case report

Solitary intestinal myofibroma – an unusual cause of neonatal intestinal obstruction

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Congenital solitary myofibroma is an exceptional tumor of newborn period and presents as solitary or multiple lesions usually confined to soft tissues. It induces intestinal obstruction or perforation, which most frequently involves the jejunum and ileum. However, jejunoileal atresia is the most frequently encountered cause of small bowel obstruction in the neonatal period. We report a new case of solitary myofibroma located in the wall of the ileum, measuring 2×3 cm^2 in size, about 25 cm from the ileocecal junction, in a 17-day-old baby girl who presented with abdominal distention and bilious vomiting. Laparotomy was performed on the patient and the tumor was removed; the patient did well after surgery. Despite all the common causes of intestinal obstruction—intestinal atresia, Hirschsprung disease, anorectal anomaly, malrotation, and meconium passage problem in the neonatal period—myofibroma of the small bowel has to be considered because treatment is fairly easy and prognosis is excellent.

Congenital solitary myofibroma is a rare tumor of newborn period, with a small number of cases reported. It was first described in 1965 by Kauffman and Stout[1] in 2 newborns. Solitary intestinal myofibromatosis induces intestinal obstruction or perforation, which most frequently involves the jejunum and ileum; however, jejunoileal atresia is the most frequently encountered cause of small bowel obstruction in the neonatal period.[2] We report here a new case in a newborn with solitary myofibroma involving the ileum without other systemic involvement such as skin or bone.

CASE

A 17-day-old baby girl was born at term after an uneventful pregnancy. The birth weight was 2.8 kg, and the Apgar score was 9 and 10 at 5 and 10 minutes, respectively. The passage of meconium was observed soon after the birth. The baby presented on day 17 of age to our emergency room with abdominal distention and projectile bilious vomiting after each feed. The baby was febrile and had no diarrhea or any other complaint. On examination, the baby looked well; she was neither dehydrated nor distressed. Her weight was 3.2 kg (25th centile), length was 48 cm (10th centile), and head circumference was 35 cm (25th centile). Her vital signs were normal for age. The abdomen was distended and lax; it was not tender and had no hepatosplenomegaly. No palpable masses could be felt. The bowel sounds slightly increased. A nasogastric tube was inserted where bilious aspirate was observed. On investigation, the plane abdominal x-ray showed slight dilated loops of small bowel with some fluid levels. An abdominal ultrasound was not conclusive. Barium swallow showed a free flow of contrast throughout the entire part of the esophagus and the duodenum to the jejunum. The patient was taken to operation room for exploratory laparotomy, where a mass measuring 2×3 cm^2 was found in the wall of the ileum (**Figure 1**). The appendix, cecum, and large bowel showed normal appearance. Resection and end-to-end anastomosis were performed. The postsurgical course was uneventful, and the patient did well. A specimen comprising a portion of the ileum (2×1 cm^2 in diameter) was pathologically examined. The overview of the specimen showed a luminal obstruction by the thickened wall of the ileum (**Figure 2**). Microscopically, the specimen showed a polyploid mass with focal ulceration associated with a granulation tissue (**Figure 3**). The mass was seen in the submucosa, involving the muscularis propria reaching to serosa, hypercellular-

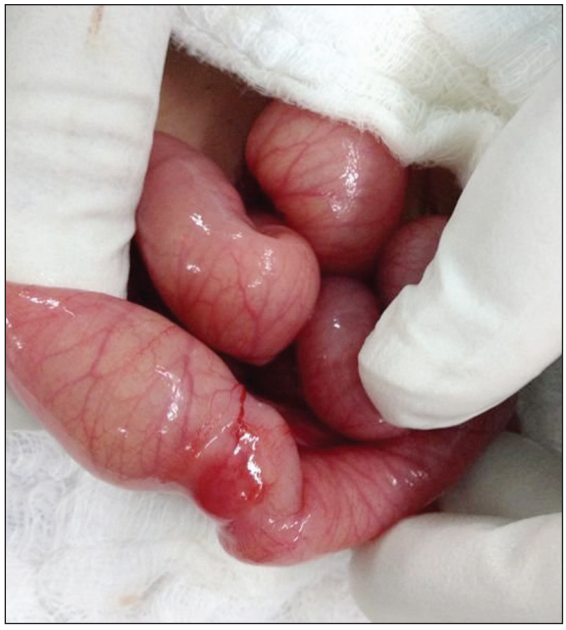

Figure 1. Operative view of the lesion.

