Case Report

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Spindle Cell Carcinoma of Maxilla Mimicking Pyogenic Granuloma–A Rare Entity

Akhilanand Chaurasia

Abstract

Spindle cell carcinoma is a rare malignancy of the head and neck region. It is a biphasic neoplasm consisting of epithelial and mesenchymal components and accounts for less than 1% of all tumours of orofacial region. It is a unique variant of squamous cell carcinoma which is very aggressive in nature and associated with relatively poor prognosis. The most common site for occurrence is upper aerodigestive tract, esophagus, salivary glands, thyroid, thymus, lung, breast, gastrointestinal tract, hepatobiliary system, genitourinary tract and uterus. Involvement of maxillary sinus by spindle cell carcinoma is a rare occurrence and follows aggressive course with a high mortality rate. We report a rare case of spindle cell carcinoma of maxilla with immunohistochemical studies. We present a case of spindle cell carcinoma involving right maxillary antrum.

Keywords: Sarcomatoid Carcinoma; Spindle Cell Carcinoma; Squamous Cell Carcinoma; Biphasic Keratin; Vimentin.

Introduction

Spindle cell carcinoma is also known as sarcomatoid carcinoma. It is a rare malignancy of the head and neck regions and a rare variant of squamous cell carcinoma. It occurs mainly in upper aerodigestive tract involving larynx, thyroid, nasal cavity, hypopharynx, oral cavity, esophagus, trachea, lung, hepatobilliary system, genitourinary tract,uterus, skin and breast [1]. It is a unique variant of squamous cell carcinoma consisting of sarcomatoid proliferation of plemorphic spindle-shaped cells with a biphasic appearance presenting as a part of frank squamous cell carcinoma [2, 3]. According to WHO's classification of tumours, this tumour is placed under malignant epithelial tumours of squamous cell carcinoma and named as spindle cell carcinoma [4]. On the basis of histopathological variations of this rare entity several terminologies has been used to describe this tumour such as carcinosarcoma, Lane tumour, pseudosarcoma, pleomorphic carcinoma, sarcomatoid squamous cell carcinoma and polypoid carcinoma [2]. Many of these tumours can be easily confused with true sarcomas unless immunohistological analysis is performed. These analyses reveal concurrent presence of malignant epithelial and homologous sarcomatoid spindle cell components by co-expression of cytokeratin, epithelial membrane antigen and vimentin to various degrees [5, 6]. The clinical course follows an aggressive course with high incidence of metastases [2].

Author's Affilation: Assistant Professor, Department of Oral medicine & Radiology, Faculty of Dental Sciences, King George Medical University, Lucknow-226003 Uttar-Pradesh (India).

Reprints Requests: Akhilanand Chaurasia, Flat No. 701, New Faculty Residence, T. G. Hostel Campuse, Khadra, Lucknow (U.P.)
E-mail: chaurasiaakhilanand49@gmail.com

Case Report

A 60 year old female (Fig.1) complaining of a swelling involving right side of upper jaw since last 6 months. The swelling was progressively increasing in size and had ulcerative surface and causing difficulty in eating and chewing food. There was no significant past medical or dental history. The patient was a pan chewer since last 30 years. There was no significant extra-oral finding. However, occasionally nasal stuffiness and blockade of right side of nose was reported by patient. Intra-oral examination showed an exophytic, well defined, ball shaped, nontender, soft to firm swelling of 3x3 cm extending from right maxillary 2nd premolar to right maxillary tuberosity region (Fig.2). Palatally the swelling was involving lingual gingival and hard palate also. The

surface of swelling was yellowish red in color having smooth surface. The centre of swelling had ulceration having rough surface and covered by yellowish white necrotic slough. There was no associated bleeding.

