



## Case Report

# Spindle cell carcinoma of the parotid- A rare case entity with review of literature

Kafil Akhtar<sup>1,\*</sup>, Sumbul Warsi<sup>1</sup>, Fauzia Talat<sup>1</sup>, Mohd Talha<sup>1</sup>

<sup>1</sup>Dept. of Pathology, Jawaharlal Nehru Medical College, Faculty of Medicine, Aligarh Muslim University, Aligarh, Uttar Pradesh, India



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### ABSTRACT

Spindle cell carcinoma of the salivary gland is very rare. Spindle cell carcinoma is a variant of squamous cell carcinoma with an aggressive course. We present a rare case of spindle cell carcinoma of the parotid in a 54-year-old male, who presented with a rapidly enlarging mass in the right parotid gland. Histopathologically, the tumour was composed of foci of squamous cell carcinoma with sheets of malignant squamoid cells and spindle cell sarcoma with scattered pleomorphic, plump to spindle-shaped cells with foci of bone formation. Immunohistochemical studies revealed cytokeratin positivity and vimentin negativity in the squamous cell carcinoma component with focal positivity for epithelial membrane antigen and negative expression of cytokeratin and vimentin positivity in the sarcomatoid component. A final diagnosis of primary spindle cell carcinoma of the parotid gland was given. Our patient is alive and well with no clinical or radiological evidence of recurrence or metastatic disease after 12 months of follow up period.

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## 1. Introduction

Parotid malignancies comprise 1.0% to 3.0% of all head and neck cancers, while sarcomas comprise only 0.3% to 1.5% of all major salivary gland neoplasms.<sup>1,2</sup> Spindle cell carcinoma are rare variants of squamous cell carcinoma and constitute about 3.0% of squamous cell carcinomas arising from the head and neck region.<sup>1,2</sup> Though Rudolf Virchow first described sarcomatoid carcinoma, the term spindle cell carcinoma was first used by Shervin et al.<sup>2,3</sup>

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 arise from a single stem cell.<sup>3</sup> Spindle cell carcinoma is considered to be a monoclonal epithelial neoplasm with the sarcomatous component derived from squamous epithelium with divergent mesenchymal differentiation.<sup>2</sup> We describe an unusual parotid spindle cell carcinoma with review of

literature of this rare lesion.

## 2. Case Summary

A 54-year-old man presented to the ENT clinics with a 6-month history of swelling in the right parotid gland region. The patient had a history of smoking and alcoholism for 30 years. The oropharyngeal examination was normal with no other mass lesion in his neck and no facial nerve dysfunction. Chest radiographs, complete blood counts and serum chemistry were within normal limits. Computed tomographic scan of the neck revealed a 4 x3 cm, irregular hypoechoic mass in the superior lobe of the right parotid gland without any deep lobe involvement. No obvious nodal involvement was also made out.

Fine-needle aspiration of the mass was performed which showed dyscohesive clusters of pleomorphic, round to oval to plump cells admixed with atypical polygonal cells (Figure 1). A right total parotidectomy with right upper node neck dissection was performed taking care to preserve the branches of the facial nerve. The post operative period of

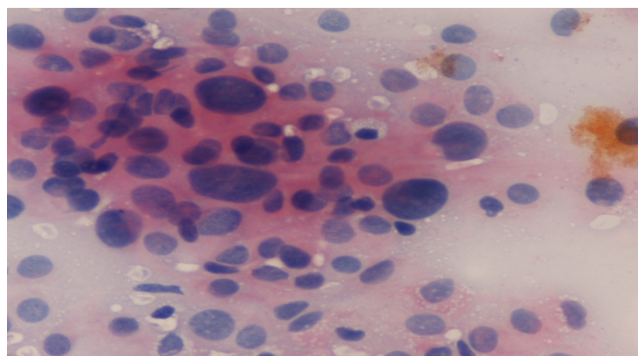
\* Corresponding author.

E-mail address: [drkafilakhtar@gmail.com](mailto:drkafilakhtar@gmail.com) (K. Akhtar).

the patient was uneventful except for minimal facial paresis which improved subsequently.

