Journal of Neonatal Surgery 2018; 7:7
Doi: 10.21699/jns.v7i1.676

OPEN ACCESS

CASE REPORT

Neonatal Solitary Intestinal Myofibromatosis

Rachida Laamiri,*[1] Nahla Kechiche,[1] Meriem Braiki,[1] Rim Hadhri,[2] Lassaad Sahnoun,[1] Mongi Mekki,[1] Mohsen Belghith,[1] Abdellatif Nouri[1]

[1] Department of Pediatric Surgery, Fattouma Bourguiba University Hospital- Monastir, Tunisia

[2] Department of Anatomy and Pathologic Cytology, Fattouma Bourguiba University Hospital-Monastir, Tunisia

How to cite: Laamiri R, Kechiche N, Braiki M, Hadhri R, Sahnoun L, Mekki M, et al. Neonatal solitary intestinal myofibromatosis. J Neonatal Surg. 2018; 7:7.

ABSTRACT

Solitary intestinal myofibromatosis (SIF) is a very rare condition affecting the pediatric population and carries good prognosis following adequate management based on segmental resection. We describe a rare case who presented with features of neonatal intestinal obstruction due to a solitary stenosing fibrotic lesion originating from the ileum and compatible with SIF.

Keywords: Solitary intestinal fibromatosis-;Neonatal intestinal obstruction; Pediatrics

INTRODUCTION

Infantile myofibromatosis is a rare disorder of the fibroblastic/ myofibroblastic proliferation in children and one of the most common benign fibrous tumors of infancy [1]. Most intestinal myofibromatosis (IM) occur in neonates or infants less than 24 months of age [1]. This disorder can present as solitary or multifocal lesions with several implications. Intestinal involvement is described as one of the least common differential diagnoses for a neonatal intestinal obstruction [2]. In the present case, we report a patient with SIF presented with features of intestinal obstruction.

CASE REPORT

A 3-day-old, 2.7Kg, full-term male neonate delivered vaginally after an uncomplicated pregnancy was referred to our department with features of neonatal intestinal obstruction- abdominal distension, bilious vomiting and meconium retention. Several prenatal ultrasonic investigations were reported as normal. Clinical examination revealed generalized abdominal distension and tenderness with exaggerated bowel sounds. The perineum was normal. The laboratory data was within normal limits. Abdominal X-ray showed multiple air-fluid levels; there was no evidence of pneumoperitoneum. Abdominal ultrasonography showed an intraperitoneal ovoid and well-circumscribed hyperechogenic soft-tissue mass measuring 40x35 mm located in the right lumbar fossa between the liver and the right kidney. Emergency exploratory laparotomy revealed a 4 cm calcified long mass arising from the ileum wall located at 10 cm proximal to the ileocoecal valve (Fig.1).

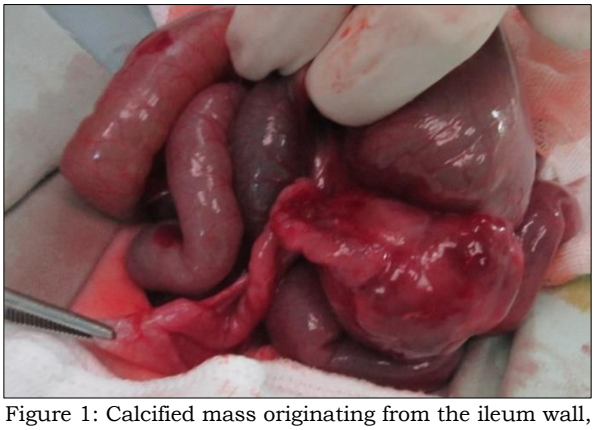

Figure 1: Calcified mass originating from the ileum wall, measuring 4 cm long axis and located at 10cm proximal to the Ileocoecal valve.

Resection of the bowel mass and end-to-end anastomosis was done. Histopathology revealed exten-

Correspondence*: Rachida Laamiri, Department of Pediatric Surgery, Fattouma Bourguiba University Hospital- Monastir, Tunisia
E mail: lamiri.rachida@yahoo.fr

Submitted: 12-10-2017 Accepted: 19-11-2017
Conflict of interest: Nil Source of Support: Nil

sive proliferation of fibromatous spindle cells consistent with the diagnosis of infantile myofibromatosis; immunohistochemistry was positive for vimentin and smooth muscle actin. The patient's post-operative recovery was uneventful.

DISCUSSION

Fibromatosis have been described in different parts of the body including the skin, muscles, viscera, bone and subcutaneous tissue. Although IM is the most common fibrous proliferation in infancy, it remains a less known entity and it is rarely described as a cause of neonatal intestinal obstruction [3]. It was first reported in 1965 by Kaufmann and Stout in 2 newborns [4].

Fibromatosis can occur as focal fibroma or as multi-centric lesions and solitary lesions occur more frequently than the multiple one [4].

The etiology remains unclear; probably there are multiple coinciding predisposing and causative agents, such as hereditary influences, trauma, viral infections, neuropathy and may be metabolic disturbances [4].

Because SIF is a rare entity and has been documented as a rare intestinal obstruction and perforation's etiology in infancy. Preoperative diagnosis is not reported in the literature and the diagnosis is often made shortly after birth [4]. The small bowel is commonly more involved than the colon and obstructing lesions are commonly found on exploratory laparotomy [1, 5].

Optimal treatment requires early recognition and segmental bowel resection removing the obstructing lesion followed by primary digestive anastomosis. If presentation is delayed, complications such perforation and sepsis may occur and the management would be staged with initial stoma [6].
Histopathologically, short bundles and fascicles of spindle shaped cells associated with myofibroblastic features characterize the lesion [6]. Variations in microscopic appearance including nuclear atypia, necrosis or calcifications can be found but have no implications on prognosis [3]. Immunohistochemically, tumor cells react positively and have strong cytoplasm staining for actin and vimentin [3].

The solitary forms usually have a good prognosis after complete resection with no reported local or distant recurrence [2, 7].

It is documented that prognosis is excellent when intestinal fibromatosis is solitary and completely removed. However, patients with visceral involvement have a worse prognosis [4].

Consent: Authors have submitted signed consent form from legal guardian of the patient and available with editorial office.

Authors' contribution: All the authors equally contributed in concept, design, drafting of manuscript, and approved final version of the manuscript.

REFERENCES

1. Mashiah J, Hadj-Rabia S, Dompmartin A, Harroche A, Laloum-Grynberg E, Wolter M, et al. Infantile myofibromatosis: a series of 28 cases. J Am Acad Dermatol. 2014; 71:264-70.
2. Jones VS, Philip C, Harilal KR. Infantile visceral myofibromatosis--a rare cause of neonatal intestinal obstruction. J Pediatr Surg. 2007; 42:732-4.
3. Behar PM, Albritton FD, Muller S, Todd NW. Multicentric infantile myofibromatosis. Int J Pediatr Otorhinolaryngol. 1998; 45:249-54.
4. Arets HG, Blanco C, Thunnissen FB, Heineman E. Solitary intestinal fibromatosis as a cause of bile vomiting in a neonate. J Pediatr Surg. 2000; 35:643-5.
5. Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues; clinicopathologic and immunohistochemical study of 54 cases. Am J Surg Pathol. 2006; 30:104-13.
6. Numanoglu A, Davies J, Millar AJ, Rode H. Congenital solitary intestinal fibromatosis. Eur J Pediatr Surg. 2002; 12:337-40.
7. Choo KL, Borzi PA, Mortimore RJ. Neonatal intestinal obstruction from solitary intestinal fibromatosis. Pediatr Surg Int. 2001; 17:467-9.