrence and death
Ratan et al. [1[a]]	1999	6 months	M	Normal	18	Posterior gastric wall	3	Complete excision	6	No recurrence
Chandrasekharam et al. [1[a]]	2000	5 months	M	NA	NA	NA	1 at least	Complete excision	12	No recurrence
Gupta et al. [1[a]]	2000	6 months	M	100	Large	Lesser curvature, liver, transverse colon	3	Complete excision	18	No recurrence
		3 months	M	1750	Large	Posterior gastric wall, regional lymph node	3	Complete excision	4	No recurrence
Yoon et al. [1[a]]	2000	3 months	M	NA	14	Greater curvature	1 at least	Complete excision	3	No recurrence
Utsch et al. [1[a]]	2001	5 months	M	697	12	Lesser curvature	2	Complete excision	16	No recurrence
Park et al. [1[a]]	2002	Congenital	M	33,456	10	Greater curvature	2	Complete excision	30	No recurrence
Wakhlu et al. [1[a]]	2002	4 years	M	Normal	Massive	Posterior gastric wall	1 at least	Complete excision	24 at least	No recurrence
Hook et al. [1[a]]	2003	25 days	M	Normal	9.5	Posterior gastric wall	3	Complete excision	6	No recurrence
Saleem et al. [1[a]]	2003	Congenital	M	NA	Massive	NA	1 at least	Complete excision	NA	NA
Corapçioglu ;et al. [1[a]]	2004	5 months	M	189	15	Lesser curvature	2	Complete excision and chemotherapy	15	No recurrence
Ukiyama et al. [1[a]]	2005	4 days	M	80,050	5.5	Lesser curvature	1 at least	Incomplete excision	24	Recurrence
Bhat et al. [1[a]]	2007	7 months	M	154	8.5	Greater curvature	2–3	Complete excision	NA	NA
Yadav et al. [4]	2007	NA	NA	NA	NA	NA	NA	NA	NA	NA
Herman et al. [5]	2008	Congenital	M	47.4	13	Lesser curvature	1 at least	Complete excision	NA	NA
Akram et al. [6]	2009	Congenital	M	255,496	10	Posterior gastric wall	3	Complete excision	9	No recurrence
Bhattacharya et al. [7]	2010	2 days	M	NA	8	Anterior gastric wall	2 at least	Complete excision	24	Recurrence with GP and hepatic metastasis
Mohta et al. [8]	2010	20 days	M	690	6.6	Anterior gastric wall	1 at least	Complete excision and chemotherapy	6	No recurrence
Sharif et al. [9]	2010	45 days	M	110	Huge	Posterior gastric wall	3	Complete excision	6	No recurrence
Sharma et al. [1]	2010	5 months	M	NA	15	Posterior gastric wall	3	Complete excision	NA	NA
Valenzuela-Ramos et al. [10]	2010	6 months	M	Normal	4	Lesser curvature	1 at least	Complete excision	36	No recurrence
Yeo et al. [11]	2010	14 days	M	352	12	Greater curvature	3	Complete excision	7	Recurrence with GP
Singh et al. [12]	2011	4 months	M	Normal	23	Lesser curvature	1	Complete excision	12	No recurrence
Jeong et al. [13]	2012	Congenital	M	>60,500	15.5	Posterior gastric wall	3	Complete excision	0.5	No recurrence
Anilkumar et al. [14]	2013	3 months	M	Normal	15	Posterior gastric wall	3	Complete excision	6	No recurrence
Kumar et al. [15]	2013	2 months	F	54,000	20	Posterior gastric wall	3	Complete excision	NA	NA
Junhasavasdikul et al.	Presented case	8 months	M	50	13	Lesser curvature	1	Complete excision	12	No recurrence

[a]Original reference cited in reference.

AFP, alpha-fetoprotein; M, male; F, female; NA, not available; GP, gliomatosis peritonei.

gastrointestinal bleeding has been reported [1]. Gastric teratoma can also cause respiratory distress due to a pressure effect to the diaphragm [13]. The immature gastric teratomas more often originate from the posterior wall and greater curvature of the stomach [1, 3–15] and can be exogastric and endophytic growth in 58–70% and 30%, respectively [12]. Endoscopy and imaging procedures such as radiography, ultrasonogram, and CT may allow early recognition of gastric teratoma.