The resected specimen measured 8x5x3 cms and weighed 60g. Cut sections revealed an ill-defined, tan, firm lesion measuring 4x3x2 cm with indistinct lobules. Microscopically the tumor showed solid sheets of tumor cells completely effacing the normal parotid gland. The individual tumor cells were plump to spindle shaped with pleomorphic nuclei with coarse clumped chromatin, prominent nucleoli and abundant ill defined eosinophilic cytoplasm (Figure 2). Foci of sheets of atypical polygonal squamous cells were also seen. Mitotic activity of 4/10 high-power field was noted with no vascular or lymphatic invasion. The pleomorphic spindle tumor cells showed cytoplasmic positivity for vimentin (Figure 3). Focal cytoplasmic cytokeratin positivity was seen in the foci of squamous cell carcinoma (Figure 4). Based on the cytomorphology and immunohistochemical features a diagnosis of Spindle Cell Carcinoma of the Parotid was given.

The patient was given adjuvant therapy with 50 Gy of Co-60 teletherapy and six cycles of combination chemotherapy of Vincristine 50 mg/m<sup>2</sup>, Adriamycin 25mg/m<sup>2</sup> and Cisplatinum 50 mg/m<sup>2</sup>. Our patient is alive and well with no clinical or radiological evidence of recurrence or metastatic disease after 12 months of follow up period.

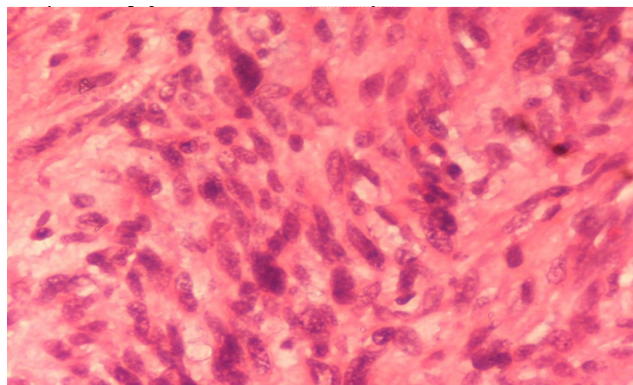


**Fig. 1:** Fine-needle aspiration showed dyscohesive clusters of pleomorphic, round to oval to plump cells admixed with atypical polygonal cells. Hematoxylin and Eosin x 40 X.

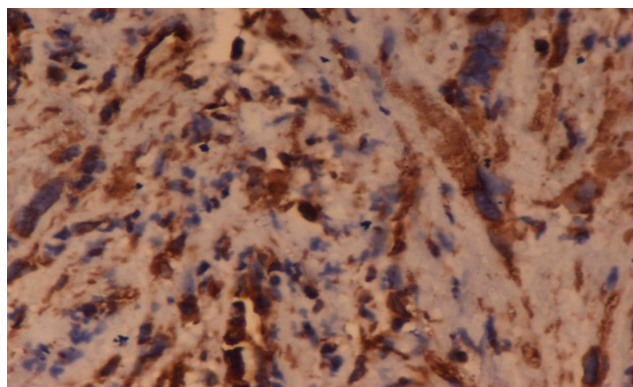
### 3. Discussion

Spindle cell carcinoma of the salivary gland is an extremely rare tumor.<sup>4,5</sup> In this biphasic tumor, the epithelial and mesenchymal components are admixed with each other, as seen in our case. These tumors mostly involve the parotid glands followed by submandibular glands and the hard palate.<sup>6</sup>

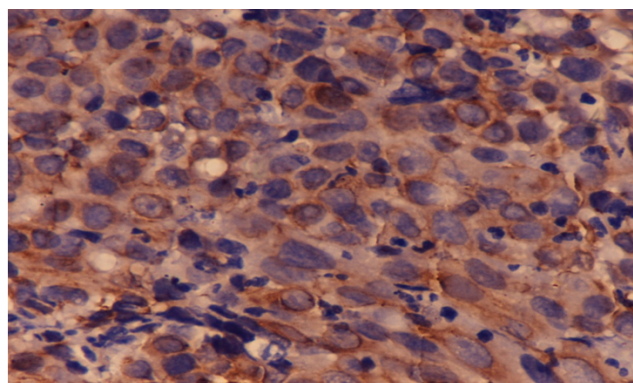
It is an aggressive, high-grade neoplasm with frequent metastasis.<sup>1</sup> It constitutes approximately 12.0% of



