

# Spindle Cell Carcinoma of the Maxilla: A Case Report of Rare Entity

Montasir Junaid<sup>1\*</sup>, Maliha Kazi<sup>2</sup>, Sadaf Qadeer<sup>3</sup>

<sup>1</sup>Department of Otorhinolaryngology-Head and Neck Surgery, Jinnah Medical and Dental College, Karachi, Pakistan

<sup>2</sup>Department of Otorhinolaryngology-Head and Neck Surgery, Royal Manchester Infirmary, Manchester, UK

<sup>3</sup>Department of Otorhinolaryngology-Head and Neck Surgery, Sir Syed College of Medical Sciences, Karachi, Pakistan

Email: \*montsj@gmail.com

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## Abstract

Spindle cell carcinoma also known as sarcomatoid carcinoma is a rare highly aggressive tumor which is histologically different from squamous cell ca and mesenchymal cancers. Only hands full of cases have been reported in literature since 1957 and hence no proper treatment protocol has been devised yet. We present such a case of thirty-four-year-old female who presented with spindle cell carcinoma of the maxilla at our department and was treated with extensive surgery followed by chemotherapy and radiation therapy. Spindle cell carcinoma is generally associated with poor prognosis and hence literature supports use of post operative Chemo and Radiotherapy for better result and decrease chance of local recurrence.

## Keywords

Spindle Cell Carcinoma, Maxilla, Maxillary Carcinoma

## 1. Introduction

Spindle cell carcinomas are considered to be highly aggressive, biphasic malignant tumor that occurs rarely in head and neck region [1]. Histologically, these tumors consist of malignant epithelial and malignant mesenchyma components [2]. The epithelial component is a squamous cell carcinoma admixed with a malignant sarcomatous component that is either non-descript or contains heterologous elements (*i.e.* osteo or chondrosarcomatous) [1]. In the past, these tumors have been given variety of names including sarcomatoid carcinoma, carcinosarcoma, pseudosarcoma, collision tumor, and pleomorphic carcinoma [3].

Spindle cell carcinoma is considered to be a poorly differentiated squamous cell carcinoma, with elongated epithelial cells that resemble sarcoma [2]. Thus

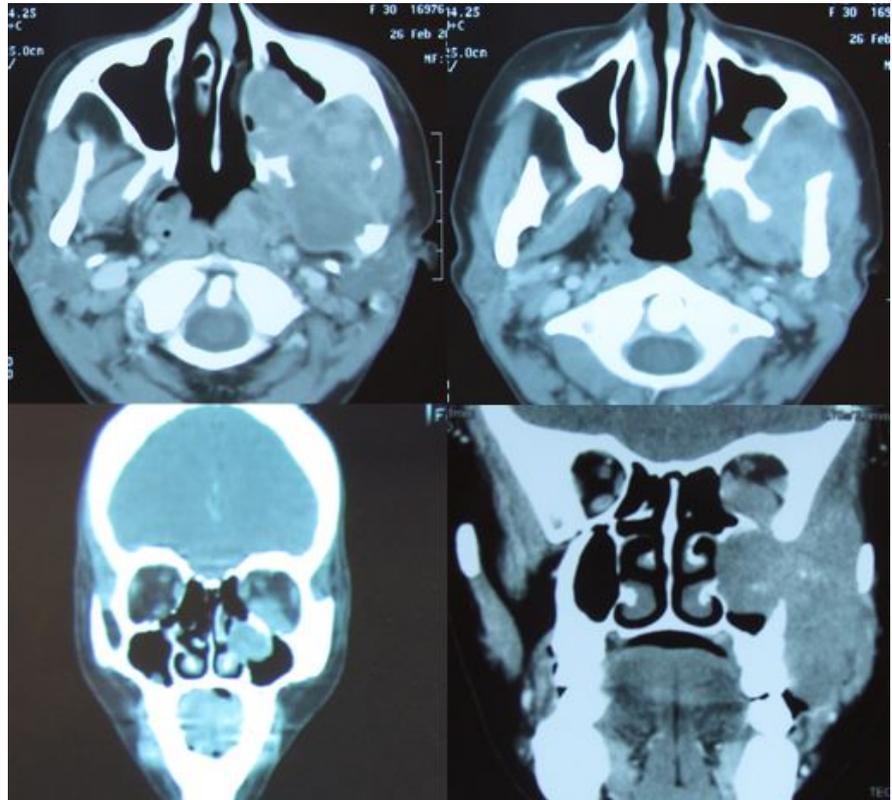
regardless of having a mesenchymal appearance, it has an epithelial origin and derived from squamous cells which can be confirmed by use of electron microscopy and immune-histochemical stains for keratins [4] [5]. Despite its rarity in the head and neck region, the most frequently involved site is the larynx, with reports of cases found in nasal cavity, hypopharynx, oral cavity, esophagus, trachea, maxilla and skin also present in the literature [6]. Spindle cell carcinoma arising in maxilla is much rarer occurrence in literature [7] with few cases reported since 1957 (**Table 1**).

