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RESEARCH ARTICLE

CASE REPORT- PRE AURICULAR SWELLING PRESENTED AS DERMATOFIBROSARCOMA

Dr. Rekharaj S.¹, Dr. Roshan J.² and Dr. Ashok Naik³

1. Research Scholar, Junior Resident in Department of Otorhinolaryngology at SDM College of Medical Sciences and Hospital, Manjushree Nagar, Sattur Dharwad 580001 Karnataka.
2. Professor in Department of Otorhinolaryngology at SDM College of Medical Sciences and Hospital, Manjushree Nagar, Sattur Dharwad 580001 Karnataka.
3. Professor in Department of Otorhinolaryngology at SDM College of Medical Sciences and Hospital, Manjushree Nagar, Sattur Dharwad 580001 Karnataka.

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Key words:-

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[DFSP]

Abstract

Background: Dermatofibrosarcoma protuberans a rare soft tissue tumour that involves the dermis, subcutaneous fat, and in rare cases muscle and fascia.

Case Report: 34 yr old female patient presented with history of swelling in front of right ear since 2 years, associated with pain since 2 weeks, HRUSG neck reported as Neoplastic pathology and FNAC reported as benign spindle cell tumor. Patient underwent surgical excision of tumour. Frozen section sent was reported as synovial cell sarcoma or schwannoma. Hence on table superficial parotidectomy done. HPE showed Dermatofibrosarcoma protuberans.

Conclusion: Dermatofibrosarcoma protuberans is considered intermediate-grade malignancy with low likelihood of metastasis hence treatment is complete surgical excision. As chance of recurrence is high regular follow up required.

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Introduction:-

1. Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia. The tumor typically presents as a slowly growing.
2. The cause of dermatofibrosarcoma protuberans is not clearly understood. Studies have implicated a chromosomal translocation, resulting in the fusion protein COL1A1-PDGFB, which promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF).
3. The majority of Dermatofibrosarcoma protuberans occur on the trunk (50%), followed by the extremities (35%) and then head and neck (15%).^{[1][2][3][4][5]}

Case Report:-

1. A 34 yr old female patient presented with c/o swelling in front of right ear since 2 years, associated with pain since 2 weeks. H/O excision of the right preauricular swelling 1 year ago.
2. O/E: solitary swelling measuring about 4x4 cms in right preauricular region, scar over the swelling present, local rise of temperature and tenderness present, firm to hard in consistency.
3. HRUSG neck- Neoplastic pathology. FNAC of the swelling- -benign spindle cell tumor.

Corresponding Author:- Dr Ashok naik

Address:- Professor in Department of Otorhinolaryngology at SDM College of Medical Sciences and Hospital, Manjushree Nagar, Sattur Dharwad 580001 Karnataka.

4. Planned for surgical excision , on table frozen section , reported as synovial cell sarcoma or schwannoma. Hence on table superficial parotidectomy was taken and performed, specimen sent for HPE, reported as Dermatofibrosarcoma protuberans (DFSP)



Figure 1:- preauricular swelling.

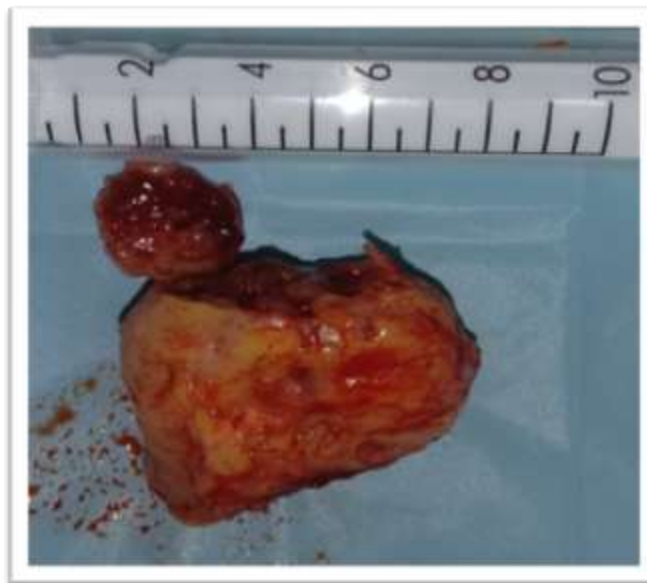


Figure 2:- Pre auricular swelling [parotid].

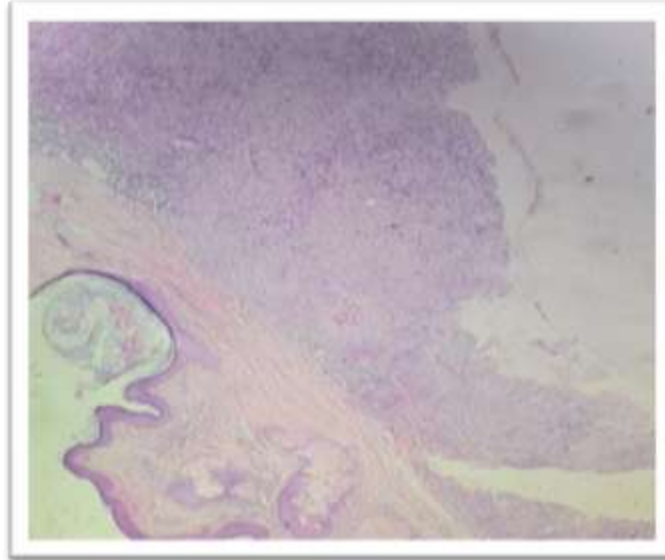


Figure 3:- Histopathological examination.

Discussion:-

1. Dermatofibrosarcoma protuberans is a rare tumor with an incidence rate of 0.8 to 4.5 cases per million persons per year.
2. Dermatofibrosarcoma protuberans occurs most often in adults in the third to fifth decades of life.
3. Diagnosis is made via skin biopsy.
4. Given its propensity for a subclinical extension, the optimal treatment modality for dermatofibrosarcoma protuberans is Mohs micrographic surgery (MMS), a surgical technique that allows complete margin assessment and tissue preservation.
5. Alternatively, dermatofibrosarcoma protuberans can be treated with wide local excision.
6. The chemotherapeutic agent imatinib mesylate is currently FDA-approved for adults with unresectable, recurrent, or metastatic dermatofibrosarcoma protuberans. [
7. The prognosis for patients with completely excised lesions is good.

Conclusion:-

1. Dermatofibrosarcoma protuberans is considered an intermediate-grade malignancy with a low likelihood of metastasis but a high local recurrence rate.
2. Our patient presented with clinically a firm swelling in the right preauricular region and HRUSG neck- FNAC of the swelling showed-benign spindle cell tumor for which she underwent superficial parotidectomy and specimen sent for HPE and HPE confirmed dermatofibrosarcoma protuberans.

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