Case Report

El **Abdominal wall desmoid tumour: revisited**

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Introduction: Desmoid tumours are histologically fibrous neoplasms originating from the musculoaponeurotic structures throughout the body. The term desmoid, was first used by Muller in 1838, was derived from the Greek word *desmos,* which means tendon like. Desmoid tumours often appear as infiltrative, well-differentiated, firm overgrowths of fibrous tissue, and are generally slowly locally invasive.
Nevertheless, the synonym aggressive fibromatosis describes the marked cellularity and aggressive local behavior. The course and the tendency for recurrence make the treatment of this relatively rare fibrous tumour challenging.
Here we report of a 17-year old female presented with a desmoid tumour of the anterior abdominal wall.

Case report:

A 17 years old female presented to our outpatient clinic with painless lump beneath an appendicectomy scar. She claimed that this lump was slowly increasing in size. She had had appendicectomy one year ago. At presentation she had good appetite, regular bowel habits, regular menses and normal frequency of micturition. There was no family history of similar condition or malignancy. Abdominal examination revealed a firm lump of five centimeters in diameter medial to the site of the appendicectomy scar. It is not attached to the skin, but to the muscles of the anterior abdominal wall. Complete blood count, urinalysis, plain x ray of the abdomen and chest were normal.

Ultrasound scan of the abdomen and pelvis showed a right sided pelvic mass with normal pelvic organs. Computerized tomography showed a well defined oval shaped low density mass in the pelvis, adjacent to the rectum surrounded by loops of small bowel (Fig1).

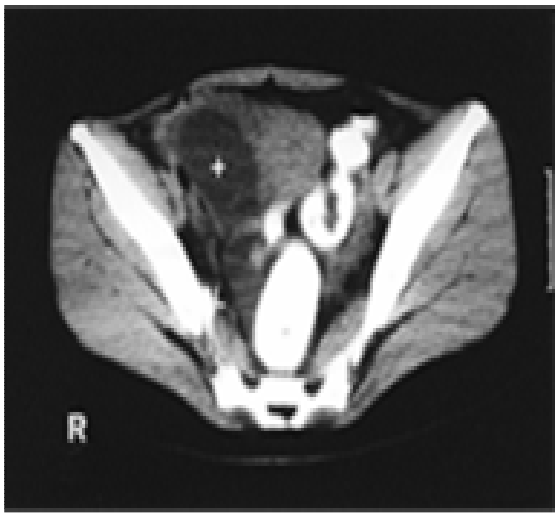

Fig 1: Solid mass extending from the appendicectomy scar into the pelvis

Another spindle shaped mass was also seen within the rectus sheath.
At operation a firm, fibrotic mass was found arising from the anterior abdominal wall. The pelvic cavity, adenexia, small and large bowels were normal. Wide local excision with safety margin was done, and the incision was closed in layers. Histopathology reported whorls and bundles of fibroblasts, producing varying quantities of collagen in different areas. The lesion was infiltrating the surrounding muscles. The appearance was consistent with desmoid tumour. We didn't

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refer for refer her for radiotherapy because we thought that the resection was curative.

She was followed up for one year where investigations for possible associated conditions were ruled out i.1 colonoscopy showed normal; colonic mucosa with normal vascular pattern. 18 months after surgery she presented with a recurrent tumour at the lower end of the same scar. The recurrence was excised with wide safety margin. During this operation there was no pelvic recurrence. Thereafter we referred her for local radiotherapy.

Discussion:

Desmoid tumours are uncommon slowly growing soft tissue neoplasms. Their incidence was reported to be 2–4/1.000.000[1]. Although we are aware that there were few cases operated by our colleagues, this is the first case of desmoid tumour to be reported in Sudan. Depending on the site of location of the desmoid tumour they are classified as: extra abdominal, abdominal wall desmoids occurring typically in women during or following pregnancy; and intra-abdominal desmoids either in the pelvis or mesentery. Accordingly, our case has a combined component of abdominal wall and pelvic desmoid tumours.

Although the cause of desmoid tumours is uncertain, it may be related to trauma (mainly surgical), hormonal factors, and may have a genetic association. This makes it fairly sound that the previous appendicectomy was probably the predisposing factor in our case.