We report here a case of Spindle cell carcinoma arising from the maxilla in a patient who was treated at our institute.

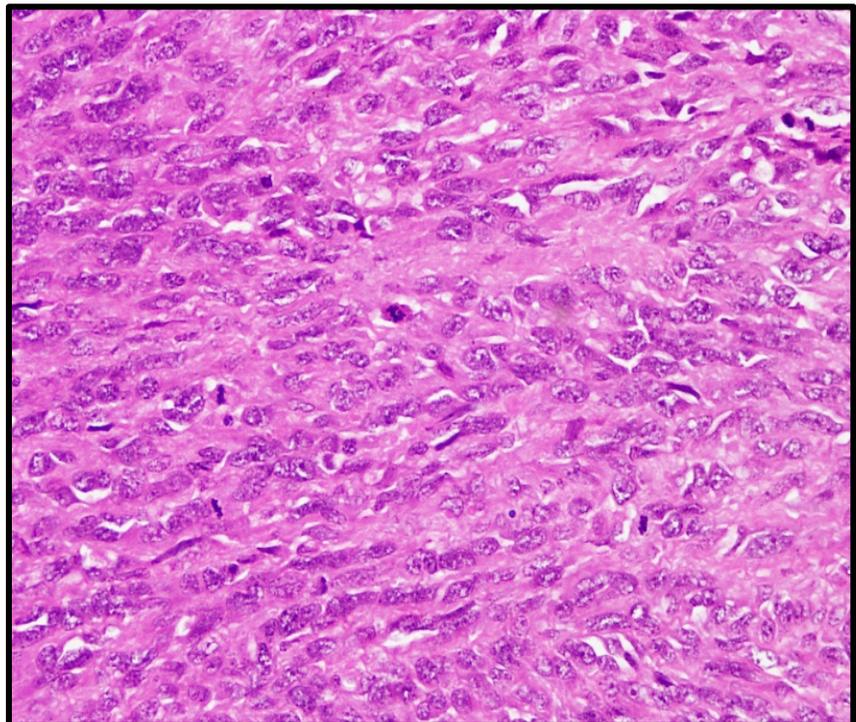
## 2. Case Presentation

A 34-years-old female, presented in ENT clinic with complaints of left nasal blockage which progressed over 4 months associated with diffuse swelling, pain and hypoesthesia of left cheek. There was left Eye proptosis and decrease vision for 6 months accompanied with loss of left upper molar and premolar tooth. Patient's past surgical or medical history was not significant for any pathology and no history of addiction to tobacco or other drugs was identified. On examination, there was a non-tender left sided facial swelling which was firm to touch and non-indurated. On anterior Rhinoscopy Nasal cavity failed to show any mass/lesion in the left side while rigid nasopharyngeal scope was also normal. There was loss of left upper molar teeth on left side with granulation tissue which bled on manipulation. The left eye had obvious proptosis with no light perception and no ocular movements while there was no light or pupillary reflex as well. Rest of ENT and neck examination was unremarkable. Patient was advised Routine labs and C.T head and neck, chest with contrast. After obtaining a computed tomographic image, biopsy was taken from left upper maxillary granulation tissue. **Figure 1** C.T scan showed the lesion to be destructive in nature, occupying the whole left maxilla but surprisingly sparing left nasal cavity. Superiorly tumor extended and eroded the orbital floor in antero-posterior fashion making its way in to orbital cavity proper and apex, while laterally extending to temporal and infra-temporal fossa, with posterior extension into pterygoid plates. The histology confirmed the presence of atypical cells with mitotic figures. The tissue was further reviewed under immuno-histochemical stains and the diagnosis of spindle cell carcinoma was confirmed. **Figure 2 & Figure 3** patient underwent metastatic work-up which was negative and no other systemic diseases were identified.