Radiographic evaluation of gastric teratomas can be differentiated from other common abdominal masses by the presence of associated calcification about 40–60% of all gastric teratomas [12]. The differential diagnoses of plain abdominal radiograph with the left upper quadrant soft tissue mass containing internal calcification include mesoblastic nephroma, nephroblastoma (Wilm tumor), neuroblastoma, ganglioneuroblastoma, ganglioneuroma, and teratoma [12].

Ultrasonography can reveal an internal content of the mass which shows heterogeneous echogenicity, mixed solid-cystic component, and internal calcification [6], but the origin of the mass, especially in the huge one, is hardly demonstrable. Nevertheless, a normal kidney on the ultrasonographic finding can exclude the primary renal tumor.

Computed tomography is more useful in demonstrating the component of mass, its intragastric location, and its extension. Both teratoma and neuroblastic tumor may contain solid and cystic components as well as internal calcification [6, 12]. In our case, however, the presence of gastric invasion and internal fat component favor gastric teratoma while these features are rarely presented in neuroblastic tumor. In conclusion, radiographic findings of the intragastric mass with internal fat component and calcification suggest the diagnosis of the gastric teratoma.

Gastric tumor is an uncommon neoplasm in pediatric patients. Endoscopic evaluation and gastric tissue biopsy must be performed. The more common tumor-mimic lesions including foreign body and bezoars must be initially excluded. The differential diagnoses of gastric tumor include juvenile polyp, hematologic malignancy, gastrointestinal stromal tumor (GIST), smooth muscle tumor, inflammatory myofibroblastic tumor, and teratoma [1].In our case, the gastric punch biopsy of teratomatous components yielded brown to dark-brown tissue, and histological examination showed that most cellular components were immature myeloid cells, compatible with granulocytic sarcoma. The following gastrectomy specimen showed immature teratoma with extramedullary hematopoiesis. A possible reason for the misdiagnosis in gastric punch biopsy specimen is the interpretation of immature myeloid cells to granulocytic sarcoma, which is found in the area of extramedullary hematopoiesis of immature gastric teratoma. The granulocytic sarcoma made up of immature myeloid cells histologically indistinguishable from that occurring in the extramedullary hematopoiesis. Moreover, individuals with down syndrome have an increased predisposition to acute leukemia, predominantly myeloid type including granulocytic sarcoma. Extramedullary hematopoiesis can be misinterpreted as representing a pathologic or neoplastic process. Besides awareness and purely histologic criteria, a false-positive identification of immature hematopoietic cells as granulocytic sarcoma may be avoided by the use of immunohistochemical stains for the maturing hematopoietic cells including myeloperoxidase and lysozyme for the granulocytic line, hemoglobin A and glycophorin A for the erythroid line, CD41, CD61, and factor VIII for the megakaryocytic line, which are highlight the extramedullary hematopoietic cells.

Complete surgical resection remains the cornerstone of treatment of gastric teratoma. Immature gastric teratoma has an excellent prognosis after a complete surgical resection. Adjuvant chemotherapy or radiotherapy is not recommended. Follow-up consists of regular observation and serum AFP measurement to monitor for recurrence or malignant transformation. In case with rising AFP level after surgical resection of gastric teratoma, chemotherapy is recommended. Some authors suggest aggressive postoperative chemotherapy to prevent local recurrence, if there is histopathologic evidence of grade III immature teratoma or malignancy demands including nephroblastic elements. However, the role of chemotherapy in immature gastric teratoma is still not explicitly clear, because of the rarity of cases.

Therefore, it is noteworthy to keep gastric teratoma in mind when dealing with mass lesion in the stomach. The biopsy of teratoma can reveal extramedullary hematopoiesis that may simulate hematologic malignancy, and the context of the specific radiologic feature, and index of suspicion should be maintained, tumor marker obtained, and repeat biopsies performed before committing to intensive chemotherapy. Early diagnosis and prompt medical treatment with careful follow-up are essential. Further genetic and molecular investigation is needed to provide pathogenesis of immature gastric teratoma.

Conflict of Interest

The authors declared that there is no conflict of interest.

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