

megakaryocytes, thus ruling out megakaryocytic hypoplasia(11). Iron deficiency and megaloblastic anemia have been implicated to cause thrombocytopenia and/or defective platelet functions(12,13). Presence of disseminated intravascular coagulation might explain occurrence of thrombocytopenia in some of the cases(4).

The results of this study show that in PEM even in absence of acute infection, mild thrombocytopenia and some abnormalities of platelet functions exist. Though no clinical evidence of bleeding was noticed, these abnormalities render the PEM cases more prone to bleeding with some other additional insult to hemostatic mechanisms.

REFERENCES

1. Dorantes S, Barren I, Arias N, Vazquez J, Seto R. Pathogenesis of purpura in the child with severe malnutrition. *Trop Pediatr* 1964, 65: 438-445.
2. Bhattacharya AK, Basu AK, Chatterjee S, Mandal JM. Hemorrhagic manifestations in Kwashiorker and marasmus. *Bull Calcutta Sch Trop Med* 1967, 15: 96-98.
3. Hassanein EA, Tankovaky I. Disturbances of coagulation mechanism in protein caloric malnutrition. *Trop Geogr Med* 1973, 25: 158-162.
4. Bhushan V, Bhagwat GP. Disseminated intravascular coagulation in malnutrition: Preliminary communication. *Center Afr J Med* 1980, 26: 180-181.
5. Bello A, Dermates S, Marques JL, Jaimes ML. Physical and biochemical characteristics of platelets in severely malnourished children with purpura. *Scand J Hematol* 1971, 8: 321-327.
6. Neame PB, Kotton JG, Walker IF, *et al.* Thrombocytopenia in septicemia: The role of disseminated intravascular coagulation. *Blood* 1980, 56: 88-94.

7. Nutrition Subcommittee of the Indian Academy of Pediatrics. Report of the Convenor. *Indian Pediatr* 1972, 4: 360.
8. Borchgrevink CF. A method of measuring platelet adhesiveness *in vivo*. *Acta Med Scand* 1960, 168: 157-164.
9. Zuckay MB, Borolli J. Platelet clumping produced by connective tissue suspension and by collagen. *Proc Soc Exper Biol Med* 1962, 109: 774-787.
10. Hardisty KM, Hutten RA. Kaolin clotting time of platelet rich plasma: a test of platelet factor-III availability. *Br J Hematol* 1965, 11: 258-265.
11. Bhattacharya AK, Basu AK. Thrombocytopenic purpura in Kwashiorker. *Bull Calcutta Sch Trop Med* 1968, 16: 1-2.
12. Marwaha RK, Singh S, Garewal G, Marwaha N, Walia BNS, Kumar L. Bleeding manifestations in megaloblastic anemia. *Indian J Pediatr* 1989, 56: 243-247.
13. Malhotra RK, Saraya AK, Kumar R, Chowdhary VP, Ghai OP. Platelet aggregation in iron deficiency anemia. *Indian J Pediatr* 1985, 52: 139-146.

Gastric Teratoma in Children

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Gastric teratoma is a rare benign tumor, found most frequently in boys. World literature yields only 55 reports of gastric

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teratoma since 1922(1) of whom only three cases are in girls(2-4). The most extensive recent review is that of Matias and Huang(5) published in 1973 in which they tabulated 30 cases. In 1986, 4 cases were reported by Meerabai(6) in the 8th Congress of the Asian Association of Pediatric Surgeons. Our series of five children is perhaps the largest in literature and takes the total number of cases to 62 as two cases of this series have been reported earlier(7,8).

Case Reports

The details of the patients and their outcome are summarized in the Table. All the tumors were large of varying shapes, lobulated and variegated in consistency. Cut section showed cystic spaces with clear fluid, mucoid material, bone and cartilage. Histologically they contained tissue derived from tridermal components (alimentary epithelium, respiratory epithelium, glandular structures, fibrofatty tissue, cartilage, new bone). Case 2 contained female gonadal tissue and Case 4 neural tissue and choroidal layer of eye ball. All tumors were benign histologically.

Discussion

Gastric teratoma is usually a disease of the male child though teratoma in other sites (sacroccygeal, gonadal, mediastinal and retroperitoneal) are more common in females. In our series, 4 of the 5 cases presented before the age of 1 year, consistent with earlier observations (37 out of 47 presented before 1 year)(2). Cases have been reported in newborns(3,9).

Abdominal swellings were the common modes of presentation in our series; Purvis *et al.*(2) reported that 39 out of 47 cases presented with such swellings. Rapid dete-

rioration of general condition, pallor, malnutrition and fever were other modes of presentation(2,5). The tumor also produced difficulty during child birth(5,9). Gastrointestinal bleeding, fever, vomiting and respiratory distress were also noted in younger patients(2,4,9).

Calcifications were found in 4 cases. Huge conglomerate calcifications diagnostic of teratoma were found in 2 of our cases, and in 60% of cases reported.

Excretory pyelography was performed in 3 cases. It was normal in 2 cases but misleading in Case 2(8) which was preoperatively diagnosed as Wilms' tumor.