Fig. 1

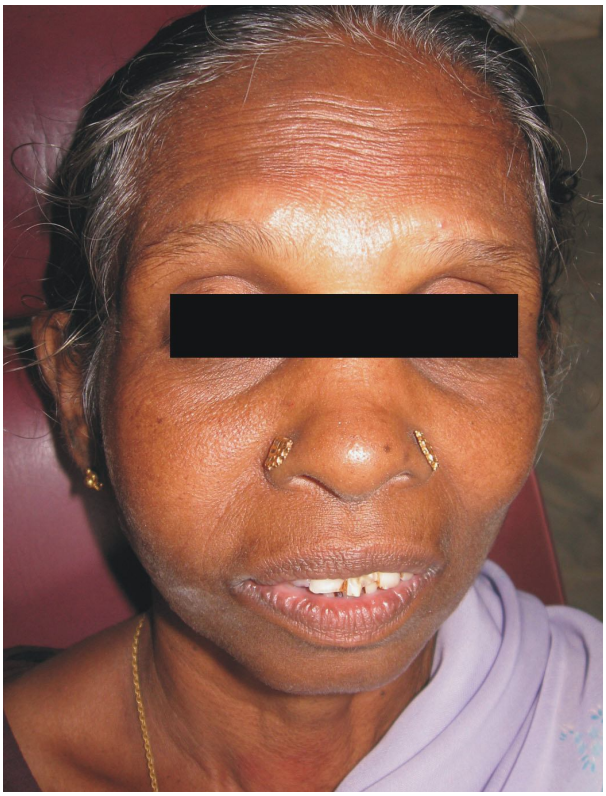
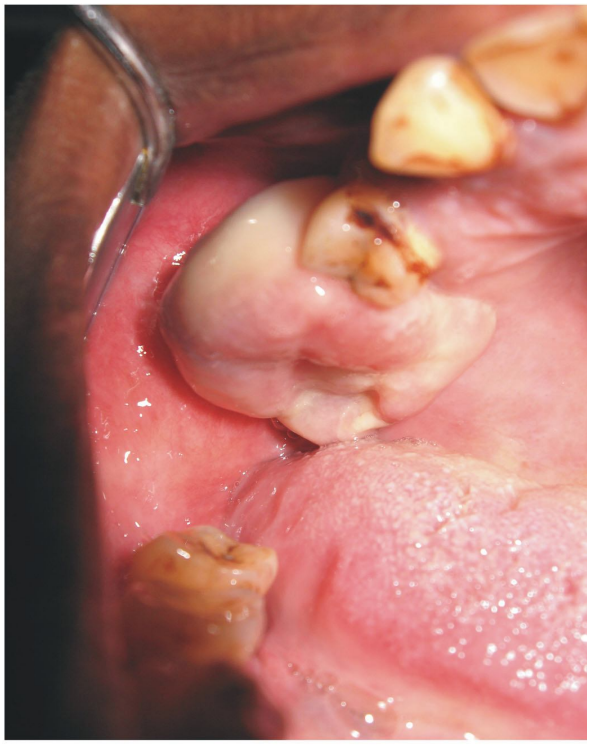


Fig. 2



The swelling was interfering with chewing and eating of patient and sometimes causing pain. The patient was advised for radiographic investigation involving panoramic radiograph and contrast

