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Oncology

A CASE OF GIANT DERMATOFIBROSARCOMA PROTUBERANS OCCURING IN THE ANTERIOR THIGH

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ABSTRACT We present a typical case of A 68 years old male patient presented with swelling in the anterior aspect of Right upper thigh since 10 months, on clinical examination swelling of 12 × 10 cm present below the right inguinal region, firm in consistency, non-tender, margin irregular, surface nodular. USG showed highly vascular swelling? Soft tissue origin. Patient underwent wide local excision with rotation flap under spinal anesthesia. Grossly the tumor was firm with lobulated surface. Histopathology of excisional biopsy suggestive of Dermatofibrosarcoma protuberance. IHC-CD34 showed positive. Patient is been on regular fallow up.

KEYWORDS :

INTRODUCTION

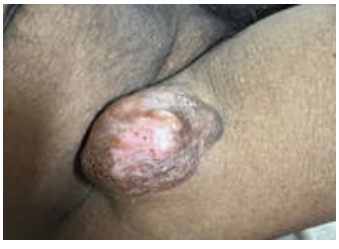
Dermatofibrosarcoma protuberans (dfsp) is an intermediate grade soft tissue tumor occurring in the dermis and subcutaneous tissue which accounts for ~1% of soft tissue sarcoma [1] .the tumor particularly in early stages resemble keloid[2] and is often misdiagnosed .

As they grow larger some can ulcerate and become painful [3]. studies have implicated a chromosomal translocation that results in fusion protein that promotes growth through overproduction of platelet derived growth factor (pdgf) [4]

Case Report

68 years old male presented with swelling in the anterior aspect of right upper thigh since 10 months and pain associated with the swelling since 3 months initially it was small in size 1×2 cm and have progressed to present size of 12× 10 cm over a period of 10 months Upon presentation patient had pain over the swelling. O/E it was size of 10x12 cm with nodular surface, well defined margins, firm in consistency, not fixed to underlying structures. Skin over swelling not pinchable.

Distal peripheral pulses felt well with no focal neurological deficits. usg showed highly vascular well defined solid lesion in the right inguinal region ? soft tissue origin. on the basis of clinical presentation and supporting investigations an diagnosis of dfsp was made and patient was taken for surgery.



SURGERY

An elliptical incision was made over the swelling. Swelling was not fixed to the surrounding structures. Swelling excised in Toto and sent for hpe and primary closure done. hpe showed features suggestive of dfsp and ihc cd34 resulted positive. Patient have been advised Tab. Imatinib [5] 400mg hs for 3 months and is on regular fallow up.

DISCUSSION

Dermatofibrosarcoma protuberans (dfsp) is a rare soft tissue tumor that involves the dermis, subcutaneous fat and in rare cases muscle and fascia. Chromosomal translocation that results in fusion protein that promotes growth through overproduction of platelet derived growth factor [4]. It is histologically characterized by bland spindle cells in a storiform pattern with multiple variants [5]. This needs to be differentiated from other benign and malignant lesions. a vast majority harborst(17;22) (q22;q13) resulting in the formation of col1a1-pdgfb fusion gene transcript [6], which holds not only diagnostic value , but also therapeutic significance. Dermatofibrosarcoma protuberans being intermediate grade malignancy with low likelihood of metastasis but high local recurrence rate.

CONCLUSION

Dermatofibrosarcoma protuberans (dfsp) is an intermediate grade soft tissue sarcoma. Most often been reported in third to fifth decade of life but had been reported in all age groups including congenital presentations.[7] because of their slow growth , lesions are commonly 1 to 5 cm at diagnosis . These tumors are characterized by a proliferation of bland appearing spindle cells in reticular dermis and subcutaneous fat. The cells intercalate between the adipocytes in the subcutaneous fat, resulting in honey comb appearance. The standard treatment is complete surgical excision. The surgical procedure include wide local excision (wle) with tumor free margins[7]. Adjuvant therapies including radiation and targeted therapy should be chosen for patients who are unsuitable for surgical excision.

REFERENCES

1. DuBay D, Cimmino V, Lowe L, Johnson TM and Sondak VK: Low recurrence rate after surgery for dermatofibrosarcoma protuberans: A multidisciplinary approach from a single institution. Cancer. 100:1008–1016. 2004. View Article : Google Scholar : PubMed/NCBI
2. Ucak M. A rare case of misdiagnosis: recurrence of dermatofibrosarcoma protuberans that was treated surgicallyas a keloid. Medical Archives. 2018 Feb;72(1):74.
3. LeBlanc J, Chan C, Zedlitz A. Dermatofibrosarcoma protuberans. Cutis. 2017 Jul 1;100(1):E6-e7.
4. Pereira E, Sahu S, Tambekar M, Dhar R, Mahore K. Myxoid Dermatofibrosarcoma Protuberans: A Diagnostic Dilemma.
5. Choi JH, Ro JY. Cutaneous spindle cell neoplasms: pattern-based diagnostic approach. Archives of pathology & laboratory medicine. 2018 Aug 1;142(8):958-72.
6. Pedeutour F, Simon MP, Minoletti F, Barcelo G, Terrier-Lacombe MJ, Combemale P, Sozzi G, Ayraud N, Turc-Carel C. Translocation, t (17; 22)(q22; q13), in dermatofibrosarcoma protuberans: a new tumor-associated chromosome rearrangement. Cytogenetic and Genome Research. 1996 May 16;72(2-3):171-4.
7. Topar G, Hönlinger B, Eisendle K, Zelger B. An unusual congenital lesion in a 26.year.old woman. Clinical and experimental dermatology. 2008 May 1;33(3):373-4.