ORIGINAL RESEARCH PAPER

General Surgery

EXTRA ABDOMINAL FIBROMATOSIS (DESMOID TUMOR)

KEY WORDS:

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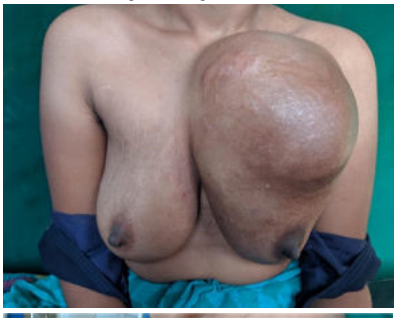
INTRODUCTION

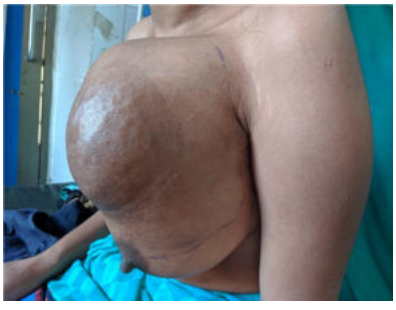
- Desmoid tumors, also known as aggressive fibromatosis, are an extremely rare entity. They are slow growing and histologically benign, but tend to be locally invasive at various anatomic sites. Desmoid tumors originate most frequently from abdominal fascial or musculoaponeurotic structures, although they may appear at extra-abdominal sites. The most common extra-abdominal locations include shoulder, chest wall, back, thigh, and head/neck . Extremity desmoid tumors are extremely rare.

- Desmoid tumors pose a clinically challenging problem because of their tendency to mimic vascular neoplasms. Generally, histopathological examination is necessary for definitive diagnosis, as in our case. Radical resection is necessary for successful excision since desmoid tumors tend to recur locally. However, surgery, radiotherapy, or both are regarded as the treatment(s) of choice for lesions

CASE REPORT

- A 27 year old female presented with c/o painless lump over left breast since 3.5 year.
- No H/o discharge from nipple/retraction of nipple
- No H/o change in overlaying skin colour
- No H/o fever /weight loss
- No significant past history
- No significant family history





PHYSICAL EXAMINATION

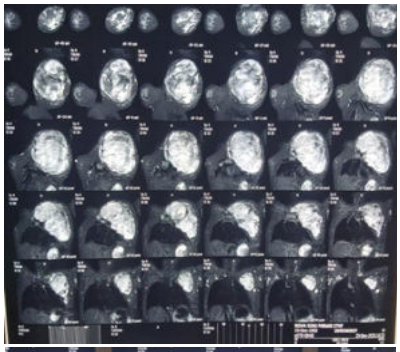
- Vitals were within normal limits
- Lump of approx 25×15×10 cm in size present over the upper outer and inner quadrant of left breast with downward displacement of nipple
- Lump have smooth surface,regular margin,firm in consistency.
- Not fixed to skin but fixed to chest wall
- No lymphadenopathy.

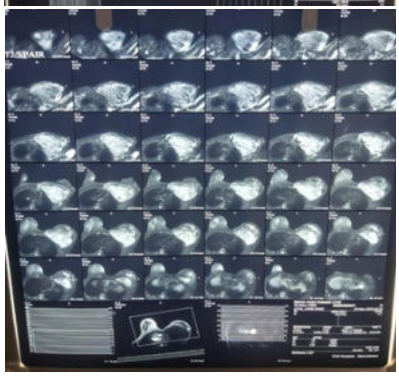
INVESTIGATIONS

USG -approx 13×16×15 cm sized lobulated heterogeneously echotextured lesion s/p/o giant fibroadenoma/phyllodes tumour

MRI –approx 12×15×17 cm sized multilobulated heterogeneously enhancing mass lesion within the left pectoralis major muscle,mass causes

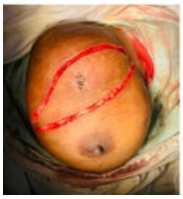
posterolateral displacement of left pectoralis mini muscle
FNAC –phyllodes tumor
Incisional biopsy –lymphocytic mastitis

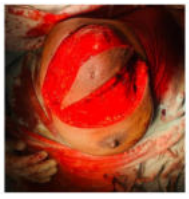


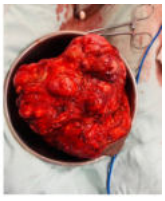


MANAGEMENT

- Wide excision under general anaesthesia planned.
- Elliptical incision kept from upper inner to upper outer quadrant Of left breast towards axilla.
- Complete excision of tumor along with pectoralis major muscle with surrounding margin of 1cm all around.
- Drain kept.







POST OPERATIVE COURSE

- Entire course was uneventful.
- Drain removed on POD-10.
- Suture removed on POD-14.
- Histopathological examination Showed Extra abdominal fibromatosis (borderline fibroblastic tumor with possibility of recurrent behaviour.)

DISCUSSION

- The incidence of desmoid tumors has been reported as 2–4 cases per 1 million. They are typically derived from the abdominal wall, the bowel and its mesentery, or in extra-abdominal sites such as chest wall, shoulder girdle, inguinal region, and neck.

- Possible risk factors for the development of desmoid tumors include female sex, or a previous history of surgery, trauma, or pregnancy(increased estrogen states).

- Desmoid tumour may occur in association with familial adenomatous polyposis,a presentation that is referred to as Gardner's syndrome and is linked to germline mutations in APC gene.

- Clinically, patients are usually in the third to fourth decade of life and have pain, a chest wall mass, or both. The tumor is usually fixed to the chest wall, but not to the overlying skin.

CONCLUSION

Desmoid tumors do not metastasize, but they have a significant propensity to recur locally, with rates as high as 5% to 50%,sometimes despite complete initial resection with histologically negative margins. Such locally aggressive behavior is secondary to microscopic tumor infiltration of muscle and surrounding soft tissues and prompts some to consider them a low-grade form of fibrosarcoma.