Fig. 3

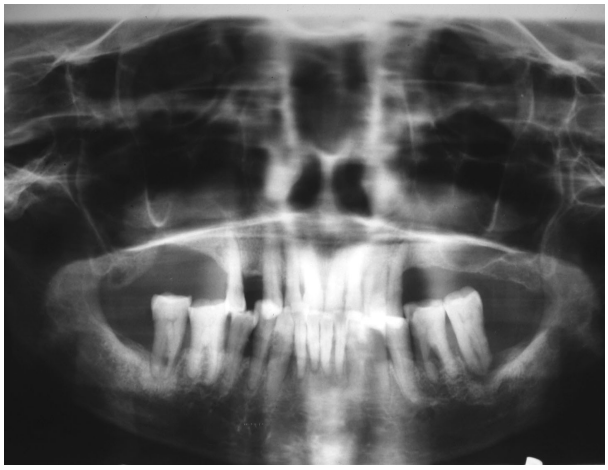


Fig. 4a

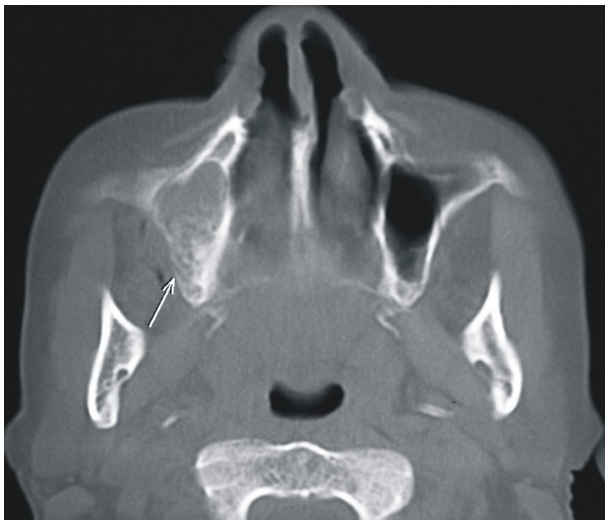
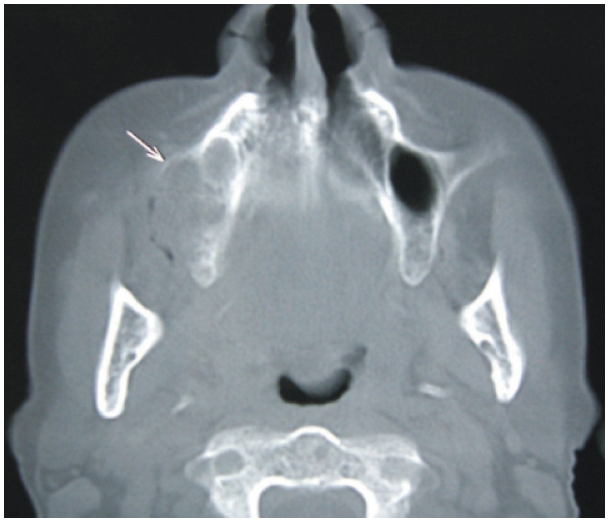


Fig. 4b



enhanced CT of right maxilla. The panoramic radiograph (Fig.3) showed breaching in floor and haziness of right maxillary sinus and loss of lamina dura around mandibular 1st premolar. The contrast enhanced CT showed homogenous mass density lesion with attenuation value of 45-60HU noted involving right maxillary alveolus causing erosion of alveolar process and extending into floor right side maxillary sinus (Fig. 4a, 4b, 4c, 4d). The homogenous mass is extending into soft tissue plane of right buccal space (Fig.5).The air pockets are noted within lesion suggestive of ulceration. Nasal septum was deviated towards left. The homogenous mass was closely related to right masseter muscle while pterygoid process was intact. On the basis of clinical and radiographic findings a provisional diagnosis malignancy has been reached. The patient was further advised for incisional biopsy. The routine haemogram, HbsAg, HIV was done. All the findings were within normal limits.The patient has undergone incisional biopsy (Fig. 6a & 6b) under LA and biopsied specimen was sent for histopathological examination. The histopathology showed both squamous and sarcomatoid component. Variable patterns of growth are seen including fascicular and storiform composed of atypical spindle cells with increased mitotic figures. The squamous cell carcinoma consisted of polygonal cells exhibiting intracellular keratiniztion, moderate to marked nuclear polymorphism and hyperchromatic nuclei(Fig.7a & 7b). On immunohistochemistry cells showed strong positivity to both cytokeratin (Fig.8) and vimentin (Fig.9). The patient was referred to

