CASE REPORT

Desmoid Tumour of Lower Abdomen

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ABSTRACT

A desmoid tumour is slow growing fibromatosis with aggressive infiltration of adjacent tissue and extremely unusual systemic metastases. We report a case of a female patient aged 25 years who had a mass in lower abdomen for 2 years. There was no previous history of any surgical intervention. Preoperative evaluation included ultrasound and computed tomography. Patient underwent primary resection with wide margins. Histology revealed a desmoid tumour.

Key words: *Desmoid tumour. Fibromatoses. Clonal neoplasm. Wide resection. Palliation. Bard Composix.*

INTRODUCTION

Desmoid tumours refer to aggressive fibromatoses, which are benign in nature. Despite their aggressive local infiltration, systemic metastases are extremely unusual.[1] They constitute 3% of all soft tissue tumours and 0.03% of all neoplasm. In the normal population the overall incidence is about 2.4-4.3 cases per million.[2] The risk increases to about 1000 folds (i.e. 50-70%) in patients with familial adenomatous polyposis (FAP); this association is referred to as Gardner syndrome.[3] The neoplasm occurs sporadically in location of trauma, scars and irradiation.[1]

Molecular studies demonstrated desmoids tumours in FAP as clonal neoplasm arising from germ line mutation or changes in APC alleles.[3] Abdominal wall desmoids typically occur in young females during gestation or frequently within a year of childbirth. This has been correlated with detection of estrogen receptors in the substance of these tumours.[2] Typical presentation is that of a painless mass. Symptoms arise due to compression of the adjacent neurovascular bundles.[1] Treatment modality includes resection of tumour with a wide margin. Even with wide local resection the recurrence rate approaches 40%. Antiproliferative agents and cytotoxic chemotherapeutic agents like doxorubicin, dactinomycin and carboplatin used to palliate the aggressive nature of desmoid tumours with variable results.

This report described desmoid tumour in a young parous female.

CASE REPORT

A young female aged 22 years, presented with mass in the right lower abdomen for the last 2 years. The rest of the history did not provide any suggestive clue. She was married, with a spontaneous vaginal delivery 4 years back.

On examination, abdomen was distended at the lower quadrant, more at the right as compared to the left. On palpation it was a single, lobulated, firm, non tender mass extending from the right iliac fossa, hypogastrium and left iliac fossa downwards up to the mons pubis. It had well defined margins.

Pre-operative investigations including ultrasound (U/S), computerized tomography of abdomen and pelvis and fine needle aspiration cytology FNAC were done. U/S showed irregular heterogeneous mass with micro lobulations in suprapubic region causing thickening of subcutaneous tissue. CT abdomen and pelvis (Figure 1) revealed a large rounded lesion in hypogastrium arising from the recti muscle which had infiltrated the anterior abdominal wall measuring 9.0 x 8.5 x 10 cm. Posteriorly it was pressing on the bladder, abutting the adjacent bowel loops wihtout invasion. There was a small solid lesion also attached to the anterior abdominal wall on the left side adjacent to the above lesion. Uterus and ovaries were normal. There was no lymphadenopathy.

FNAC was concluded as a hemorrhagic aspirate showing spindle cell lesions suggestive of soft tissue neoplasm.

Trucut biopsy followed by histological examination revealed multiple linear cores showing proliferation of

Figure 1: CT scan abdomen and pelvis with contrast enhancement demonstrates the desmoid tumour infiltrating the anterior abdominal wall.

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spindle shaped cells with esinophilic cytoplasm and elongated nuclei without any evidence of nuclear pleomorphism, increased mitosis or necrosis.

Per operatively, the mass was adherent to anterior rectus sheath and rectus muscle, and to the parietal peritoneum with no intra abdominal extension. Strong adhesions were found between the mass and pubic symphsis. Tumour measured about 10 x 10 cm in size. Wide excision was done and using polypropylene mesh the abdominal wall defect was repaired. Cross- section had a cauliflower appearance (Figure 2).

