Rare disease

Immature extragastric teratoma of infancy: a rare tumour with review of the literature

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Summary

A full term otherwise healthy, 4-months-old male infant presented with progressive distension of abdomen from 2 months. The clinical examination showed shifting dullness only but no definite lump palpable. The abdominal radiography revealed calcification in right hypochondrium. Serum α -feto protein (AFP), neuron specific enolase, β human chorionic gonadotrophin and urinary vanillylmandelic acid (VMA) were appropriate for age-range. Contrast enhanced abdominal CT showed predominantly multicystic lesion in right hypochondrium with central solid component and calcification, but no definite organ of origin determined. The exploratory laparotomy showed extra gastric multilobulated cystic mass sized $23 \times 15 \times 8$ cm lesion arising from the lesser curvature of body of stomach. The tumour was feeding through short pedicle based on left gastric artery. There was no infiltration to adjacent areas. Histopathology of excised specimen showed immature teratoma. The child was discharged with appropriate advice and had no recurrence in 1 year of follow-up.

BACKGROUND

Gastric teratoma (GT) is a very rare tumour among infantile teratomas. We here report this rare tumour which had unusual clinical presentation of huge abdominal distension due to fluid. The case supports the dictum, that every infant if had abdominal distension due to fluid should always be suspected to have cystic abdominal tumour teratoma/neuroblastoma in the differential diagnosis, because of high prevalence of these tumours in this age group. A simple abdominal radiogram adds much clue to the diagnosis, as calcification is usually present in more than 50% of cases of teratoma and neuroblastoma.^{1 2} The infants should investigate for the same, as earlier surgical intervention can markedly change the prognosis. The delay in the treatment increases the risk of rupture and malignant transformation of tumour.

CASE PRESENTATION

A 4-month-old male was admitted to our department with painless progressive abdominal distension since 2 months noticed by the mother. The infant was otherwise healthy, having adequate weight for age. There were no other complaints of intestinal obstruction, gastrointestinal haemorrhage, fever or decreased urine output, etc. He was the product of an uneventful pregnancy, labour and vaginal delivery. Physical examination revealed huge abdominal distension with fluid thrill, but no clinical sign of definitive lump.

INVESTIGATIONS

Serum α -feto protein (AFP) was within the range of age limits. Other tumour markers as human chorionic gonadotrophin (HCG) 0.5 mIU/ml (normal level <5.3 mIU/ml), neuron specific enolase (NSE) 10.9 g/l (normal level <12.5 g/l) and urine vanillylmandelic acid 1.2 mg/24 h (normal level <2 mg/24 h), were within normal limits.

Ultrasonography (US) revealed a multicystic swelling occupying whole of the abdomen with bowel displaced inferiorly probably mesenteric cyst. Abdominal radiography showed calcification in right hypochondrium (figure 1). 64-slice contrast enhanced abdominal CT scan showed a predominantly cystic lesion sized $20 \times 12 \times 6$ cm present in right hypochondrium, epigastric, periumbilical region with central solid component and calcification. The tumour had well defined margin and was feeding through a vascular pedicle originating through gastric or falciform ligament (figure 1). No evidence of metastases or any other anomaly.

DIFFERENTIAL DIAGNOSIS

The preoperative clinical diagnoses more common are encysted ascitis, cystic neuroblastoma, pancreatic cyst, omental cyst, mesenteric cyst, cystic abdominal lymphangioma and cystic teratoid tumour.¹

The differential diagnoses of a calcified mass in the left upper quadrant in infant are mesoblastic nephroma, congenital neuroblastoma, teratoma, primary calcifying extra peritoneal neoplasms like malignant fibrous histiocytoma, malignant mesenchymoma leiomyosarcoma, liposarcoma, osteosarcoma and rarely hemangiopericytoma.²

TREATMENT

The exploratory laparoscopy was performed under general anaesthesia keeping in view the principles of tumour surgery. An extra gastric tumour sized $23 \times 15 \times 8$ cm was seen arising from the lesser curvature of the body of stomach. The tumour had short single vascular pedicle from left gastric artery. The central solid component was $10 \times 8 \times 3$ cm and multiple cystic structures were arising from that. The cystic component had multiple concentric growing layers within each cyst. Fluid was clear, serous and had normal AFP levels. There was no lymph node or invasion to adjacent structure. The tumour was completely excised

Figure 1 GT based on narrow pedicle on left gastric artery pointed by arrow (A), stomach primarily repaired (B), excised specimen sized $23 \times 15 \times 8$ cm (C), teratoma was predominantly cystic having clear fluid (D), Tumour had uniform concentric growing layers (E).

