A RETROPERITONEAL DESMOID TUMOR IN LOWER ABDOMEN - AN UNUSUAL CASE

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ABSTRACT

An extensive tumour in a 35-year-old women with gradual swelling of abdomen. On examination, solid mass over the abdomen of 28 weeks size of gravid uterus. Tumor was fixed with irregular surface and proved to be an aggressive fibromatosis [Desmoid tumour] on histological examination. Twelve months after surgery there was no evidence of recurrent disease. This suggests that tumour resection had been successful.

Keywords: Desmoid tumour, Retroperitoneal

INTRODUCTION

Desmoid tumors (also called aggressive fibromatosis) are benign fibrous growths that occur rarely in the general population (5 to 6 per 1 million per year) but frequently in one of the familial cancer predisposition conditions known as familial adenomatous polyposis (FAP) or Gardner syndrome, affecting between 3.6% and 20% of patients. About 2% of all desmoids arise in patients with FAP. Desmoids may occur in any musculoaponeurotic tissue structures of the body, although they tend to be in extremities and spinal areas in the general population and in the abdomen in FAP. Desmoid tumour or aggressive fibromatosis are locally malignant tumour without any metastatic potential. These tumours are primarily located abdominally but retroperitoneal location is extremely uncommon. These tumours are associated with women of child bearing age, especially during and after pregnancy. Desmoid tumors are not considered sarcomas; however, they have been classified as aggressive fibromatosis, and the optimal treatment plan usually requires multidisciplinary teams only found in sarcoma centers. Surgeons with expertise in sarcomas should guide the approach to extra-abdominal lesions, while intra-abdominal desmoids are best considered by teams including sarcoma surgeons together with GI surgeons, oncologists and radiation therapists all with desmoids experience and expertise. The diagnosis of desmoid tumor, or aggressive fibromatosis, should alert members of the sarcoma team to the potential underlying diagnosis of FAP. This recognition is imperative to the recruitment of gastroenterologists and geneticists who can provide a more focused approach to polyposis screening and genetic testing, which could benefit the extended family as well as the patient.

CASE REPORT

A 35 years old women, gravida 6, presented with complain of dyspepsia, anorexia, gradual swelling of lower abdomen for last 6-7 months. She had no menstrual abnormalities. There was no significant past medical or surgical history.
Examination – she was thin built with average nutrition. Pallor was mild. There was a solid mass over the abdomen of 28 weeks size of gravid uterus. Tumor was fixed with irregular surface and there was no tenderness on palpation. On per speculum- cervix appeared healthy and on per vaginal examination – both the fornices and pouch of Douglas was occupied by tumor.

Investigation - Her Hb% was 8.6gm/dl. All other base line investigations and kidney function test was normal. Ultrasonography of abdomen and pelvis showed a huge solid mass which is retroperitoneal, likely to be malignant. Uterus and both ovaries were normal.

Management – under general anesthesia abdomen was opened with right Para median incision and the tumor was found to be adherent with the entire posterior abdominal wall. Tumor was removed intact with great effort and homeostasis was secured. Then parietal peritoneum cut to open abdominal cavity. Uterus with adnexae, liver, G I system was found to be normal. The abdomen was closed in layers and a drain was put below rectus sheath and above rectus muscle.

The postoperative course was uneventful and the patient was discharged at the 9th postoperative day.

PATHOLOGY
On gross examination the specimen measured 14 × 12 × 10 cm with irregular surface. No lymph node was detected. The cut surface was tan, glistening, white, whorled and firm. There was no necrosis, hemorrhage or any cystic changes.

Microscopic examination showed a lesion Composed of uniform-appearing, spindle-shaped fibroblasts which are loosely and haphazardly arranged [Fig 1]. These plump cells have bland, oval nuclei with tapering ends and moderate amounts of ill-defined eosinophilic cytoplasm. Mitotic figures were extremely rare. There were many Delicate, thin-walled vessels with open lumens. There were no cells with epithelioid features, inflammatory cells, calcification, osseous metaplasia or necrosis [Fig 2]. All the resected margins were free of tumor.

FOLLOW UP
Two monthly follow up was advised for clinical examination and 6 monthly USG of abdomen, especially looked for local recurrence. During one year follow up she was normal clinically and radiologically.

DISCUSSION
Desmoid tumours are benign deep fibromatoses, originating from fascia and muscle aponeurosis with an infiltrating growth [1]. They are primarily located abdominally or intra abdominally, whereas only sporadic cases describe a localisation within the thoracic wall or retroperitoneally [2]. The desmoid tumour is often associated with female gender, Familial Adenomatous Polyposis (FAP) and occasionally with surgical trauma [3]. It has a higher prevalence in women during and after pregnancy [4]. The fibroblast has been shown to exhibit a proliferative response to estrogen. Women with desmoid tumors have regression of their lesions after attaining menopause. Depending on the tumors size, on the therapy and on the negative resection margins, recurrence occurs in up to 45% [5]. Histopathological examination is the only confirmatory method which demonstrates long fascicles of spindle cells of variable cell-density with few mitoses and absence of atypical nucleus. Characteristically, there is a diffuse cell infiltration of adjacent tissue structures. [6]. The most effective treatment of accessible and smaller desmoid tumors is still the resection with negative margins, although it may not prevent local recurrence [2, 3]. The effectiveness and indication of initial and adjuvant radiation is not proven yet. In a comparative analysis, a significantly better local recurrence control was described with radiation in combination of surgical resection in comparison to resection only. However, these results of radiation therapy can
only be achieved due to a higher complication rate. [7].
Athanassios Economou et al studied that the combination of features, such as the history of previous surgery, the age and sex of the patient, the location of the mass within the anterior abdominal wall and the imaging features, make desmoid tumor a strong primary diagnostic consideration even if it is a rare entity and especially in men. The treatment approach remains aggressive and includes complete surgical resection [8].

CONCLUSION

In conclusion, the treatment of desmoid tumors remains enigmatic. Non-surgical treatment resulted 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.

REFERENCES


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Fig 1: Hyalinized hypocellular collagenous tissue x 100

Fig 2: Cells are spindled or stellate in shape and have bland nuclear features x 400