Fig. 4c

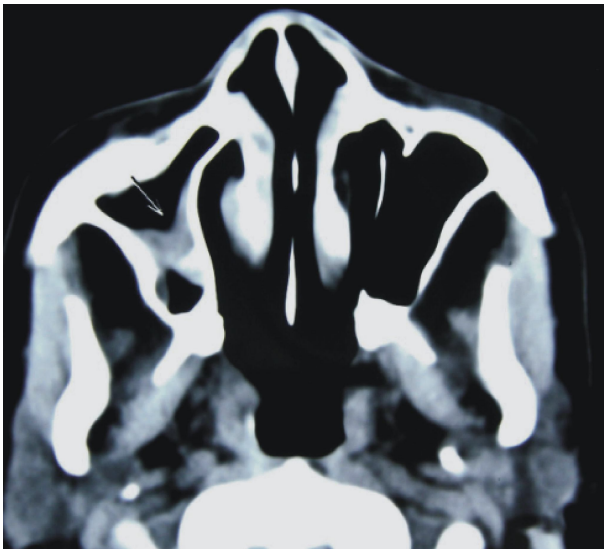


Fig. 4d

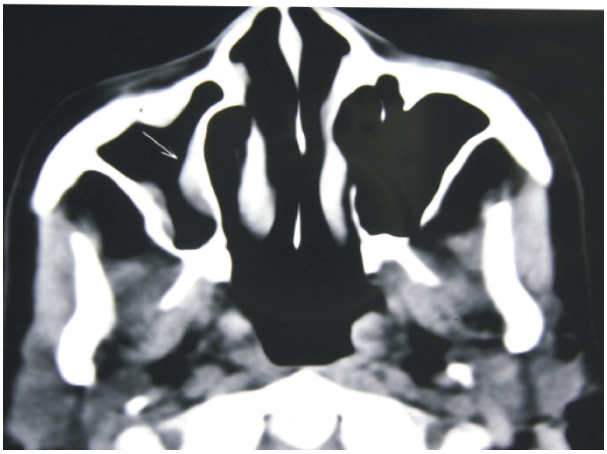


Fig. 5

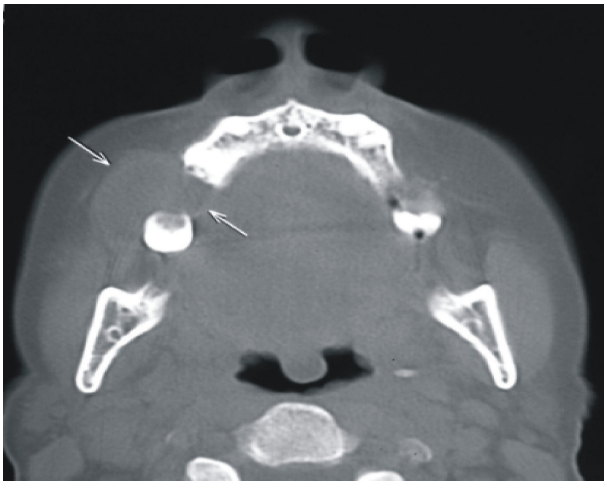


Fig. 6a

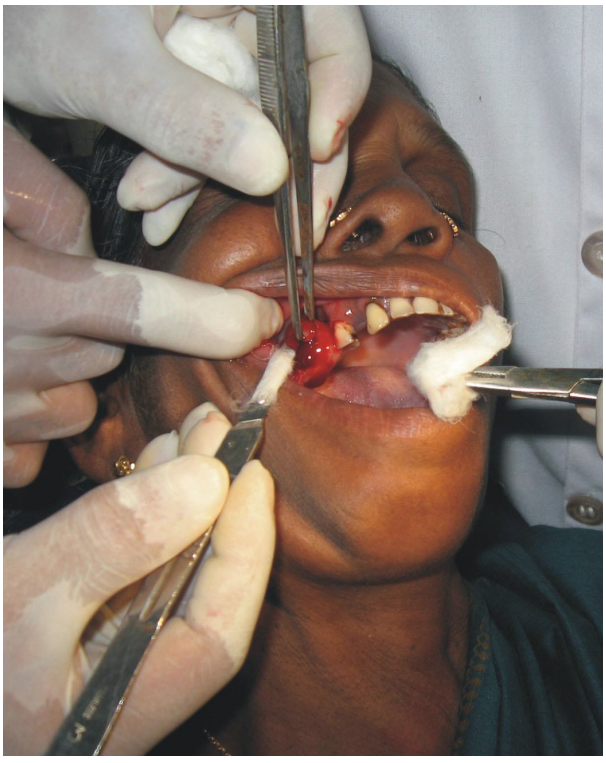


Fig. 6b

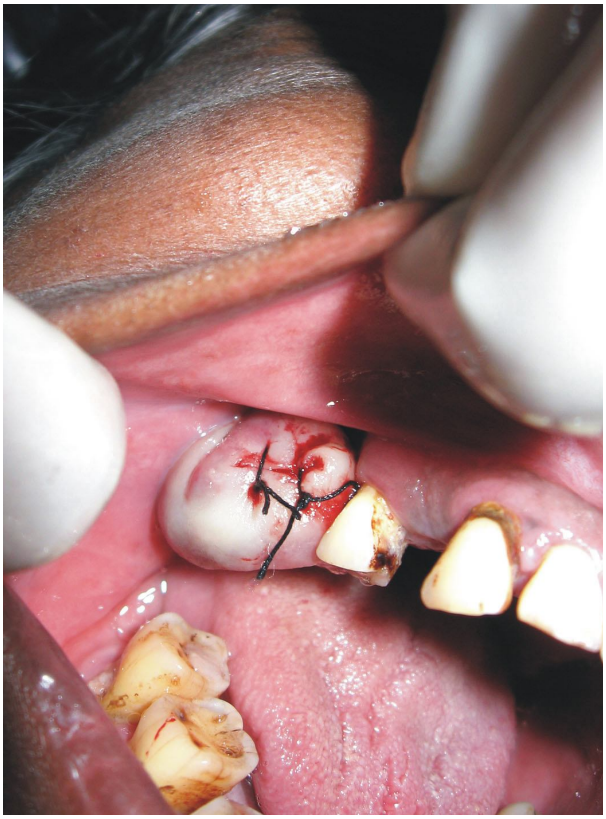


Fig. 7a

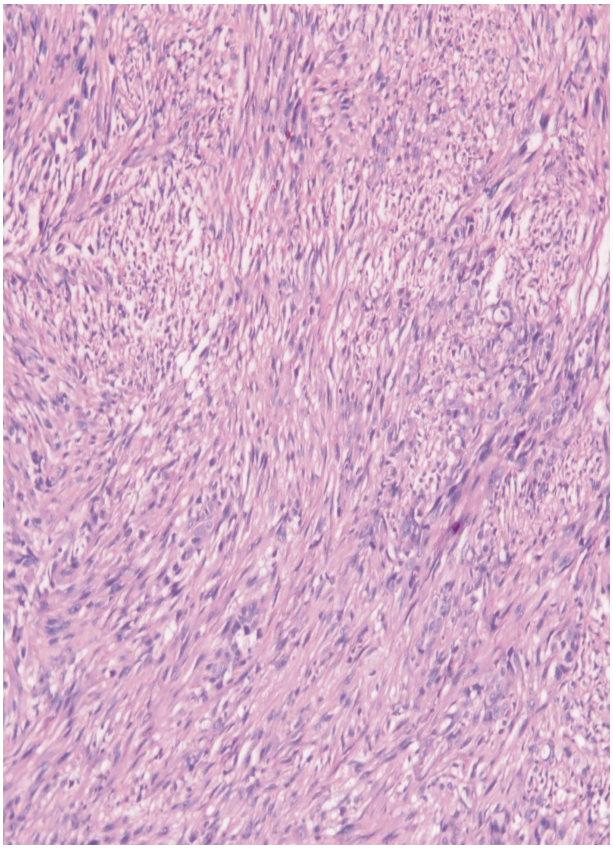


Fig. 7b

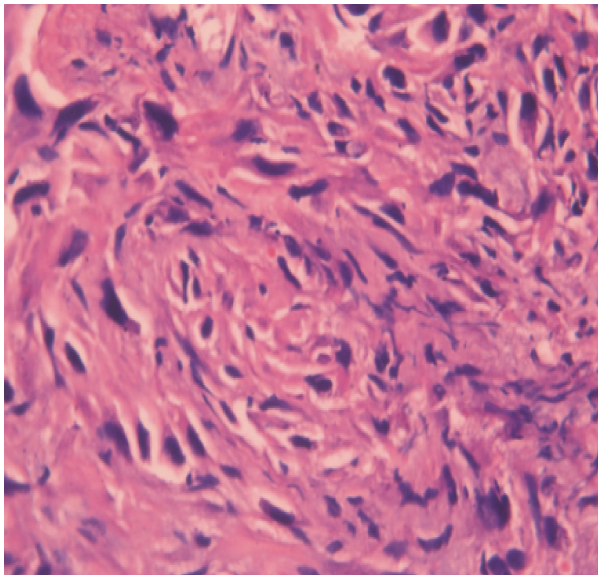


Fig. 8

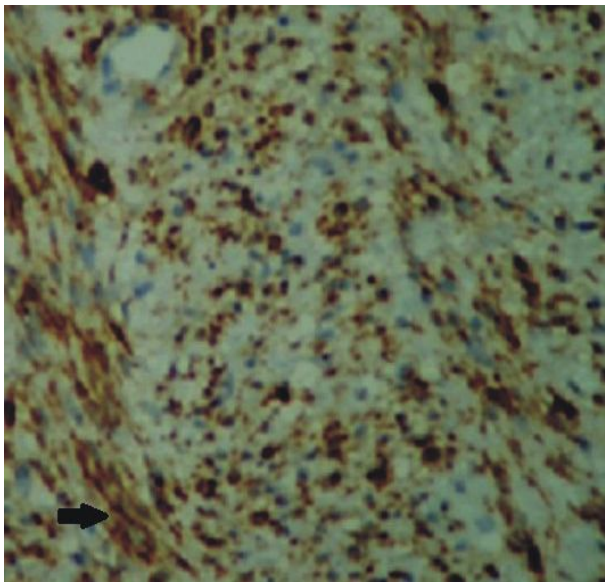
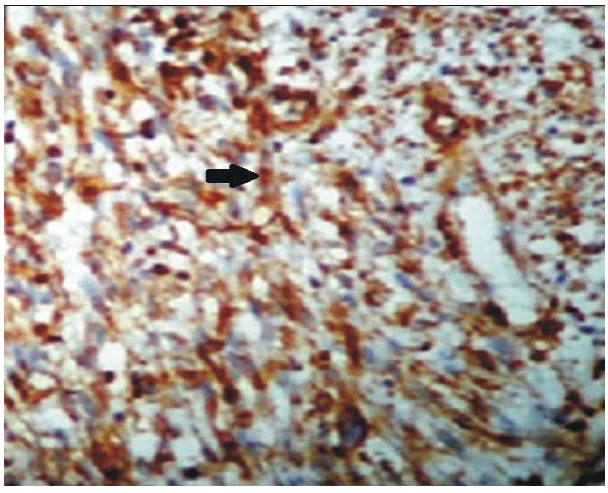


Fig. 9



surgical oncology for further management where she underwent partial maxillectomy on the right side with prophylactic right radical neck dissection. Histopathology and immunohistochemistry of excised specimen confirmed the diagnosis of spindle cell carcinoma of right maxillary sinus. Postsurgery