Most teratomas were diagnosed after laparotomy, though abdominal calcification in plain X-ray was suggestive. Upper abdominal masses need differentiation from neuroblastoma, nephroblastoma, omental or mesenteric cysts, teratoma and duplication cyst(4,10). Preoperative diagnosis is difficult if, as in our series, facilities for ultrasonography and CT scan are not available.

The sites of origin of all but one of the tumors were the greater curvature of the stomach, one took origin from the lesser curvature. They were usually large in size (10-20 cm in diameter), arising from any portion of the stomach (endo or exogastric). Case 2 in our series was exogastric.

The operative procedure consisted of local excision with closure of the defect in the stomach wall. Enucleation from the gastric wall was done in Case 2, and splenectomy required in Case 3 due to extensive adhesion of the tumor with the spleen.

Partial gastrectomy has been reported in 10 cases, total gastrectomy in one(2,4), and no surgery in 3 cases due to associated anomaly and complications(2,5).

Out of 62 patients, 56 patients recovered, 3 died after surgery(5,8). The high

TABLE—Details of 5 Cases

Case No.	Age & weight	Clinical presentation	Plain X-ray	Excretory urography	Operative findings	Treatment	Pathology		Outcome
							Size of tumor	Weight of tumor	
1.	2 yrs 12 kg	Abdominal mass and vomiting	Calcification	Normal	Arising from posterior wall at the lesser curvature near the fundus of stomach	Excision	18×16×6cm	—	Alive
2.	5 months 6.5 kg	Abdominal mass and Rt inguinal hernia	-do-	Distorted & displaced pelvicalyceal pattern (L)	Arising from greater curvature near the cardioesophageal junction of stomach	Enucleation	16×15×7cm	1.4 kg	Died due to diarrhea and poor general condition following pre-operative chemotherapy
3.	2 months 3.5 kg	Upper abdominal mass & discharge of pus from protruding mass following incision elsewhere	-do-	—	Arising from greater curvature of stomach and adherent to spleen	Excision with partial gastrectomy and splenectomy	20×10×8cm	0.5 kg	Died due to septicemia
4.	8 months 7.9 kg	Upper abdominal mass	-do-	Normal	Arising from posterior wall of stomach and adherent to pancreas	Excision & repair of stomach	17×15×11cm	1 kg	Alive
5.	2 days 2.5 kg	Abdominal swelling, pallor, gastric bleeding, respiratory distress	No	—	—	—	Autopsy showed firm lobulated mass arising from greater curvature of stomach	—	Died due to gastric bleeding before operation

mortality in our series was due to misdiagnosis and associated complications.

All our cases were benign histologically. Malignant changes have not been reported in literature either.

REFERENCES

1. Eusterman GB, Sentry EG. Benign tumors of the stomach. *Surg Gynec Obstet* 1922, 34: 5.
2. Purvis JM, Miller RC, Bernard I. Gastric teratoma—first reported case in a female. *J Pediatr Surg* 1979, 14: 86-88.
3. Esposito G, Cigliano B, Paledetto R. Abdominothoracic gastric teratoma in a female newborn infant. *J Pediatr Surg* 1983, 18: 304-305.
4. Senocak ME, Kale G, Buyukpamukcu N, Hicsonmez A, Caglar M. Gastric teratoma in children including the third reported female case. *J Pediatr Surg* 1990, 25: 681-684.
5. Matias IC, Huang YC. Gastric teratoma in infancy: report of a case and review of world literature. *Ann Surg* 1973, 170: 631-636.
6. Meerabai D. Four cases of gastric teratoma. Presented at the 8th Congress of the Asian Association of Pediatric Surgeons, Calcutta, 1986.
7. Chatterjee SK, Banerjee RK. Teratoma of the stomach. *Indian Pediatr* 1964, 1: 328-329.
8. Nandy AK, Sengupta P, Chatterjee SK. Teratoma of the stomach. *J Pediatr Surg* 1974, 9: 563-564.
9. De Anglis VC. Gastric teratoma in a newborn infant. *Surgery* 1969, 16: 794-797.
10. Agarwal UK, Parashar SK, Srivastava SK. Gastric teratoma in an infant. *Indian J Surg* 1972, 34: 87-88.

Kostmann Syndrome

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Kostmann syndrome, which was first described in 1956, is a group of closely related disorders characterized by congenital or infantile neutropenia resulting in frequent episodes of fever, skin infection, boils, aphthous stomatitis and other infections presenting in early life(1,2). Most of the cases described were autosomal recessive(1) and hence consanguinity was common. About 50 cases have been described in the literature so far with no case report in Indian literature. We report a young girl with Kostmann syndrome seen by us at the age of 7 years.

Case Report

A girl aged 7 years, belonging to Christian family from Goa, was referred to us for recurrent infections. She was a product of consanguinous marriage and born full-term by Cesarean section. At the age of 3 months, she developed first episode of fever with otitis externa. Since then, she developed frequent infections affecting skin, ear, throat, mouth, gums and urinary tract at regular intervals. She had spontaneous abscesses at three occasions needing

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