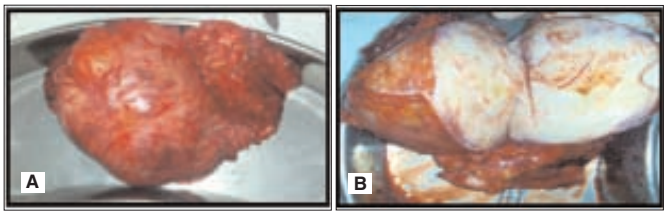

Figure 2 (A and B): Specimen of the resected tumour, gross (A), cut section showing cauliflower appearance (B).

DISCUSSION

Desmoid tumours are benign deep fibromatoses, originating from fascia and muscle aponeurosis with an infiltrating growth. They are primarily located abdominally or intraabdominally, whereas only sporadic cases describe localization within the thorax wall or retroperitoneal. Depending on the tumours size, therapy and the negative resection margins, recurrence occurs in up to 45%.[1]

According to the site of origin it can be superficial arising from the fascia or deep arising from the musculoaponeurotic layer. The superficial tumour is slowly growing and small in size. The deep variant is relatively rapid growing, larger in size and associated with high local recurrence rate. Main locations of the tumour occurrence are the proximal extremities particularly the shoulder girdle, the abdominal wall and the mesentery of intestine in patients with FAP.

CT scan localizes the tumour and excludes metastasis. MRI reveals the tumour's hypointensity on T1 and demonstrates variable signal intensity on T2 weighted imaging,[4] depending on the accumulation of mucoid structures. Therefore, differentiation from other solid tumours is impossible using morphological criteria.

Histology is the only evidentiary method which demonstrates long fascicles of spindle cells of variable cell-density with few mitoses and absence of atypical nucleus-separations. Characteristically, there is a diffuse cell infiltration of adjacent tissue structures. Immuno-histochemical response for actin can be partially positive and immunohistochemical muscle cell markers delimit desmoid tumours from fibrosarcoma.[5]

The therapy of choice is still controversial. Anti-inflammatory treatment, hormone-therapy and chemo-therapy were not shown to be effective.[6] These therapies are limited to patients, in whom resection is technically impossible because of a widespread tumour infiltration.

In case of restricted access to surgical resection desmoid tumours can lead to death. Especially in patients with familial adenomatous polyposis (FAP) undergoing colectomy desmoid tumours are the leading cause of morbidity and mortality.

The indication of adjuvant radiation seems to have a little effect on recurrence.[7] In a comparative analysis, a significantly better local recurrence control was described with radiation and combined surgical resection in comparison to resection only. However, these results of radiation therapy can only be achieved due to a higher complication rate. Other studies show a tumour progress after radiation therapy and a high local recurrence rate.

Therefore, the most effective treatment of accessible and smaller desmoid tumours is still the resection with clear margins, although it may not prevent local recurrence.[8] However, massive mesenteric lesions often occur in FAP patients after colectomy and non surgical treatment has variable and unpredictable objective efficacy.[9]

Surgery always aims at radical tumour resection with free margins, which, depending on localization of surgery, may leave major soft tissue defects behind. Although abdominal wall integrity after full-thickness surgery can be restored with direct sutures, and reconstruction with synthetic materials is a common technique in major abdominal wall defects. In this case, the small tumour resection with tumour free margins and reconstruction of the abdominal wall was performed with a Bard Composix-Mesh. Recent data recommends distant or free muscle flaps for greater abdominal wall defect coverage that is not accessible to local flaps.[10] Additionally, prosthetic material is more susceptible to bacterial infection and other complications, although new developed material showed encouraging experimental results. After resection of the desmoid tumour the abdominal wall defect is replaced with a Bard Composix-Mesh®.

In conclusion, the treatment of desmoid tumours remains enigmatic. Non-surgical treatment results in diverse and unpredictable outcome and is considered to be an opportunity in patients with unresectable lesions or for adjuvant therapy. Radical resection with clear margins remains the principal determinant of outcome with the risk of local recurrence.

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