**Fig. 2:** Microscopically the mesenchymal component was seen as individual tumor cells, plump to spindle shaped with abundant ill defined eosinophilic cytoplasm, pleomorphic nuclei and prominent nucleoli. Mitotic activity of 4/10 high-power field was noted. Hematoxylin and Eosin x 40 X.



**Fig. 3:** The pleomorphic spindle tumor cells showed diffuse cytoplasmic positivity for vimentin. IHC Vimentin x 40X.



**Fig. 4:** Diffuse moderate cytoplasmic cytokeratin positivity was seen in the foci of squamous cell carcinoma. IHC Cytokeratin x 40X.

malignant salivary gland tumors.<sup>1,7</sup> Usually seen in the 6<sup>th</sup> and 7<sup>th</sup> decades of life with a male to female ratio of 12:1.<sup>4</sup> Several theories show a multifactorial causation of the disease with a strong association with cigarette smoking, alcohol abuse and ionizing radiation with the radiation risk complicated by the dose and duration of exposure.<sup>3</sup> Most of the cases are seen to arise in a pre-existing benign mixed tumor.<sup>6</sup> Volker et al put forward the hypothesis of common stem cell monoclonal origin of spindle cell carcinoma that could be the myoepithelial cell and an inactivated tumor suppressor gene on chromosome 17 other than p53.<sup>5</sup>

The epithelial components can be ductal adenocarcinoma or squamous cell carcinoma whereas the most common mesenchymal component is chondrosarcoma. Our case showed admixture of leiomyosarcoma and squamous cell carcinoma. Malignant spindle cell tumors within salivary glands are a diagnostic challenge. The differential diagnosis include pure carcinoma, spindle cell carcinoma, malignant fibrous histiocytoma (MFH), malignant melanoma, fibrosarcoma, leiomyosarcoma, osteosarcoma, liposarcoma and rhabdomyosarcoma.<sup>8,9</sup> Immunohistochemistry is instrumental in narrowing the differential diagnosis.<sup>10,11</sup>

Sarcomas such as rhabdomyosarcoma or neurosarcoma show positivity for S-100 and HMB-45 with characteristic features on electron microscopy.<sup>10,11</sup> Fibrosarcoma has the typical herring bone histologic appearance and indistinct rough endoplasmic reticulum on ultra structural study. The possibility of an osteosarcoma could be entertained owing to the presence of bone formation with lacelike osteoid with confirmation by osteonectin expression. Malignant fibrous histiocytoma can occur in the deep soft tissues and in salivary glands with microscopic features of cellular pleomorphism and a focal storiform pattern of growth.<sup>12,13</sup> Since cellular pleomorphism was moderate and a storiform pattern was not prominent in our case, the overall features were felt to be inconsistent with MFH. Furthermore, spindle cell carcinomas usually have a glandular component with positivity for epithelial markers in the neoplastic cells.

Ishibashi reported a case of spindle cell carcinoma of the parotid, who had fatal recurrence after radical surgery.<sup>14</sup> This indicates the need for adjuvant therapy in the form of radiotherapy and/or chemotherapy to decrease the incidence of recurrence. Huang et al highlighted the metastatic potential of spindle cell carcinoma of the parotid to the adjacent cervical lymph nodes, which further emphasizes the need for prompt postoperative loco-regional adjuvant therapy.<sup>15</sup> 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 similar to their squamous cell carcinoma counterparts.<sup>16</sup> Better understanding of the nature of sarcomatoid carcinoma could help in the selection of appropriate therapies to hinder its progression.<sup>14</sup>

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#### 5. Conflict of Interest

The authors declare they have no conflict of interest.

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#### Author biography

**Kafil Akhtar**, Professor

**Sumbul Warsi**, Resident

**Fauzia Talat**, Resident

**Mohd Talha**, Senior Resident

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