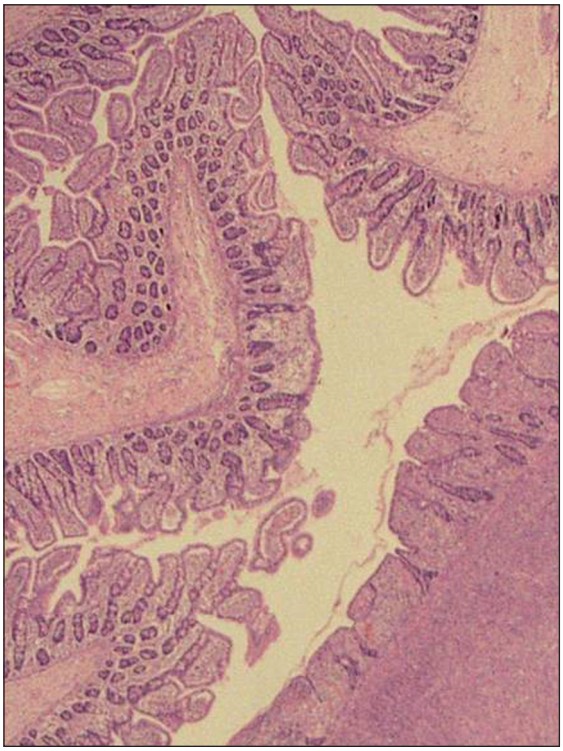

Figure 3. On the left side, unremarkable small bowel is present with normal mucosal villi, submucosa, and muscular smooth muscle wall. On the right side, there is a lesion that causes thickening of the bowel wall and in the picture it extends from the mucosa through the submucosa into the muscularis propria. The mucosa overlying the lesion is ulcerated (hematoxylin and eosin, 20×).

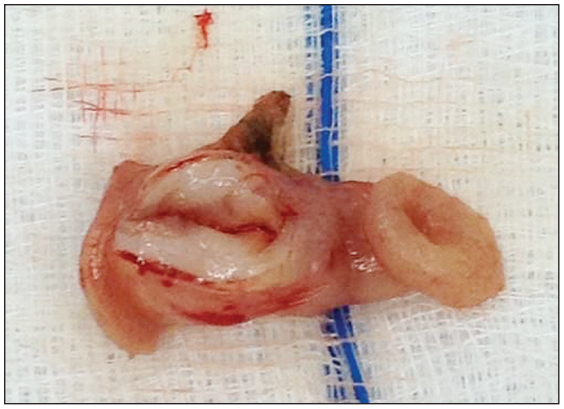

Figure 2. Overview of the specimen shows the luminal obstruction by thickened wall.

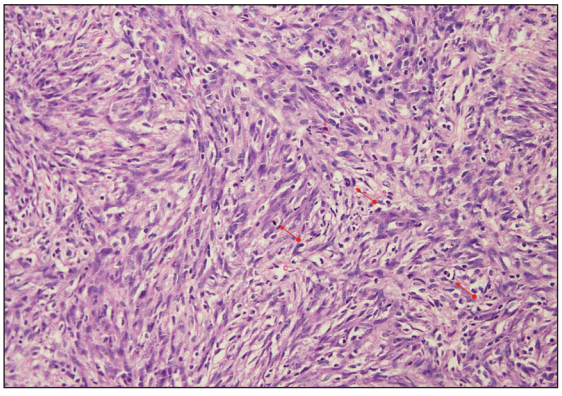

Figure 4. A high microscopic power of the lesion showing hypercellularity and spindle tumor cells with elongated plump nuclei containing several mitotic figures (arrows). Tumor necrosis and nuclear pleomorphism are not present (hematoxylin and eosin, 40×).

