Sarcomatoid Carcinoma of Maxillary Sinus: Report of an Unusual Case and Review of Literature
Pankaj M Shirsat, Rajiv S Desai, Shivani Bansal, Mohan Deshpande

Abstract
Sarcomatoid carcinoma is an uncommon variant of squamous cell carcinoma. Sarcomatoid carcinoma of maxillary sinus constitutes a rare occurrence and follows an aggressive course with only 13 reported cases. This case report describes sarcomatoid carcinoma of maxillary sinus in a young adult with a review of literature.

Keywords: Sarcomatoid Carcinoma; Maxilla; Sinus; Immunohistochemistry; Squamous Cell.

Introduction
Sarcomatoid carcinoma is a biphasic tumor composed of a squamous cell carcinoma, either in-situ or invasive, and a malignant spindle cell component with a mesenchymal appearance, but of epithelial origin. Sarcomatoid carcinomas have been reported from diverse sites such as upper aerodigestive tract, esophagus, salivary glands, thyroid, thymus, lung, breast, gastrointestinal tract, hepatobiliary system, genitourinary tract and uterus. The head and neck subsites include larynx, pharynx, oral cavity and nasal area. Sarcomatoid carcinoma of maxillary sinus constitutes an uncommon occurrence and follows aggressive course with a high mortality rate.

Case Report
A 22- year-old apparently healthy male presented to the Department of Oral Pathology, with a chief complaint of asymptomatic swelling in right maxillary anterior region since one month. Extraoral examination revealed mild elevation of right ala of nose. No lymphadenopathy or draining sinuses were noted. Intraoral examination revealed exophytic growth in right labial sulcus in relation to the permanent maxillary right central incisor, lateral incisor and canine measuring about two centimeter in size, blackish in color, firm in consistency and slightly tender to palpation without associated tooth mobility (Fig 1a). Oral hygiene was fair. Orthopantomograph revealed an ill-defined radiolucency between the roots of permanent right maxillary lateral incisor and canine (Fig 1b). The patient's medical and family history was non-contributory. Social history revealed absence of tobacco or alcohol habits. A differential diagnosis of benign inflammatory lesions like pyogenic granuloma and peripheral giant cell granuloma and gingival malignancy were included in our differential diagnosis.

The incisional biopsy of the lesion was performed under local anesthesia. The histopathological examination showed spindle shaped cells in a fascicular pattern. The cell nuclei were elliptical and vesicular (Fig 1c). Mitotic figures were present in few cells. Variable number of rounded cells with abundant cytoplasm and round vesicular nuclei were scattered among spindle cells. Multinucleated atypical tumor giant cells were seen in few places. Immunohistochemical investigations of tumor cells showed positivity for CK (Clone MBF116, Dakocytomation) (Fig 1d) and focal positivity for EMA (Clone GP1.4, DBS) (Fig 1e). Based on immunohistochemical findings a diagnosis of sarcomatoid carcinoma was made.

The patient was referred to medical oncology centre for further treatment. where CT scan revealed a lesion in the right maxillary sinus (Fig 1f). The patient underwent partial maxillectomy on the right side with right radical neck dissection. Histopathology and immunohistochemistry of excised specimen confirmed the diagnosis of sarcomatoid carcinoma of right maxillary sinus. Postsurgery the patient underwent radiotherapy. Chemotherapy was also planned. Four months later there was a
recurrence in the maxillary anterior region. Patient succumbed to the disease eight months after his first clinical presentation.

<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>sex</th>
<th>Histopathologic appearance</th>
<th>Immunohist ochemistry</th>
<th>Treatment</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Leventon et al. 1981</td>
<td>81</td>
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<td>Squamous cell carcinoma</td>
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<td>Asbury et al 1995</td>
<td>37</td>
<td>F</td>
<td>Sarcomatoid Carcinoma</td>
<td>-</td>
<td>Surgery + Postoperative radiotherapy</td>
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<tr>
<td>Sadaba et al 2006</td>
<td>45</td>
<td>M</td>
<td>Sarcomatoid carcinoma + Orbital apex syndrome</td>
<td>Performed for confirmation</td>
<td>Chemoradiotherapy</td>
<td>8 months disease free survival. Died 4 months later</td>
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<tr>
<td>Howard et al 2007</td>
<td>54</td>
<td>M</td>
<td>Malignant spindle and epithelioid cells in a myxoid background . Foci of squamous cell differentiation in spindle cell carcinoma</td>
<td>Pan-cytokeratin positive</td>
<td>Concomittent Chemotherapy + Radiotherapy</td>
<td>Residual disease at 4 month follow up</td>
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<tr>
<td>Kumar et al 2008</td>
<td>25</td>
<td>M</td>
<td>Polygonal squamous cell component blending with sarcomatous component of tumor</td>
<td>Strong positivity for cytokeratin and vimentin</td>
<td>Radiotherapy</td>
<td>Alive at 6 months with disease</td>
</tr>
<tr>
<td>Vishwanathan S 2010</td>
<td>50</td>
<td></td>
<td>Epithelial differentiation was seen in (20%) maxillary lesions</td>
<td></td>
<td>Immunopositivity for epithelial markers identified in (75%) maxillary lesions</td>
<td>- No definite conclusion due to limited follow-up</td>
</tr>
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<td>Terada T 2011</td>
<td>60</td>
<td>M</td>
<td>Malignant spindle and round cells</td>
<td>Tumor cells positive for AE1/3, KL-1, CAM 5.2, CK 18, Vimentin, CD 68, p 53Ki-67, α1-antitrypsin and α1-antichymotrypsin</td>
<td>Chemoradiation</td>
<td>Died at 9 months</td>
</tr>
<tr>
<td>Shirsat et al 2011 (present case)</td>
<td>22</td>
<td>M</td>
<td>Malignant spindle cells</td>
<td>CK positive, EMA - focal expression</td>
<td>Right partial maxilectomy with right radical neck dissection + Radiotherapy</td>
<td>Died at 8 months</td>
</tr>
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Table 1: Reported cases of sarcomatoid carcinoma of maxillary sinus.

