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# Spindle cell carcinoma of the parotid gland - a rare case report

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**Abstract :** Spindle cell lesions of the head and neck, offers a dizzying array of pathologies ranging from benign reactive lesions to malignant highly aggressive lesions. Spindle cell carcinoma is a variant of squamous cell carcinoma with particularly aggressive outcome. Spindle cell carcinoma of the mucosal lining of the oral cavity and the aerodigestive tract though not a rare entity, the occurrence of this particular histology in the salivary glands is very rare with very few similar cases published in the literature so far. Due to its rarity, no strict guidelines that would aid in their management have not been established. Here we are presenting a rare case of spindle cell carcinoma of the parotid gland, elaborating on the diagnosis and the management of these rare tumours.

**Keyword :**Spindle cell carcinoma , Squamous cell carcinoma , Sarcomatoid carcinoma , surgery , radiotherapy

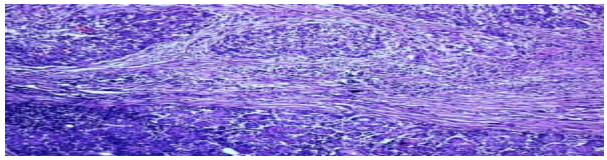
Introduction :

Salivary gland neoplasms are a common class of malignancies that present frequently to the operating surgeon. Very few malignancies in the body have a plethora of histological diversities like the salivary gland. Here we are presenting a rare case of spindle cell carcinoma of the parotid gland. Though many such cases have been reported in the oral cavity, this is only the third case of spindle cell carcinoma involving the parotid gland, after extensive search of the medical literature.

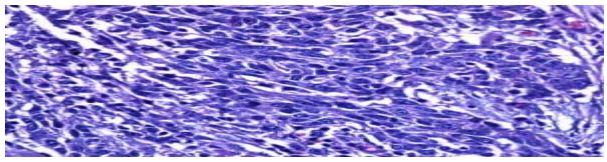
Case history:

A 49 year old gentleman presented with a swelling in the region of the right parotid of 4 months duration. Clinically the patient had a 2x2 cm hard mobile swelling, arising from the superficial lobe of the parotid. Fine needle aspiration cytology was suggestive of malignant neoplasm with cells showing spindled appearance. CT scan revealed a small nodular lesion in the superior lobe of the right parotid gland without any deep lobe involvement. No obvious nodal involvement was also made out. The patient underwent right total parotidectomy taking care to preserve the branches of the facial nerve. The post operative period of the patient was uneventful except for minimal facial paresis which improved subsequently.

The biopsy of the parotidectomy specimen was reported as a 1.2x1 cm nodule with features suggestive of malignant spindle cell neoplasm. Hematoxylin and Eosin staining revealed irregular atypical spindled cells widely distributed in a loose stroma of epithelial cells.(Fig 1,2 )



**H and E image of the slide at 10x**



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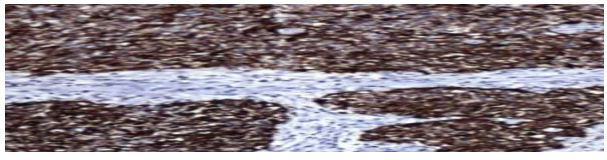
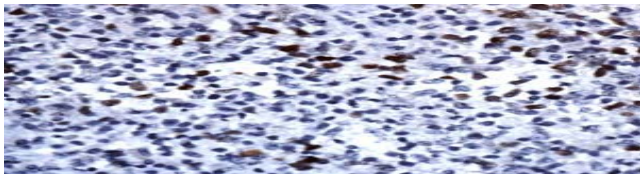
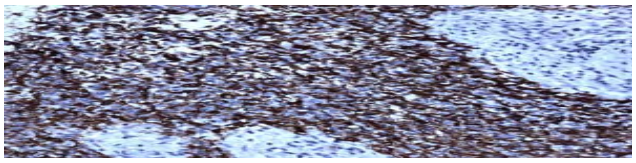
Immunohistochemistry revealed strong positivity for keratin (Fig 3). Vimentin, Smooth muscle actin and calponin were negative. Positivity for TTF1(Fig 4) and S100P was also established. Epithelial cells showed strong positivity for High molecular weight cytokeratin(HMWK, Fig.5). The proliferation marker Ki 67 was 60% positive. Markers for neuroendocrine differentiation namely, Neuron specific enolase , chromogranin , synaptophysin were negative. The final immunohistochemistry report was given as spindle cell carcinoma of the parotid, high grade.



