Mesenteric fibromatosis with ureteric stenosis

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Summary: We report a case of a 37 year old man who presented with vomiting and colicky abdominal pain and who was found to have microscopic haematuria. Radiological examination showed a right hydronephrosis apparently caused by a paracael tumour extending to involve the right ureter. This was resected and proved histologically to be a mesenteric fibromatosis. The patient has none of the factors predisposing to the development of this lesion, in particular Gardner's syndrome. Eight months following surgery he appears to have made a full recovery. Previous publications on this rare intra-abdominal neoplasm are reviewed.

Introduction

The fibromatoses are a group of benign fibrous tissue proliferations with similar microscopic appearances and behave in a fashion intermediate between that of benign fibrous lesions and fibrosarcomas. Like fibrosarcomas they infiltrate and recur but unlike them they do not metastasize. The fibromatoses occur predominantly in adults and consist of differentiated fibrous tissue forming firm poorly circumscribed nodules.

The concept of the fibromatoses as a group of similar lesions was first advanced by Stout in 1961. They have been subdivided by Weiss & Enzinger into 2 groups, superficial or fascial, and deep (musculoaponeurotic) fibromatoses. The superficial group includes palmar, plantar and penile fibromatoses and the deep group extra-abdominal, abdominal and intra-abdominal fibromatoses or desmoids. The intra-abdominal lesions include pelvic and mesenteric fibromatosis and Gardner's syndrome. Electron microscopic studies have shown that the proliferating cell type in these lesions is the myofibroblast.3

In this paper we report a rare example of an intra-abdominal fibromatosis in mesentery which had extended to involve the ureter.

Case report

A 37 year old man presented with a colicky right sided abdominal pain and a feeling of fullness. On examination he was found to be well built and fit looking. No significant abnormalities were found in any system.

Routine haematological and biochemical screens were normal as was a stool examination. The erythrocyte sedimentation rate (ESR) was 3 mm/hour. Urinalysis showed microscopic haematuria. An abdominal ultrasound showed a dilated right renal pelvis and an intra-abdominal mass. An intravenous pyelogram confirmed the presence of hydronephrosis (Figure 1). A computed tomographic (CT) scan showed a paracael tumour apparently causing stenosis of the middle third of the ureter (Figure 2). No other radiological abnormalities were demonstrated. A fine needle aspiration showed no evidence of malignancy but could not contribute a definitive diagnosis.

The patient underwent a laparotomy at which a mass was found in the mesentery and wall of the caecum with a tail-like extension towards the ureter and retroperitoneum. A right hemicolecotomy was performed with resection of the affected segment of ureter. Fourteen centimetres of terminal ileum and 13 cm of large intestine were received. In the wall of the caecum there was a firm mass 9.5 × 6.5 × 4 cm which extended to the submucosa. The cut section showed a grey-white tumour with an irregularly whorled appearance. The border of the lesion was ill-defined. The segment of ureter received was surrounded by tissue with similar appearances.

Histologically the neoplasm was composed of bundles of fairly uniform spindle shaped cells with intervening collagen fibres. There was no evidence of cytological atypia and few mitoses were seen. At the edge of the lesion it showed an infiltrative growth pattern. It was seen to extend up to the large...
Figure 1 Intravenous pyelogram showing stenosis of the middle third of the ureter with proximal hydronephrosis.

Figure 2 CT scan showing paracael mass extending posteromedially to involve the ureter (arrow).

Figure 3 Ureter being encroached upon by tumour (H. & E. x 25).

intestinal submucosa and around the ureter at its deep aspect (Figure 3). The mucosal surface of both structures appeared normal. There was no evidence of inflammation. The appearances were those of a fibromatosis and excision appeared to be complete.

Eight months after surgery the patient remains asymptomatic and entirely well and is being followed up with computerized tomography to screen for recurrence.

Discussion

Mesenteric fibromatosis is a rare intra-abdominal tumour. It can occur as a single lesion, with retroperitoneal extension or accompanied by a separate retroperitoneal fibromatosis. The tumour usually presents as an asymptomatic abdominal mass with some reaching a considerable size before diagnosis, the largest reported being 50 cm in diameter. Symptoms may however be produced by compression of small and large intestine, and of the ureters as in this case.

In the small number of series of these tumours which have been reported a wide age range has been described, from 10–60 years in that of Yannopoulos & Stout. The sex incidence varies, with Yannopoulos & Stout reporting an equal frequency of males and females and Kim et al. and Weiss & Enzinger reporting a female predominance.

The preoperative differential diagnosis of these tumours may include intestinal carcinoma, carcinoid and lymphoma, and also idiopathic retroperitoneal fibrosis. For the latter there were, in this case, none of the systemic manifestations often associated with retroperitoneal fibrosis such as weight loss, fever and elevated ESR. As there are no certain clinical or radiological pointers to the diagnosis of fibromatosis, the nature of the pathology may only be confirmed, as in this case, at laparotomy.

The histological differential diagnosis includes reactive fibrosis and fibrosarcoma. For the former, there is no evidence of previous injury or
haemorrhage and the absence of nuclear pleomorphism, significant mitotic activity or atypical mitoses assists in the differentiation of fibromatosis from fibrosarcoma. There is no evidence of the inflammation, fat necrosis and vascular involvement seen in retroperitoneal fibrosis.

Treatment has generally been by local excision which can be curative. However repeated recurrences can occur. Radiotherapy, hormonal therapy and chemotherapy have been used with variable results in fibromatoses. In this and other anatomical sites Weiss & Enzinger in their series report no close correlation between the histological picture and behaviour, and that some cases appear to be unresponsive to all forms of treatment.

Although it is not apparently the case with our patient the most likely cause of these lesions is tissue injury in persons with a genetic predisposition towards excessive fibrous growth. In the series of Simpson et al. 15 of 22 patients had a history of previous intestinal surgery and in that of Hayry et al. 13 of 40 patients with abdominal fibromatosis had been operated upon in the region of the subsequent growth.

Mesenteric and retroperitoneal fibromatoses whilst seen as isolated tumours may also be found in association with Gardner's syndrome. Weary et al. found fibromatosis of all forms in 45% of patients with Gardner's syndrome and Naylor et al. found 4 of their 28 cases developed mesenteric and retroperitoneal fibromatosis, 3 of whom had a history of previous surgery.

The bony and soft tissue tumours of Gardner's syndrome, including mesenteric fibromatosis, may precede gastro-intestinal polyposis. These may lead to the subsequent development of gastro-intestinal carcinoma. Suarez & Hall therefore recommend screening of the bowel in all patients with mesenteric fibromatosis. In our case there is no evidence of any of the stigmata of Gardner's syndrome.

References
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