After counseling, detail discussion and consent; patient underwent left subtotal maxillectomy with orbital exentration while the pterygoid muscle and plates were found to be devoid of disease and temporal fossa extension of tumour was cleared via tunnel created through lateral wall of orbit. Surgery was followed by 2 cycles of cisplatin (30 mg/m<sup>2</sup>) of chemotherapy and intensity-modulated radiation therapy (IMRT) over the left maxilla for a total of 70 Gy in 35 fractions. Patient remained free of any local or regional recurrence since as

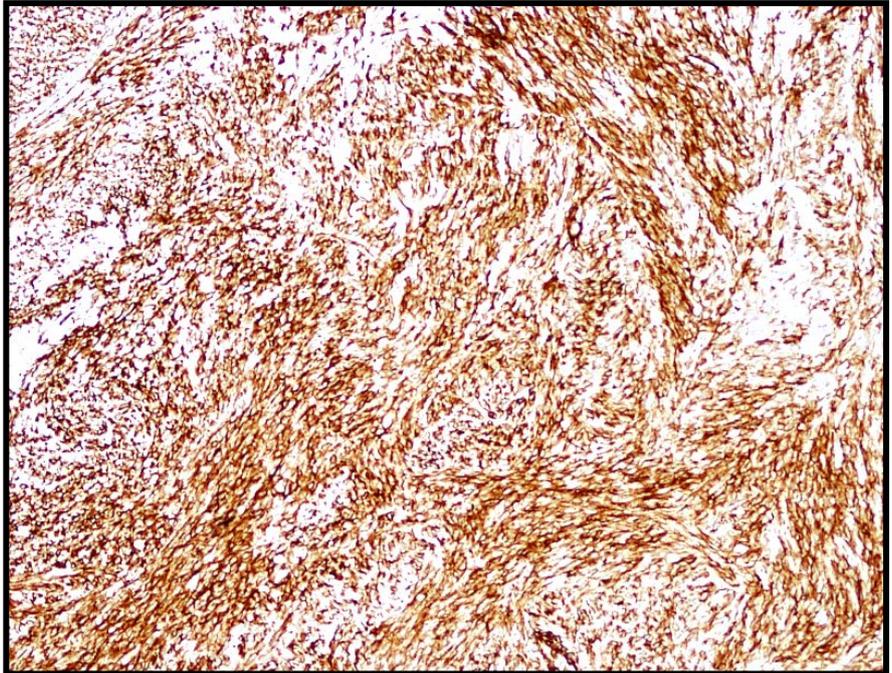


**Figure 1.** C.T Scan (Axial and Coronal Views) showing the extensive lesion.

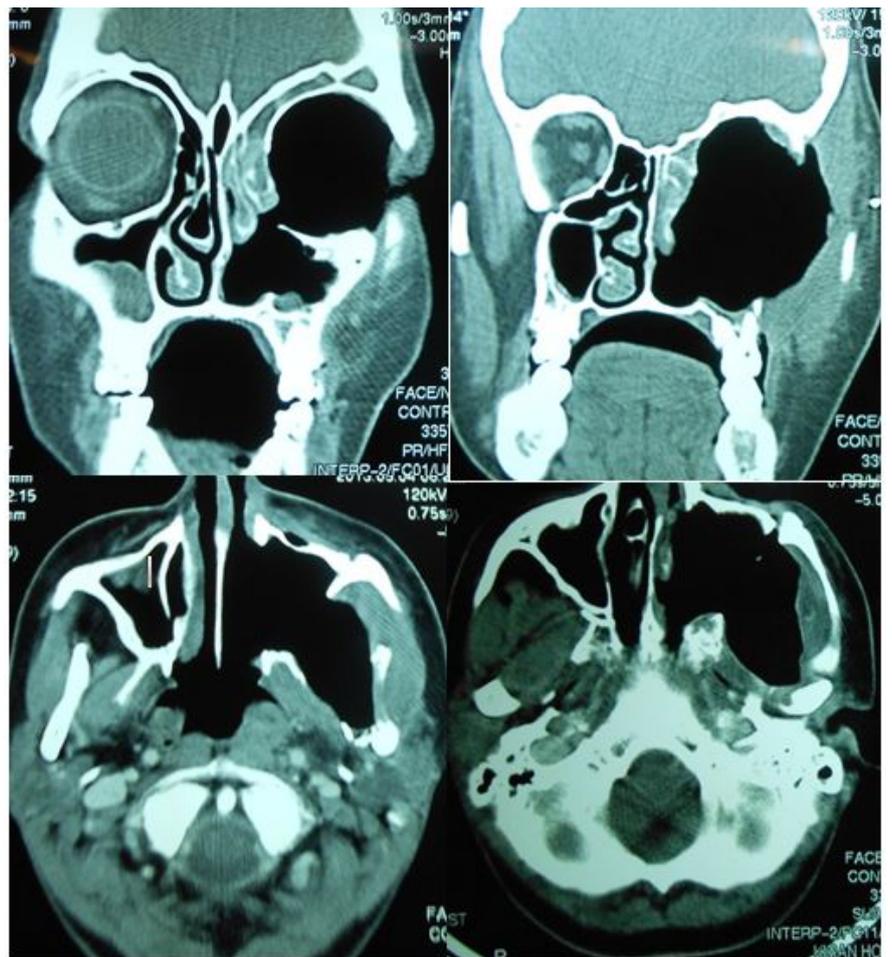


**Figure 2.** Medium power view showing fascicles and bundles of spindle cells.

reviewed on regular follow up visits and C.T scan at one year post surgery **Figure 4.**



**Figure 3.** Positive staining of smooth muscle actin indicating spindle cell carcinoma.



**Figure 4.** Follow-up scan after 1 year (Coronal and Axial views).

**Table 1.** List of published articles on spindle cell carcinoma of maxilla with treatment.

No.	Age in years	Gender	Treatment	Year of publication	Author
1.	62	Female	Radiotherapy	1957	Meyer
