



**Case Report**

# **Rare Spindle Cell Lesion Of Parotid Gland: Solving The Dilemma**

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**How to cite:** *Medha Mani Gupta., Rare Spindle Cell Lesion Of Parotid Gland: Solving The Dilemma. Int J Histopathol Interpret 2023; 12(2);11-16*

**DOI:** <https://doi.org/10.56501/intjhistopatholinterpret.v12i2.889>

Received : 14/07/2023

Accepted: 24/11/2023

Web Published: 02/12/2023

## **Abstract**

Dermatofibrosarcoma of parotid gland is rare. Microscopic examination showed spindle cells arranged in storiform pattern, having monomorphic ovoid nuclei, moderate to abundant eosinophilic cytoplasm with staghorn-shaped blood vessels. Differential diagnosis of solitary fibrous tumour, DFSP, schwannoma, synovial sarcoma were kept and finally Immunohistochemistry solved the dilemma as Vimentin, CD34, STAT-6 & S-100 were applied. Tumor showed Vimentin & CD 34 positivity, while STAT-6 & S-100 were negative.

**Key words :** *Dermatofibrosarcoma, staghorn vessels, storiform pattern, spindle cells, CD34*

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## BACKGROUND

Dermatofibrosarcoma protuberance is a fibrohistiocytic tumor with low to intermediate malignant potential [1]. It is a locally aggressive tumor originating from dermal and subdermal tissue having high recurrence rate [2]. It usually presents as patch which progresses to nodular cutaneous mass gradually. Most common sites include torso (50-60%) followed by the proximal extremities (20-30%) and less commonly head and neck (10-15%) [3]. Most common age of presentation is 2nd to 5th decade of life with slight female predilection. Head and neck with salivary gland involvement is the rare site for DFSP to occur [4]

## CASE REPORT

A 31year old male came to General Surgery OPD at JNMCH, A.M.U Aligarh with the complaint of slowly enlarging mass on the right side of his face, antero-inferior to the tragus of right ear since 4 years. He had past history of similar swelling 5 years back which was excised by a local practitioner without undergoing any investigation.

On physical examination, a mass of size 6x5 cm, firm to hard, non-mobile, non-tender was palpated over right parotid gland with no evidence of inflammation, discharge, facial nerve and lymph node involvement. Rest of the head and neck examination was unremarkable. Chest X-ray was normal and other laboratory investigations were within normal limit showing no evidence of metastasis.

After laboratory investigations patient was advised USG neck which revealed an irregular, heterogenous mass of 6.8x5.2 cm with no extra-parotid extension. Patient also underwent Fine Needle Aspiration Cytology from the swelling which revealed clusters of spindle cells suggesting spindle cell neoplasm.

Swelling was excised and sent for Histopathological examination. An irregular, creamish- white firm to hard mass measuring 7.0x6.3 cm was received in the department of Pathology, JNMCH AMU. The cut surface of the mass showed creamish-white areas with no areas of necrosis. On microscopic examination, H and E stained sections showed spindle cells arranged in storiform pattern. Spindle cells exhibit monomorphic ovoid nuclei, moderate to abundant eosinophilic cytoplasm (Figure 1-3). Some areas of normal salivary gland and fat entrapment were also seen along with staghorn-shaped blood vessels. No areas of necrosis noted.

On the basis of microscopic features, differential diagnosis of DFSP, solitary fibrous tumour, schwannoma, synovial sarcoma were made.

A panel of IHC markers consisting of vimentin, CD34, STAT-6 & S-100 was applied. Tumor showed vimentin & CD 34 positivity, while STAT-6 & S-100 were negative (Figure 4-7). Hence, the diagnosis of DFSP was confirmed and other possibilities were ruled out. Patient underwent total parotidectomy for the same with an uneventful post operative course.

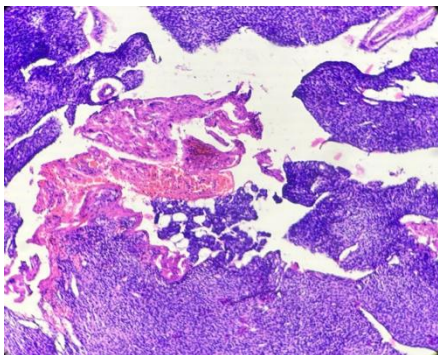
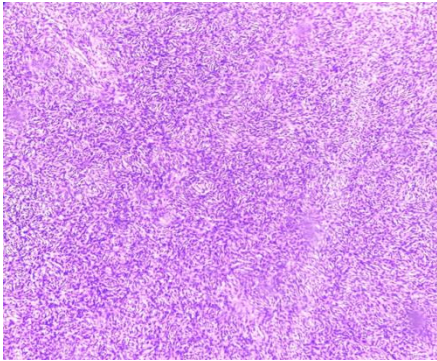
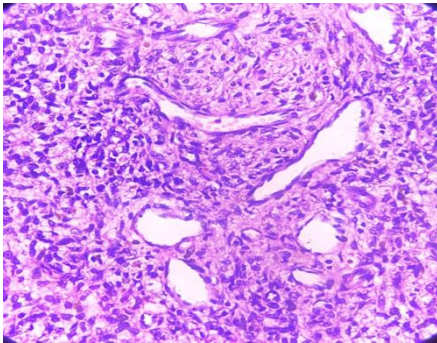


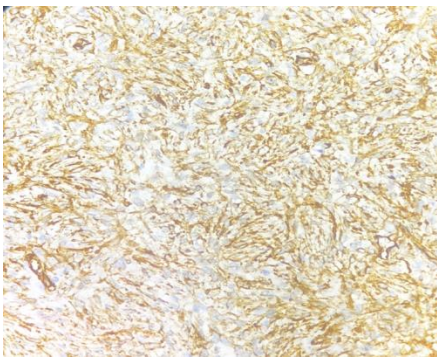
Figure 1 100X-Spindle cells along with area showing normal parotid