ity, and spindle tumor cells with several mitotic figures (**Figure 4**). Immunohistochemical stains showed tumor cells that were vimentin positive, desmin negative, smooth muscle actin negative, CD117 negative, CD31 positive (cytoplasmic), and beta-catenin positive. This result of the tumor based on the gross pathologic as well as histological and immunohistochemical features best fits as myofibroma; although most myofibromas are positive for smooth muscle actin, 5% to 10% can be negative as in our index case.

DISCUSSION

Congenital intestinal myofibromatosis is an exceptional tumor of newborn period, which involves the intestinal wall and causes either obstruction, leading to vomiting and abdominal distention as in our index case, or perforation of the small bowel. The prognosis is excellent in the case of total tumor resection, and neither

metastasis nor recurrence has been reported.[3,4] Solitary intestinal fibromatosis belongs to the group of pediatric fibromatosis including infantile fibromatosis and myofibromatosis, and presents as solitary or multiple lesions usually confined to soft tissues.[5] The etiology of myofibromatosis is obscure, but it has been associated with a hereditary autosomic dominance pattern, with reduced penetrance that can be modulated by hormonal factors such as estrogen. It has been hypothesized that a congenital defect in smooth muscle cell causes an increase in the number of estrogen receptors favoring their proliferation. This hormonal stimulation could begin in fetal life from maternal estrogens.[6] Infantile myofibromatosis with intestinal involvement has been reported and is found positive for muscle cell markers such as smooth muscle actin and desmin.[6] However, 10% of cases with myofibromatosis can be negative for these cell markers as in our index case.

Patients with solitary myofibromas in the intestinal tract are treated surgically with total resection and have excellent prognosis without metastasis or recurrence.[7]

In conclusion, bilious vomiting and abdominal distention in the newborn period should always be considered a sign of intestinal obstruction. Most common causes are intestinal atresia, Hirschsprung disease, anorectal anomaly, malrotation, and meconium passage problem. Sepsis in the newborn period may be associated with adynamic ileus and signs of intestinal obstruction.[8] Despite all of the above common causes of intestinal obstruction in the neonatal period, myofibroma of the small bowel has to be considered because treatment is fairly easy and prognosis is excellent.

REFERENCES

1. Kauffman SI, Stout AP. Congenital mesenchymal tumors. Cancer 1965;18:460-476,1965.
2. Desa DJ. The alimentary tract. In: Wigglesworth JS. Singer DB. Editors. Textbook of fetal and perinatal pathology 1991, pp 927-33.
3. Numanoglu A, Davies J, Millar AJW. Et al. Congenital solitary intestinal fibromatosis. Eur J Pediatr Surg 2002;12:337-40
4. Choo KL, Borzi PA, Mortimore RJ. Neonatal intestinal obstruction from solitary intestinal fibromatosis. Pediatr Surg int 2001;17:467-9.
5. Weiss SW, Goldblum JR . Fibrous tumors of infancy and childhood. In: Strauss M. editor. Enzinger and Weiss,s soft tissue tumors 4th: 2001,pp.357-63
6. Rodrigo Menendez-Arzac, David Valdez-Mendez, Rodolfo Landa. Solitary infantile gastrointestinal myofibroma: case report. J Pediatr Surg 2005;40:1361-1363.
7. Saguem MH, Brochu P, Ouirmet A. The localized intestinal form of infantile myofibromatosis. Ann Pathol 1990;10:126-9.
8. Lilien LD, Srinivasav G, Pyati SP, et al. Green vomiting in the first 72 hours in normal infants. Am J Dis Child 1986;140:662 -664.