Figure 2 X-ray abdomen showing calcification (A), mid-sagittal view of CECT abdomen showing predominantly cystic structure occupying whole of the abdominal cavity (B), non-contrast enhanced axial view at L-1 vertebra showing calcified solid, cystic structure anterior to stomach (C), contrast enhanced view at same level showing vascular pedicle by left arrow and stomach containing oral contrast pointed by right arrow (D).

with a small part of the lesser curvature, and the defect in the stomach was primarily repaired (figure 2). The whole tumour was removed in single piece. Histological examination revealed immature teratoma with no foci of malignant tumour and no staining for AFP.

OUTCOME AND FOLLOW-UP

Postoperative period was normal. The child was asymptomatic in 1 year of follow-up with normal AFP levels.

DISCUSSION

In infancy and early childhood, the most frequent location of teratomas is extragonadal, whereas after childhood, they are more commonly located in the gonads.^{3 4} The sacrococcygeal teratoma is the most common teratoma of infancy.^{1 3} GT are very rare, with a reported incidence of less than 1% of all paediatric teratomas.^{5 6} The first case of GT was reported in 1922 by Eustermann and Sentry.⁷ Since then, many cases of this rare tumour had been reported^{6–13} This tumour usually occurs in children less than 1 year of age, especially neonates. However, few cases have been reported in older children.⁹ Male predominates over female.^{6–10} This tumour arises most commonly from the posterior wall of the stomach, but in our case it was from anterior wall toward lesser curvature.⁶ The GT is exogastric in 58–70% of cases, while it is endogastric in 30% of cases.⁶ Extra-gastric GT are usually asymptomatic, while intra GT may present as an abdominal mass, gastrointestinal bleeding and/or obstructive manifestations.^{11 12}

Teratoma are usually benign, however, 1–2% of ovarian teratomas and 2–3% of testicular teratomas and few cases of GT undergo malignant transformation.^{3 5 12 14} Coincidence of GT with the Wiedemann-Beckwith syndrome and peritoneal gliomatosis has also been described, but none of these features was identified in our case.⁷

The radiograph showed calcification in 40–60% of cases of GT.^{6–8} Other usual causes of calcification in infancy are mesenchymal hamartoma (MH) of kidney or neuroblastoma.¹¹ Next in US presence of normal kidney rule out MH and serum AFP, NSE, urinary VMA differentiate the rest of two entities. Other US features of teratoma described in literature are a cystic lesion with a densely echogenic tubercle (Rokitansky nodule) projecting into the cyst lumen; a diffusely or partially echogenic mass with the echogenic area demonstrating sound attenuation owing to sebaceous material and hair within the cyst cavity, or multiple thin, echogenic bands caused by hair in the cyst cavity.⁶ Pure sebum within the cyst may be hypoechoic or anechoic, while fluid-fluid levels seen due to sebum floating above aqueous fluid, which appears less echogenic than the sebum layer.⁶ None of the above feature were identified in our case because huge cystic component containing clear fluid occupying the major portion of abdominal cavity.

In contrast enhanced CT fat attenuation within a cyst, with or without calcification in the wall, is diagnostic for mature cystic teratoma.^{13 15} A floating mass of hair can sometimes be identified at the fat-aqueous fluid interface, but due to immature nature of tumour only calcification was identified in our case.¹⁵ Further MRI scan confirms the sebaceous component as it has very high signal intensity on T1-weighted images, similar to retroperitoneal fat.¹⁵

Treatment of GT is total excision and primary closure of the gastric wall. Partial, subtotal and total gastrectomies can be performed depending on the extent of stomach involvement. Because of short and narrow pedicel we excised the tumour with small stomach bite only. There were no features of recurrence of tumour in follow-up of 1 year.

We had tried to summarised in brief all the aspects of GT because although the tumour is very rare but had excellent outcome if promptly treated.

Learning points

- Any infant with cystic abdominal swelling should be suspected to have cystic teratoma in the differential diagnosis.
- Abdominal x-ray is very useful and adds much to the diagnosis and to plan further investigation, so it should be a part and parcel of routine investigation in every child with distension of the abdomen.
- AFP adds to the diagnosis, but not exclude the diagnosis of teratoma as its value is normal in pure mature teratoma and rises only if immature teratoma had foci of malignancy or component of well formed intestinal epithelium within the tumour. Further it is the earliest marker of tumour recurrence.
- Exogastric GT can be safely removed with small portion of gastric mucosa only if it has a pedicel, with good long-term results.

Competing interests None.

Patient consent Obtained.

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