**IHC showing keratin positivity**



**IHC showing TTF 1 positivity**



**IHC showing HMWK positivity**

A PET CT done subsequently revealed no other areas of metastasis. The patient then received post operative irradiation of 60Gy to the locoregional site. The patient has since then been on follow-up and has had no evidence of any recurrences so far.

Discussion:

Spindle cell carcinoma are rare variants of squamous cell carcinoma and constitute about 3% of squamous cell carcinomatous tumours arising from the head and neck region [1, 2]. Many terminonlogies like carcinosarcoma , pseudosarcoma , Lane's tumour , sarcomatoid carcinoma, pleomorphic sarcoma etc. have been used for this fascinating malignancy. The term spindle cell carcinoma was first used by Shervin et al. The cell of origin of the sarcomatoid tumours have been a matter of debate, though in recent times the conventional squamous carcinoma and the spindle cell component have been postulated to arises from a single stem cell [3].

Various other theories of varying ideas have been proposed to highlight the histogenetic nature of the spindle cells. One theory explains that both the spindle cells and epithelial cells arise simultaneously from separate plueripotent cells, thus rightly deserving the name as "collision" tumor. The second theory suggests, thant the tumour may have an inflammatory origin and the spindle cell component of the tumour may be a part of reactive proliferation of the inflammatory stroma and hence the name of "pseudosarcoma". Certain proponents also suggest that the spindling of cells may indicate a certain degree of dedifferentiation indication progression of the conventional squamous cell carcinomatous component. The epithelial cells are thought to undergo a series of phenotypic changes thereby acquiring a pathway akin to mesenchymal differentiation, undergoing spindling of the cells, producing mesenchymal matrix components, losing keratin expression and gaining the expression of vimentin. Loss of cohesion, invasion of the stroma by production of matrix metalloproteinase and subsequent metastasis occur like any other malignancy [4].

The outcome of the patients with spindle cell carcinoma is thought to be mainly decided by the stage and the location of the tumour, much akin to their squamous cell carcinoma counterparts [5].

The differential diagnosis for spindle cell carcinoma are a myriad of tumours, some of which may include fibromatosis, fibrosarcoma, malignant fibrous histiocytoma, leiomyoma, leiomyosarcoma, malignant peripheral nerve sheath tumour, chondrosarcoma, mesenchymal chondrosarcoma, angiosarcoma, Kaposi's sarcoma,malignant melanoma and synovial sarcoma just to highlight a few.

Immunohistiochemistry comes to our aid to a certain extent in narrowing down on the diagnosis. Spindle cell carcinoma is usually positive for both epithelial and mesenchymal markers namely keratin and vimentin. S100P is said to positive 70% of the cases, as is HMWK as demonstrated in our patient. Ki67 a marker of proliferation was 60-70%, indicating the high grade of the tumour involved. TTF1 which is a marker for thyroid and lung malignancy was also positive. .

PET CT done to clarify our doubts, confirmed no uptake in the region of the thyroid gland or the lungs, indicating the primary site of origin as the parotid based on the location of the tumourThe largest series of spindle cell sarcomas (187 patients) has been reported by Thompson et al mainly focussing on larynx as the primary[2]. Another large series from this part of the world (103 cases) which includes the entire region of the head and neck has also been published[6]. Both these large series have not mentioned about the tumour arising from the parotid gland, indicating the rarity of the tumour.

Ishibashi et al. published a case of spindle cell carcinoma of the parotid, who after radical surgery, recurred and died within 11 months[7]. This indicates the need for adjuvant therapy in the form of radiotherapy to decrease the incidences of recurrence. Another article by Huang et al, published a case report which highlighted the metastatic potential of spindle cell carcinoma of the parotid in spreading to the adjacent cervical lymph nodes, which further strengthens the need for prompt locoregional therapy immediately after surgery to maintain a prolonged disease free survival[8].

A thorough search in literature revealed only two published cases of spindle cell carcinoma of the parotid gland so far, this being the third case. This case has been presented for its rarity and the multidisciplinary approach which helped us render optimal treatment to the patient.

Conclusion:

A large array of rare histopathological entities continue to be and published in literature, adding up to our ever expanding conglomeration of malignancies. Immunohistochemistry has come a long way and has helped to a large extent in narrowing down on the diagnosis. Though many tumours may continue to be diagnosis, the basic principles of diagnosis and management holds good, and must be followed strictly for better treatment of our patients.

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