2.	62	Female	Radiotherapy	1970	Lichtiger
3.	71	Male	Radiotherapy + Total maxillectomy + Orbital exentration	1982	Feinmesser
4.	65	Female	Total maxillectomy + Radiotherapy	1985	Ampil
5.	57	Male	Wide Excision of tumor	1987	Hafiz
6.	60	Male	Total maxillectomy + Radiationtherapy + Chemotherapy	1989	Sonobe
7.	53	Male	Craniofacial resection + total Maxillectomy + Radiationtherapy + Chemotherapy	1990	Shindo
8.	80	Female	Total Maxillectomy + Radiationtherapy + Revision surgery	1998	Sanabre
9.	47	Male	Partial Maxillectomy + radiationtherapy	2001	Furuta
10.	54	Male	Radiationtherapy + Chemotherapy	2007	Howard
11.	25	Male	Refused Surgery + Radiation therapy only	2008	Kumar
12.	60	Male	Total Maxillectomy + Radiationtherapy + Chemotherapy	2009	Jeong-Ki Moon
13.	52	Male	Total Maxillectomy + Radiationtherapy + Chemotherapy	2012	Alem
14.	60	Female	Partial Maxillectomy + Supra-Omohyoid Neck Dissection	2013	Ravindran
15.	52	Female	No Treatment/Lost to follow-up	2013	Samuel
16.	61	Male	Total Maxillectomy + Modified Radical neck dissection	2014	Cheong
17.	40	Male	Partial Maxillectomy + Level I and II neck Dissection	2014	Rath
18.	34	Female	Partial Maxillectomy + Orbital Exentration + Chemotherapy + Radiotherapy	2015	Our case