the patient underwent radiotherapy. The patient was followed every 3 month for 1 year but no reccurrence have been reported.

Discussion

Spindle cell carcinoma is a subgroup of malignant mixed tumours and is extremely rare in maxilla. In 1864 Virchow first described this malignancy and coined the term 'carcinosarcoma' for this rare entity [7]. The term spindle cell carcinoma was first used by Shervin et al [2]. Spindle cell carcinoma accounts for less than 1% of all tumours of oral regions [8]. The age at the time of diagnosis range from 47 to 88 yrs. with mean age of 65.7 yrs and shows male preponderance [4]. The most pre-dilected site is the lower lip, tongue, alveolar ridge or gingiva however most tumours in head and neck region occur in the larynx [3, 9]. Spindle cell carcinoma is biphasic malignant neoplasm with controversy ever the basic nature of sarcomatoid element whether it is benign or malignant and mesenchymal or epithelial in origin. The sarcomatoid cells thought to be derived from squamous cells and the epithelial nature of the sarcomatoid component of spindle cell carcinoma was revealed by combination of immuno histochemical staining for keratins and electron microscopic demonstration of tonofilament or desmesome like structure [4]. The 4 etiological factors which predisposes to spindle cell carcinoma are tobacco, alcohol, poor oral health, previous irradiation to area of tumour. However some author emphasized that radiation or trauma induces spindle cell carcinoma [2].

