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A Case Report of Desmoid-Type Fibromatosis (Aggressive Fibromatosis).

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ABSTRACT

Desmoid tumors are rare slow growing benign and musculoaponeurotic tumors. These tumors are seen in women of fertile age, especially during and after pregnanc. We report a young female patient with a giant desmoid tumor of the anterior abdominal wall who underwent wide local excision. The patient had no history of an earlier abdominal surgery. Preoperative evaluation included abdominal ultrasound, computed tomograph and FNAC. The histology revealed a desmoid tumor. Primary surgical resection with immediate reconstruction of abdominal defect is the best management of this rarity.

Keywords: Desmoid, Fibromatosis,Abdominal, Aggressive

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INTRODUCTION

Desmoid tumors are rare monoclonal, fibroblastic proliferation which has an unpredictable clinical course. Despite their benign nature, they are known as aggressive fibromatosis, with the potential to cause damage to surrounding structures, leading to organ damage [1]. Desmoid tumours are rare, accounting for 0.03% of all neoplasms and with an incidence rate of 2-4 per million per year with a slight female preponderance and peak incidence in the third and fourth decades [2,3]. The two major groups are superficial (fascial) and deep (musculoaponeurotic). The deep fibromatoses are characterized as extraabdominal, abdominal, and intraabdominal [4]. We report a case of Desmoid tumour in the anterior abdominal wall in a youmg female patient. fnac and histopathological study were consistent with desmoid tumour.

CASE REPORT

A 27 year old female presented with swelling in the anterior abdomen for 6 months which was gradually increasing in size. The patient had no history of previous abdominal surgery or any abdominal trauma. Clinical examination revealed a 8x 5 cm swelling in the right side of the umbilicus. Abdominal ultrasound, computed tomography, and FNAC were done. Wide local excision was done and the specimen was sent for histopathological examination. Post-operative period was uneventful.

Gross examination revealed a soft tissue mass measuring 7 x 6 cm .Cut surface of the mass whitish, fibrous whorled appearance. Fibromuscular tissue measuring 6.5 cm one side and 1.5 cm on other side. Deeper surface of the tumour covered by muscle fibres.

Histopathological examination showed a Spindle cell neoplasm with elongated cells with vesicular nucleus arranged in interlacing bundles and fascicles.Stroma showed thin walled blood vessels, focal myxoid changes, cartilagenous metaplasia and occasional mononuclear cells. No necrosis or mitosis. Muscle infiltration by the tumour cells. Margins surrounded by normal muscle bundles. Depeer margins covered by a thin layer of fibromuscular tissue. Based on the gross and microscopic findings, we gave a diagnosis abdominal desmoid fibromatosis .

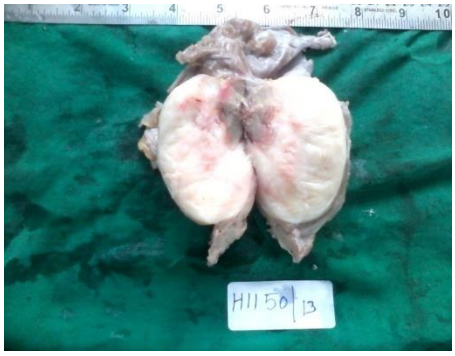
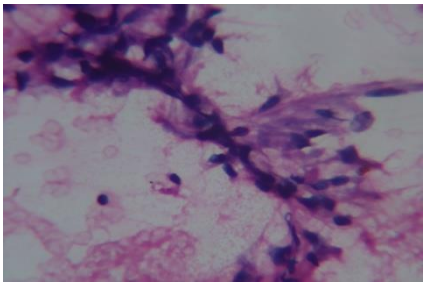


Figure 1: Gross image showing cut surface of the mass whitish ,fibrous whorled appearance.



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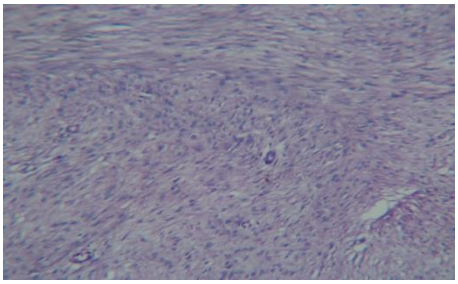

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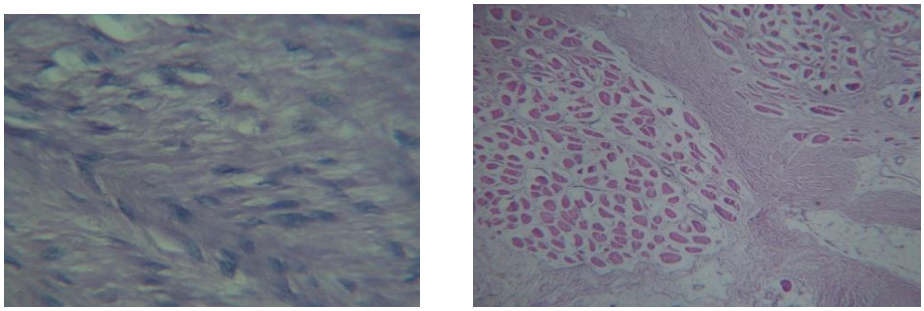

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a) High power view showing clusters of spindle shaped cells with dark staining elongated nuclei.
b) 10 x view showing spindle cells with vesicular nucleus arranged in interlacing bundles and fascicles.
c) 40 x view showing elongated cells with vesicular nucleus.
d) 10 x view showing muscle infiltration by the tumour cells.Margins are surrounded by normal muscle bundles.

DISCUSSION

Desmoid tumors can develop in any site of the body. Superficial desmoids are less aggressive when compared to deep ones and may present as slightly painful lump. The deep ones are more aggressive and have the tendency to damage the adjacent organs. The mortality is higher when they arise in smaller regions such as head and neck [5]. They occur usually between the ages of 25 and 40 years with a strong prevalence among women in the fertile age group. The most common site of predilection is the anterior abdominal wall, with an incidence of 50% [5]. They may be extra-abdominal (the shoulder girdle, trunk, and lower extremities), intra-abdominal (in the abdominal wall, especially the rectus and internal oblique muscles with their fascial coverings, and mesentery or retroperitoneum), multiple familial, and as part of Gardner's syndrome. Abdominal desmoid tumor usually presents as a firm mass with ill-defined margins and no distinct capsule [6]. On cut surface, they are gritty, glistening white and trabeculated resembling scar tissue. Histologically, desmoid tumors consist of elongated fibroblasts and myofibroblasts [7,8].

Wide local excision with reconstruction is the treatment of choice. Peritoneum, intraperitoneal organs, or adjacent bony structures involved by tumor must be resected as well. Incomplete tumor removal may lead to local recurrence [9]. Malignant transformation in desmoid tumor are reported only in few cases, and all were associated with local irradiation [10].

CONCLUSION

The history of painless abdominal mass, 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. Aggressive, wide surgical resection is the best surgical option. Complete surgical excision of desmoid tumors is the most effective method of cure, sometimes necessitating removal of most of an involved anterior abdominal wall in such a giant desmoid tumor with immediate repair of resultant huge defect and reconstruction using prosthetic mesh for better functional results.

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