Mesenteric fibromatosis of the ileocolic area: a case report

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Abstract Fibromatosis is a kind of spindle cell tumor which is characterized by the remarkable proliferation and invasive growth of fibrous tissue. It often arises from the abdominal wall or the extremities and rarely from the mesentery and abdominal organs. The patient was 35 year old female whose major complaints were an abdominal mass and pain. She underwent a right hemicolectomy after the tumor was confirmed by abdominal ultrasonography and computerized tomography. Hereby the pathophysiology, diagnosis and the treatment of the disease is discussed.

Keywords Mesenteric fibromatosis · Desmoid tumor · Surgery

Introduction

Mesenteric fibromatosis is a very rare benign fibrous lesion which is usually arises from the abdominal wall, extremities and rarely from the mesentery and abdominal organs. Incidence of this disease is 2–4 per million, approximately [1]. Its biological behaviour holds a place between benign fibrous lesions and fibrosarcoma. They may be locally aggressive but they do not metastatize and they have a well-known local recurrence tendency. These tumors exhibit benign histological features but high tendency towards involving the visceral abdominal struc-

tures during its growth. Mesenteric fibromatosis is usually localized in the mesentery of the small bowel but may arise from ileocolic mesentery, gastrocolic ligament and omentum [2].

Case

A 35-year old woman admitted to hospital with a complaint of an abdominal mass and pain in the right lower quadrant. She had underwent a sectio cesarean operation 11 years ago. Abdominal examination revealed an abdominal mass in the right lower quadrant. Laboratory data and tumor markers comprised normal values. Ultrasonography revealed a solid abdominal mass which was approximately 6 cm in diameter which was localised nearby the caecum. Moreover CT scan revealed a solid tumoral lesion which was 6.5 cm in diameter which was reported to be possibly originating from the caecum; but colonoscopy was negative. After laparotomy a tumor which was arising from the ileocolic mesentery was observed. A right hemicolectomy was performed. She was discharged from the hospital after a 7 days stay after the operation with no complication. The tumor was 6 cm. in diameter and it was localised nearby the caecum but mucosal involvement couldn't be proven (Fig. 1). The tumor had a smooth capsule-like formation around it. The tumor had a solid cut surface with gray-white colour containing hemoragic areas. Histologically, long sweeping fascicles with low to moderate cellularity was observed in the dense collagen matrix with no glandular cells. Spindle tumor cells were bland and the

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Fig. 1 The tumor was near to the caecum and 7 cm in diameter

mitotic activity was very low (Fig. 2). Actin and Desmin were locally positive, CD-117 and S-100 were found negative in immunohistochemical dying.

Discussion

Mesenteric fibromatosis is a rare benign fibrous lesion which is usually localized in the small bowel mesentery. It is the most common primary tumor of the mesentery [2]. Although it is considered as a benign tumor, local recurrence after tumor resection was reported. As a result of its biological malignancy it has a high tendency towards involving the visceral abdominal structures during its growth. In addition; the location of mesenteric fibromatosis may predispose to marked morbidity due to progression of the disease.

Most cases are sporadic but there are associations with previous intraabdominal surgery, estrogenic stimulation,

Fig. 2 Microscopic view of the tumor. Spindle tumor cells were bland and the mitotic activity was low (HE×100)

Gardner syndrome, Familial Polyposis Coli and bilateral ovarian fibromatosis [3]. However this kind of tumor could be seen in patients who didn't underwent a surgical operation before. Yet; their precise etiology and natural history remains obscure.

Data on age and gender vary; Yannopoulos reported that males and females were affected with equal frequency [2]. But Burke reported that the tumor was more commonly observed in males; reporting the mean age to be 41 years [4].

The tumor can present with a wide variety of symptoms. Most patients' have mild or no complaint; so usually the tumor is detected during routine examination. The tumor also may be presented with frightening symptoms resulting from complications, such as torsion, hemorrhage or infarction of the mass.

Sclerosing mesenteritis, mesenteric panniculitis, inflammatory fibrosarcoma of the mesentery and retroperitoneum must be considered in differential diagnosis during pathological evaluation. The lesion is composed of proliferated fibroblasts and its histological features are specific enough to allow definitive diagnosis in the majority of cases. Unfortunately in some cases the differential diagnosis between fibromatosis and well-differentiated fibrosarcoma is difficult [5]. The differential diagnosis includes tumor originating from smooth muscle, neuronal, adipose and stromal tissue of the gastro-intestinal tract, as well as tumor of histiocytic origin.

Some authors recommend an aggressive surgical approach aiming a wide excision [6] while others advocate a less radical strategy [7]. Wide excision, including the resection of the adjacent organ is recommended for these tumors as they have a tendency toward local recurrence. If the tumor is not resected, it eventually causes pain and obstructive symptoms by mass pressure on neighboring structures such as ureter, small or large intestines. Burke reported 23% of all tumors recurred and there was a strong association between patients with Gardner syndrome and recurrence [5]. Recurrent disease can be resected and patients may live for extended periods with recurrent disease, especially in cases of sporadic desmoid tumors. Smith indicates that prolonged survival is possible in patients with intraabdominal desmoid tumors, even for those with technically irresectable or recurrent lesions [8]. Other therapy modalities include treatment with antiestrogenic agents, steroids, cytotoxic chemotherapy and postoperative irradiation. Surgical excision is mentioned to be the only curative method for treatment of this kind of tumor. Medical therapy is of unclear benefit. Medical therapies should be kept in mind as complementary methods in the management of these patients but they are no substitute for surgical procedures.

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