Desmoid tumours occur at a rate of 10-20% in patients with Familial Adenomatous Polyposis (FAP)[2], however, sporadic forms do occur. In contrast colonoscopy was normal. On the other hand, like our case desmoid tumours most commonly appear in young women particularly during or after pregnancy. In the contrary, desmoid regress during menopause, after tamoxifen treatment and may regress after exposure to oral contraceptives[3]. This is explained by the fact that the proliferative response of fibroblasts occurs in presence of estrogen. The later fact may explain the presence of desmoid on our 17 year old female. Like our case desmoids are more common in the age group 10-40 years, however, they do occur in young children, older adults, but are twice as common in females as in males.

Myofibroblast is the cell incriminated to be responsible for the development of desmoid tumours. Despite the benign histologic appearance and negligible metastatic potential of desmoids, they tend to infiltrate adjacent muscle bundles, frequently entrapping them leading to their degeneration. They may cause significant morbidity, and mortality may result from pressure effects of obstructing vital structures. Fortunately this is not the case on our patient.

As in our case, desmoids grow slowly and their symptoms depend on the location of the tumour. Also, as in our case, patients with intra-abdominal desmoid may have an asymptomatic mass, however, symptoms of intestinal, vascular and urinary obstruction or neural involvement may occur.

CT scan and MRI help determining the extent of the tumour, its relationship to nearby structures, and for follow-up after surgery. MRI was reported in the literature to be superior to CT scan in defining the pattern and the extent of involvement as well as in determining if recurrence has occurred after surgical removal[3].

As in our case, most desmoid tumours on gross examination are confined to the musculature and the overlying aponeurosis or fascia. Its size varies from five to 20 cm. Desmoids are firm, cut with a gritty sensation, and on cross section reveal a glistening white, coarsely trabeculated surface resembling scar tissue[1]. Histologically the tumours are composed of abundant collagen surrounding poorly circumscribed bundles of spindle cells. The dense bundles of eosinophilic spindle cells contain regular nuclei and pale cytoplasm with neither mitoses nor giant cells. Macrophages, giant cells, and lymphocytes are present peripherally. This is in contrast to those in a fibrosarcoma, which has greater mitotic activity, an increased nuclear: cytoplasmic ratio, greater vascularity, less collagen production, and a paucity of

immune cells. Immunostaining with vimentin, alpha smooth muscle actin and desmin are also helpful in distinguishing this tumour.

Primary surgery with free surgical margins is the most successful primary treatment modality and complete surgical excision of desmoid tumours is the only effective method of cure. Positive margins after surgery reflect a high risk for recurrence. Re-operation for the treatment of recurrent disease is advocated by most authors, resulting in a cure rate similar to that of the primary surgical resection[1]. Function and structure preserving procedures should be the primary goal. In selected patients, whose only option is amputation, it may be prudent to observe them with their limb and tumour intact[5].

The combined treatment with radiotherapy showed a significantly longer free survival than surgical resection alone. Radiotherapy for recurrence has significantly worse prognoses compared with adjuvant radiotherapy[4]. We didn't refer our patient for radiotherapy because we thought that the first surgery was curative. However after the second surgery we did.

Endocrine treatment with agents such as tamoxifen has resulted in response rates of up to 50%. Other hormonal agents employed in the treatment of desmoids include medroxyprogesterone acetate, megace, toremifen, and gonadotropin releasing hormone analogue goserelin.

On the other hand, cytotoxic chemotherapeutic agents have been used in inoperable or unresectable tumours, progressive or residual disease, and in rare cases neoadjuvant therapy is used to facilitate wide surgical resection[1]. In recurrence, chemotherapeutic regimen of doxorubicin, dacarbazine, and carboplatin may be effective.

Although neither haematogenous nor lymphatic metastases have been observed, most lesions are refractory to treatment because the overall local recurrence rates range from 19% to 75%. Most recurrences are usually observed within 3 years, and nearly all by 6 years. Age may affect the recurrence rates since local recurrence may occur more likely in younger patients with extra-abdominal lesions. The region of involvement also affects local recurrence because local recurrence rates for intra-abdominal tumours are higher than those reported for extra-abdominal tumours, ranging from 57% to 86% after complete resection. However, mortality is rare in extra abdominal desmoids[1].

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