Abstract

Desmoid tumors are rare, accounting for 0.03% of all neoplasms and with an estimated incidence rate of 2-4 per million per year. Extra-abdominal desmoid is a rare tumor. On the extensive review of literature, we found only one case of extraabdominal desmoid occurring at digit. We present a case of extraabdominal desmoid-type fibromatosis which arose in index finger. A 65 year-old woman presented with gradually increasing swelling in the right index finger since one year. Clinical examination showed ulcerative cauliflower-like growth over the right index finger, which made clinician to diagnose it as a case of squamous cell carcinoma, which turned out to be desmoid-type fibromatosis on histopathological examination. Extra-abdominal desmoid tumor is a locally aggressive tumor, despite being histologically benign. We are presenting this case for its rarity.

INTRODUCTION

The term “desmoid” refers to the hard, tendon-like appearance of the tumor. Nichols in 1923 was the first to recognize and define extra-abdominal desmoid tumor.[1] Desmoid tumours are rare, accounting for 0.03% of all neoplasms and with an estimated incidence rate of 2-4 per million per year with a slight female preponderance and peak incidence in the third and fourth decades.[2] These tumours are more aggressive in younger patients, with recurrence rates up to 87%.[3] Fibromatoses are a group of benign fibrous tissue proliferations characterized by infiltrative growth and a tendency to recur locally without metastasis. The two major groups are superficial (fascial) and deep (musculoaponeurotic). The deep fibromatoses are characterized as extraabdominal, abdominal, and intraabdominal.[4] In various studies, 28–69% of desmoids were intraabdominal (mesenteric or pelvic) or abdominal wall, and the remaining were extraabdominal.[5] Extra-abdominal desmoid fibromatosis arises principally from the connective tissue of muscles and the overlying fascia or aponeurosis (musculoaponeurotic fibromatosis). It chiefly affects the muscles of the shoulder and pelvic girdles and the thigh of adolescents and young adults.

CASE REPORT

A 65 year-old woman presented with a swelling in right index finger since last 6 months. The swelling was associated with dull aching pain. Clinical examination revealed a hard, ulcerated cauliflower lesion measuring 8x7.5x4.0 cm. over the right index finger. (fig 1) The tumour was 1.5 cm away from surgical margin. Bone was not involved by the tumour. (fig 2) Axillary lymph nodes were enlarged. Clinical diagnosis made was squamous cell carcinoma of right index finger with axillary node involvement.

A surgical amputation of the finger was done and sent for histopathological examination. Cut section of mass was whitish homogenous and fleshy. (fig 3) We also received 4 lymph nodes largest measuring 2x1.8x1.5cm, smallest measuring 0.5x0.4cm.

Microscopic examination showed skin with epidermis exhibiting extensive surface ulceration. The dermis showed well-defined tumor mass composed of uniform oval to spindle cells of fibroblast type and spindle cells of myofibroblast type showing variable cellularity with collagenised areas in between. (fig 4) Also seen were few scattered lymphocytes and plasma cells, with focal lymphoid aggregates. Tumor cells showed infiltration between deeper fat and muscle fibres. (fig 5) Masson’s Trichrome showed presence of collagen intervening between tumour cells. (fig 6) Periosteal tissue and bone were free of tumor. Lymphnodes were free of tumour.
Based on these gross and microscopic findings, we gave a diagnosis of extraabdominal desmoid fibromatosis of right index finger.

**Figure 1**
Figure 1: Index finger with noduloulcerative growth

**Figure 2**
Figure 2: X ray showing growth with bone free of tumor.

**Figure 3**
Figure 3: cut surface of the growth-homogenous, greyish white, fleshy

**Figure 4**
Figure 4: Low power view (H & E stain) showing spindle shaped cells and intervening collagen pointed out by arrow.
**DISCUSSION**

Extraabdominal desmoid-type fibromatosis (also known as extraabdominal desmoid, well-differentiated nonmetastasizing fibrosarcoma, and aggressive fibromatosis) arises from the connective tissue of muscle and its overlying aponeurosis or fascia.[4]