Discussion
Neoplasms with mixed epithelial and mesenchymal features have been recognized since the time of Virchow. Kromphecher is credited with proposing the theory that carcinoma cells can undergo sarcomatous transformation. The oral cavity and larynx are predominant sites for sarcomatoid carcinoma in the head and neck region. Sarcomatoid carcinoma of maxillary sinus is a rare tumor with only 13 cases reported in english literature (Table 1). The reported cases point towards male predilection with patients ranging from...
3rd to 7th decade\textsuperscript{2,5-9} as seen in our case. Although tobacco, alcohol abuse and previous irradiation are considered predisposing factors\textsuperscript{6-8} no habits were present in our case.

Sarcomatoid carcinoma of maxillary sinus presents as a swelling of the affected maxilla along with associated facial pain, numbness or feeling of fullness of the sinus but in the present case patient had asymptomatic swelling in maxillary labial region. In advanced stages nasal examination shows either deviation of nasal septum or a mass visible through the nostril or a fleshy mass filling the nasal cavity. Buccal or palatal expansion may be present. On perforation of cortical plates the tumor appearance is usually polypoid or exophytic with surface ulceration and necrosis\textsuperscript{2,5-7} as seen in our case. The typical tumor is rapidly growing, usually polypoid sometimes bulky mass. Despite the seemingly innocuous nature, maxillary sarcomatoid carcinoma presents at an advanced stage either T3 or T4 pursuing an aggressive course.\textsuperscript{2} Neck node involvement may be present which may be due to inflammatory changes or tumor necrosis.\textsuperscript{2,6} Imaging studies of the maxillary sinus reveals presence of mass (as seen in our case) or a lytic lesion encroaching the adjacent structures.\textsuperscript{2,6,7}

Figure 1: The Intraoral photograph of exophytic growth in right maxillary anterior region (a). The panoramic radiograph showing ill-defined radiolucency between roots of permanent maxillary right lateral incisor and canine (b). The photomicrographs showing spindle shaped cells in a fascicular pattern with elliptical and vesicular nuclei under H&E stain, with magnification at x400 (c), under IHC with cytokeratin positive tumor cells (d) and focally positive tumor cells for Epithelial membrane antigen with magnification at x100 (e). Axial computed tomography scan showing lesion in the right maxillary sinus (f).
Sarcomatoid carcinoma manifests a bimorphic histologic appearance as definable squamous cell carcinoma and dominant atypical stroma composed of fusiform cells. It shows an admixture of growth patterns categorized as fasciculated, myxomatous or streaming. The fasciculated pattern (as seen in the present case) is most common, composed of highly cellular groups of elongated bipolar cells in parallel, interwoven alignment. Predominant cells are mainly plump, elongated or round and may appear epithelioid. Cellular and nuclear pleomorphism may be notable. Sarcomatoid part exhibits pronounced cellular pleomorphism, marked cellularity, mitosis and epithelial character of cells. Some tumors show apparent transitional foci, others islands of carcinoma surrounded by spindle cells.

Immunohistochemical demonstration of pan CK and EMA (as employed in our case) was found to be most useful practically. The epithelial marker expression decreases as the degree of epithelial differentiation decreases and may be lost entirely, hence a negative result does not rule out the diagnosis of sarcomatoid carcinoma. Low immunopositivity may be due to various factors like subjectivity in interpretation, sampling error, nonhomogeneous tumors, poor preservation or fixation, inappropriate antibody or technique, or epithelial features less than threshold of detection. Both the sarcomatoid as well as conventional squamous carcinoma components have now been proven to arise monoclonally from a single stem cell with further evidence to prove that the sarcomatoid component represents a dedifferentiation and suggests molecular progression of the conventional component.

The epithelial cells go through a spectrum of progressive phenotypic changes, acquiring a mesenchymal pathway of differentiation metamorphosing to a spindle shape and gaining vimentin while losing keratin expression. The phenotypic plasticity of interconversion of epithelium to mesenchyme cells is expressed by a loss of intercellular cohesion, elongation of the cells, loss of basement membrane, production of mesenchymal matrix components and stromal invasion. Wide surgical excision alone or with radical neck dissection (as done in the present case along with partial maxillectomy) seems to be most successful therapeutic modality. The clinical course appears roughly to parallel that of ordinary squamous cell carcinoma, with disease progression characterized predominantly by local recurrences (as seen in our case) and regional metastases. Clinically, distant metastases and histomorphologically depth of tumor invasion into underlying structures were found to be the only reliable prognostic indicators. The prognosis still remains poor due to lack of proper evidence based treatment options as the patient died eight months after first presentation.

Sarcomatoid carcinoma follows aggressive clinical course and is defined by malignant histopathologic features requiring immunohistochemistry for diagnosis. It is important to be aware of this type of neoplasm to ensure early detection and develop appropriate clinical management strategies by further studies.

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