### 3. Discussion

Sarcomatoid carcinoma is a rare biphasic malignant tumor, and is reported to account for less than 1% of all tumors within the oral cavity [8]. Its development is considered to be multi-factorial with poor oral hygiene, alcohol or tobacco intake and previous radiation considered as risk factors. Genetic predisposition has also been reported in some studies [4]. With a male predominance, it generally occurs in the sixth decade of life, with cases in younger ages reported as well **Table 1.**

Bulk of the tumor is made up of spindle cells with remaining part consisting of the epithelial cells [8] [9]. Various theories have been put forward to explain the histological appearance of spindle cell carcinoma. Some researchers believe that both these components arise from separate stem cells, the so called collision tumor. While others favor the concept of spindle cells being atypical reactive proliferation of the stroma, despite all arguments, the most widely accepted

theory supports the monoclonal hypothesis that states that they arise from the same stem cells and have actually undergone “dedifferentiation” [9] [10]. Some considered the development of spindle cells being derived from the epithelial cells themselves with functional loss of genes responsible for epithelial differentiation [9].

There are several differential diagnoses for spindle cell carcinoma and these include leiomyosarcoma, rhabdomyosarcoma, myoepithelial carcinoma and malignant peripheral nerve sheath tumor. For diagnosis of spindle cell carcinomas, light microscopy gives initial important clue, and immuno-histochemical stains further help in the confirmation of the diagnosis. Epithelial differentiation and staining for one or more epithelial markers supports the diagnosis. In most cases (26% - 62%) Keratin and cytokeratin are commonly found to be positive. Furthermore, positivity to mesenchymal-type markers is also demonstrated. Almost 100% of the cases show positivity for vimentin and about a third of them for smooth muscle actin [11].

In literature the treatment of spindle cell sarcoma follows same footsteps as that of squamous cell carcinoma of similar stage with wide surgical excision being the preferred choice of treatment [Table 1]. Nevertheless, in comparison to squamous cell carcinoma, within the head and neck region, spindle cell carcinoma is considered as a more aggressive tumor and has a propensity to recur and metastasize early [12]. For tumors of the oral cavity as well as the larynx, surgery is the mainstay of treatment. In comparison to radiation therapy alone, the prognosis is better for surgery [13] [14]. Chemotherapy has proven to have good results as part of neo-adjuvant and adjuvant treatment in helping to reduce the tumor size. However, when used as a sole treatment modality, it has ambiguous results [15]. Treatment also depends on various other factors; like low grade tumor, small tumor size, absence of previous radiation therapy and polypoidal growth indicate good prognosis [14], while factors that influence choice of treatment clearly depends primarily upon the extent of lesion and surgeon being able to excise the lesion completely. This is followed by Radiation and +/- chemotherapy [15]. But no clear guideline is available in literature to support one treatment modality over the other. Sarcomatoid carcinoma is considered as a more aggressive tumor, with a tendency to recur and metastasize early when compared to squamous cell carcinoma within the head and neck region [11] [12].

The presence of distant metastasis and the depth of invasion of the tumor are considered to be reliable prognostic features for the disease [16]. Regional and distant metastasis does greatly alter the prognosis as well as the overall mortality rate. Cervical lymph node involvement is present in approximately 7.5% - 26% of the cases and an even lesser distant metastasis [17]. The most frequent site of distant metastasis is the lungs. Lambert *et al* in his study, reported the incidence of lung metastasis to be about 5% with the overall mortality rate reported in literature to be 14% - 32% [17] [18].

## 4. Conclusion

In conclusion, spindle cell carcinoma of maxilla is a rare entity in head and neck. It is a potentially aggressive tumor with high recurrence rate. Due to the fact that it has both epithelial and mesenchymal tissue, it poses a diagnostic challenge for the histopathologist. Immune markers and special stains have an important role to play in its differentiation with other tumors.

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