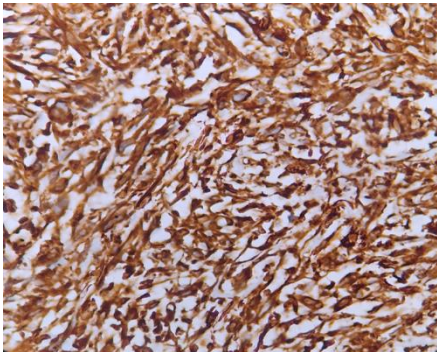
**Figure 2** 100X-Spindle cells arranged in storiform pattern



**Figure 3** 400X –Image shows spindle cells arranged in storiform pattern around staghorn blood vessels



**Figure 4** 100X-CD34 positive in spindle cells



**Figure 5** 400X- Vimentin –positive in spindle cells

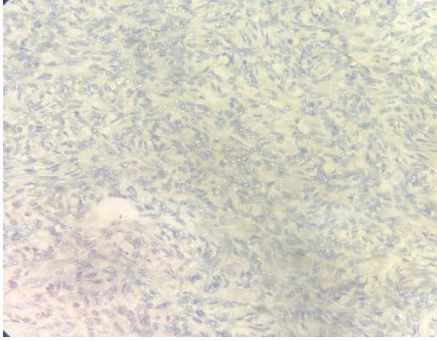


Figure 6 100X – Image shows S100- Negative in spindle cells

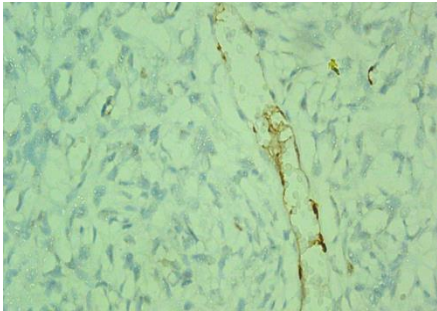


Figure 7 400X – Image shows STAT6-Negative in spindle cells

## DISCUSSION

Hoffman coined the term “dermatofibrosarcoma protuberance” and it was first described as a “progressive and recurring dermatofibroma” by Darier and Ferrand [5]. It commonly occurs in early life with a slight predilection for females. It has low to intermediate malignant potential, commonly presenting on the trunk and proximal extremities as a slow-growing, painless and firm mass [6]. “Protuberant” appearance can be appreciated in the developed lesion where cutaneous plaque develops into nodular appearance. However, unusual site for DFSP includes parotid gland. Histologically, the tumor cells are composed of uniform population of monomorphic spindle cells arranged in storiform pattern with mitotic figures not exceeding 5 per 10 HPF [7]. Cytogenetic studies of Dermatofibrosarcoma Protuberance shows two common translocations- 1) Reciprocal translocation,  $t(17;22)(q22;q13)$  2) Supernumerary ring chromosome derived from the translocation,  $r(17;22)$ . These translocations result in fusion of the collagen type IA1 gene (COL1A1) and the platelet derived growth factor B-chain gene (PDGFB) and hence chimeric COL1A1-PDGFB gene is formed, which plays an important role in the development of Dermatofibrosarcoma Protuberance [8].

In the differential diagnosis, solitary fibrous tumor, benign fibrous histiocytoma, and schwannoma should be considered. Solitary fibrous tumor is difficult to distinguish from Dermatofibrosarcoma Protuberance because of the similar histologic findings. It is characterized by a variety of growth pattern like mixture of cellular spindle cell areas with a distinct hemangiopericytic vasculature and myxoid changes while Dermatofibrosarcoma shows remarkable uniformity, a lack of the hemangiopericytic pattern and a distinct storiform pattern. Benign fibrous histiocytoma may show features similar to Dermatofibrosarcoma but it is usually negative for CD34. Schwannoma contains Antoni A and B areas, and it is S-100 positive which is not seen in Dermatofibrosarcoma [9].

Imaging findings for Dermatofibrosarcoma are nonspecific and is characterized by a subcutaneous well defined soft tissue mass on CT and MRI, showing intermediate to high enhancement on contrast enhanced studies. Histopathological examination along with immunohistochemistry showing positivity for CD34 and Vimentin may lead to final diagnosis [10].

The mainstay of treatment for Dermatofibrosarcoma is surgery with wide local excision with gross margins of 2 cm. Conventional chemotherapy also appears to offer little utility, however treatment with the molecular targeted therapy including Imatinib, has provided good results. Dermatofibrosarcoma is known to be a radiosensitive tumour. Radiation therapy may be recommended for patients if the margins of resection are positive or in case adequate wide excision is not possible due to cosmetic reasons.

### **CONCLUSION:**

In conclusion, Spindle cell lesion is quite a rare finding in the head and neck region and there is always a diagnostic dilemma as there are many different diagnosis encountered in the surrounding region. Therefore, other spindle cell tumors occurring in the salivary gland should be excluded before making a diagnosis of DFSP.

In the present case, the rarity of location of the lesion ruled out the possibility of considering DFSP in the clinical differential diagnosis. However, correlation of the clinical presentation with histopathological findings helped in arriving at the diagnosis. Using the immunohistochemical studies a definitive diagnosis of DFSP was established. The role of immunohistochemistry is very important to reach the final diagnosis in such cases.

### **Finanial Conflict of Interest:**

Nil

### **Sources of funding**

Nil

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