Masson and Soule believed the term “desmoid tumor” to be acceptable when referring to a specific form of an infiltrative fibrous growth that may appear in various locations, often recurs locally, and does not metastasize, but under certain extenuating circumstances may cause loss of a limb or more rarely death.[6]

Extra-abdominal desmoid tumour is a non-metastasizing fibrous tumour. It is characterised by infiltrative invasion of soft tissues and a high propensity for local recurrence after surgical excision.[4,7]

A case of extra-abdominal fibromatosis, is a rare entity. Although the etiology of extraabdominal desmoid-type fibromatosis is not known, some associations include genetics, hormonal status (related to pregnancy and often regressing after delivery), and prior trauma.[8] The condition most commonly manifests between puberty and age 40 years, with the highest frequency of cases occurring between 25 and 35 years of age.[4] Our patient is a 65 year old female, this age is higher than that reported in literature. Women are more likely to be affected than men and the tumors are more aggressive in younger patients. [9] Patients younger than 30 years have higher relapse rates than do those over 30 years of age.[10] The tumors are also multicentric in approximately 10%-15% of cases.[11]

Extraabdominal desmoid-type fibromatosis often begins as a painless, deep soft-tissue mass in the lower extremity (eg, thigh, foot, or ankle), upper extremity (eg, shoulder or hand), or the head and neck.[12] In our case patient presented with dull aching pain in right index finger.

Grossly extraabdominal desmoid-type fibromatosis are usually composed of nonencapsulated gray-white tissue confined to the musculature and overlying fascia or aponeurosis.[4,13] The tumors are firm and glistening white on cut section and resemble scar tissue.[4] Our case presented as noduloulcerative mass and on cut section, it was whitish homogenous and fleshy.

The tumors are composed of alternating bundles of locally infiltrating, monomorphic elongated, spindle-shaped fibroblast and myofibroblast bundles within a collagenous Stroma.[10,14] Mitotic activity and cellularity are low (four mitoses per 50 high-power field).[15] Necrosis and hemorrhage are absent features in this tumor, we had similar findings in our case.

The cells are usually actin-positive and CD34- and S100-negative. [15] Masson’s Trichrome stain and reticulin stain can be used to reveal collagen interlaced between tumor cells.[4] In our case, Masson’s Trichrome stain showed collagen between tumour cells. Extraabdominal desmoids are intimately associated with muscle and fascia, with the most common locations being the shoulder and upper extremity (33%) gluteal region and lower extremity (30%)
Extraabdominal-Desmoid-Type Fibromatosis In The Index Finger: A Rare Case Report

chested wall or back(17%), and head and neck (10%) [2, 16, 17] Isolated cases of breast desmoids have been reported.[18]

In our case tumor is seen involving right index finger which is extremely rare site. In a case study done by Easter and Halasz with a nineteen cases of desmoids tumours, twelve cases were of extraabdominal desmoids, out of which single case of digital fibromatosis with recurrence had been mentioned.[19]

To the best of our knowledge, ours is the second case of extra abdominal desmoid tumor occurring at the digital site.

Surgery and radiation therapy, either alone or in combination, have been used to treat extra-abdominal desmoids. For potentially resectable lesions, surgery provides excellent local control, even in those with recurrent disease.[20] In our case, surgical amputation was done and clinical follow up was advised.

CONCLUSION

A case of extra-abdominal fibromatosis in a female occurring at index finger is an extremely rare entity. To the best of our knowledge, ours is the second case of extra-abdominal desmoid tumor occurring at the digital site, which clinically mimicked squamous cell carcinoma. Our patient is a sixty three year-old female, an age is greater than that reported in literature. Desmoid tumors are a challenging clinical condition with locally aggressive behaviour and a strong tendency for recurrence. Histopathological confirmation is a must for diagnosis. Management options include observation, surgical resection, radiotherapy, conventional chemotherapy, hormonal agents, and newer molecular targeted agents. A multidisciplinary approach tailored to the individual patient is usually needed, depending on the location, local effects, and clinical course.

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References

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