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Solitary intestinal fibromatosis-A rare cause of intestinal obstruction in childhood

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Summary

Intestinal involvement of fibromatosis can occur as part of a generalised or multicentric disease. Solitary lesions causing intestinal obstruction are rare. A few cases have been reported in neonates, all showing an excellent prognosis following local excision. This report describes a similar lesion in a 13 year old boy.

Key words: Intestinal fibromatosis, intestinal obstruction

Introduction

Fibromatoses affecting infants and children comprise a group of lesions with a variable histological picture and behavior [2]. Infantile myofibromatosis, previously known as congenital generalized fibromatosis, has been found to be the most common of all types. Multicentric form of the disease with visceral involvement carry an unfavorable prognosis, whereas solitary lesions, or multiple lesions without visceral involvement, follow a benign course. Solitary lesions occur more commonly in boys, the main site being the soft tissues of the head and neck region, and the trunk. A few cases of intestinal involvement of solitary lesions have appeared in the literature presenting as bowel obstruction in neonates [6,8,9]. These unusual lesions have shown a completely benign behavior following local excision. Pathological findings are uniform and confirm their benignity. Our report describes a similar case in a 13 year old boy with intestinal obstruction.

Case report

A 13 year old boy presented to the Pediatric Surgery department with clear vomiting, pain in the abdomen localized to the right side, fever and loss of appetite. On physical examination, the abdomen was found to be slightly distended with tenderness and rebound over the right iliac fossa. Otherwise, the general condition of the patient was good. Peristalsis was normal. The rectal examination did not reveal any abnormality.

Plain X-rays of the abdomen showed air-fluid levels over the right flank.

Laboratory findings were: WBC 13,500, Hb 12.4, Hemocrit 38.

Clinically, acute appendicitis was suspected. At laparotmy the appendix appeared normal. Further exploration revealed a hard nodular mass of the ileal wall, 20 cm from the ileocecal valve, with dilatation of the proximal intestine. A simple intestinal resection with end to end anastomosis was performed.

Postoperative course was uneventful. Subsequent investigations including barium meal and passage, barium enema with double contrast, plain X-rays of the skeleton and isotope bone scan did not demonstrate any abnormality.

Pathological findings: The resected specimen consisted of a 5 cm small intestine with normal mucosa. In the mid-portion of the specimen the intestinal wall was thickened with a circumscribed, grayish-white hard mass protruding mostly extramurally, measuring 20x20x15 mm.

Multiple sections including the mucosa were examined by light microscopy and staining with hematoxylin and eosin. Periodic acid Schiff reagent, Masson trichrome, phosphotunostic acid and hematoxylin, and Pearl's iron. The mass was made of dense collagenous tissue with scattered spindle-shaped cells possessing uniform, elongated nuclei.

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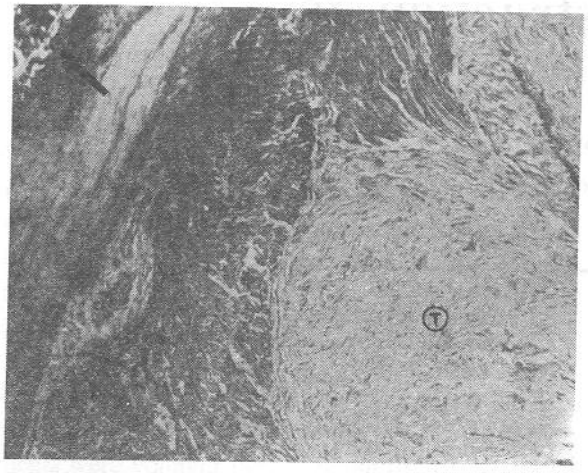

Fig. 1. Histological section showing nodular fibrous growth with compression and encroachment on the muscular layers. At the upper left corner, the thinned submucosa can be seen (Masson Trichrome x 25).

Mitotic figures were absent. Here and there foci of calcification and lymphocytic infiltration around thin walled blood vessels were seen. The lesion was mainly located in the serosa with compression and encroachment on the muscular layers and thinning of the submucosa. No hemosiderin pigment was detected (Fig 1).

Electron microscopic examination of gluteraldehide fixed tissue showed fibroblasts with prominent rough endoplasmasmic reticulum and abundant mature collagen fibrils in the extracellular space. Microfilaments were present, no condensations were detected; nor was there basement membrane-like material.

Discussion

The AFIP classification of fibrous tumors of infancy and childhood includes eight distinctive fibromatoses [2]. The congenital generalized fibromatosis described by Stout in 1954 [8] and subsequently classified as infantile myofibromatosis by Chung and Enzinger is by far the most common of all types [11]. The lesions occur as widely distributed indurative nodules anywhere in the body except in the central nervous system [1,7]. However, the AFIP review mentions the case of a 10 day old boy who had multicentric disease including lesions within the CNS [2]. It is interesting to note that one of the cases included in Allen's review [1], a female child with congenital multicentric fibromatosis had a mass in the left parental region of the brain as revealed by carotid angiogram. Histological proof as to the nature of the lesion is lacking and the case has been excluded from the series. Other reports on infantile myofibromatoses [4,10] describe the entity as a rare disorder. Clinically it presents as solitary or multicentric lesions. The latter lesions are confined to the soft tissue and bone, and in 37 per cent of cases visceral involvement has been reported [2]. Solitary lesions occur characteristically in skin, muscle or subcutaneous tissue. Only five cases have come to our notice that have presented as solitary lesions in the intestine causing bowel obstruction [6,8,9]. Histological differentiation between the two types is difficult, if not impossible, as both types show variable amounts of extracellular collagen and spindle-shaped cells possessing characteristics of fibroblasts and myofibroblasts. It may be that the myofilamentous elements present in the cytoplasm are indicative of an early phase of fibroblastic differentiation [14]. Hence, as recommended by Gonzales, Crussi and Noronha [6], solitary intestinal fibromatosis seems to us the most appropriate name for this condition.

Although the benign nature of the condition is often evident from the histopathological appearence, cellular lesions may pose difficulty in differentiating them from malignant neoplasms. Localized intestinal masses that occur in other diseases merit mention. Neurofibromatosis may involve the bowel in as many as 15-25 % of patients. The patients, usually adults, have other accompanying symptomatology. The neurogenic origin of the lesion can easily be demostrated by appropriate immunohistochemical and electron microscopic examination. Mesenteric fibromatosis can be a manifestation of Gardner's syndrome. Histologically the lesions resemble other fibromatoses, but patients have other stigmata of the disease, such as intestinal polyposis, osteoma and cutaneous cysts [5]. Another form of mesenteric fibromatosis (intraabdominal desmoid) occurs as large masses, usually following abdominal surgery. A careful evaluation of the clinical set up and morphological features with ensure a correct diagnosis in most cases [3].

In summary, a rare case of intestinal obstruction caused by a solitary lesion of fibromatosis in a 13 year-old-boy is presented. Histopathological differential diagnosis is discussed at it bears important implications as regards the immediate and long-term patient management. Simple intestinal resection is adequate for this benign disorder.

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