The histogenesis of spindle cell carcinoma is still controversial. Some authors believe that spindle cell component is a benign stromal reaction against squamous cell carcinoma while others proposes that spindle cell are metaplastic change of malignant squamous cells [10]. Three dominant pathogenetic theories have been proposed. The 1st theory proposes that spindle cell carcinoma represents a "collision tumour" (carcinosarcoma) while according to 2nd hypothesis it is a squamous cell carcinoma with an atypical reactive stroma (pseudosarcoma). The 3rd theory states that spindle cell carcinoma is of epithelial origin with de-differentiation or transformation to a spindle cell morphology (sarcomatoid carcinoma) [9]. Clinically spindle cell carcinoma is characterized by exophytic, polypoid or nodular sessile growth. The most common symptoms are swelling, pain and presence of non-healing ulceration. The spindle cell carcinoma shows extensive surface ulceration with friable, fibrinoid necrosis of variable thickness or shaggy exudates [11, 12]. The list of differential diagnosis for spindle cell carcinoma is long which includes a number of benign and malignant tumors such as squamous cell carcinoma, fibrosarcoma, leiomyosarcoma, mesenchymal chondrosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma ,osteosarcoma, Kaposi's sarcoma, angiosarcoma, synovial sarcoma, malignant melanoma, fibromatosis, leiomyoma, nodular fasciitis and reactive epithelial proliferations [11]. Microscopically spindle cell carcinoma is typically characterized by a biphasic growth pattern with both spindle cell and squamous component. The squamous component may be represented by dysplasia, carcinoma in situ or invasive carcinoma. The spindle cell component may assume various histological patterns. The pleomorphic (malignant histiocytoma like) and spindle cell sarcoma (fibrosarcoma type) are most common histological patterns [13]. However, histological studies alone cannot explain the spindle cell components. Recent immunohistochemistry studies explained that histogenesis of spindle cell elements in spindle cell carcinoma is of epithelial origin. This fact is proved by positive keratin and vimentin immunostaining and presence of desmosomes and tonofilaments in spindle cells [14]. Immuno histochemically most sensitive and reliable epithelial markers for demonstration of epithelial components are keratin and epithelial membrane antigen which is useful in differential diagnosis from other sarcomatous lesions. P63 has been reported as useful marker for spindle cell carcinoma [4]. Kudo et al reported that spindle cell squamous carcinoma cells expressed wnt-5a and vimentin mRNA at high levels but did not express E-cadherin mRNA. This pattern was similar to that of fibroblast but not to oral squamous carcinoma. These findings suggested that the nature of spindle cell squamous carcinoma cells may be similar to mesenchymal cells. However the positivity for cytokeratin showed epithelial nature of spindle cell carcinoma cells [1]. The spindle cell carcinoma of oral cavity and oropharynx is a potentially aggressive tumor and seems to recur easily and to metastasize [15]. Some authors are of the opinion that wide surgical excision is the treatment of choice while others are opinion that surgery with radiotherapy is needed.

Radiation therapy is considered an acceptable alternative for inoperable patients. Adjuvant irradiation might be of benefit in cases in which the surgical margins are positive or in patients with nodal metastasis at the time of diagnosis [3]. Prognosis is related to location, tumour size, depth of invasion, stage of diseases and with the presence of any keratin staining in the spindle cells [12]. The biologic behaviour of spindle cell carcinoma varies from patient to patient. The deeply invasive tumours tend to have poor prognosis whereas those with early stage tumour usually have excellent prognosis [15]. The reported incidence of metastases was 36% for spindle cell carcinoma involving oral cavity [2]. The reported 5 year disease free survival rate is ~30% [16].

Conclusion

Spindle cell carcinoma is a biphasic tumour 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. The spindle cell carcinoma of maxillary sinus constitutes an uncommon occurrence and follows aggressive course with a high mortality rate. Sarcomatoid carcinoma 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 for a better prognosis and thereby reducing morbidity and mortality.

References

1. Kudo Y, Ogawa I, Kitagawa M, Kitajima S, Siriwardena BSMS, Aobara N et al. Establishment and characterization of a spindle cell squamous carcinoma cell line. J oral pathol med 2006; 35: 479–83.
2. Munakata R, Cheng J, Nakajima T, Saku T. Spindle cell carcinoma of the gingiva: report of an autopsy case. J Oral Pathol Med 1998; 27: 180–184.
3. Rizzardi C, Frezzini C, Maglione M, Tirelli G, Melato M. A look at the biology of spindle cell squamous carcinoma of the oral cavity: report of a case. J Oral Maxillofac Surg 2003; 61: 264–8.
4. Parikh N, Desai N. Spindle cell carcinoma of oral cavity: A case report of a rare entity and review of literature. J Academy Adv Dental Research 2011; 2: 31–36.
5. Weidner N. Sarcomatoid carcinoma of the upper aerodigestive tract. Semin Diagn Pathol 1987; 4: 157–68.
6. Batsakis JG, Suarez P. Sarcomatoid carcinomas of the upper aerodigestive tracts. Adv Anat Pathol 2000; 7: 282–93.
7. Berthelet E, Shenouda G, Black MJ, Picariello M, Rochon L. Sarcomatoid carcinoma of the head and neck. Am J Surg. 1994; 168: 455–8.
8. Jordan RCK, Regezi JA. Oral Spindle Cell neoplasms: A review of 307 cases,Oral surg,Oral Med,Oral Pathol,Oral Radiol,Endo 2003; 95: 717–24.
9. Huang SF, Chen IH, Liao CT, Chen TM, Lee KF. Sarcomatoid carcinoma of the parotid gland with apparent metastasis of epidermoid elements to cervical lymph nodes. Acta Otolaryngol 2006; 126: 667–71.
10. Nilima P, Harish Kumar MS,Sharada P,Pradeep GL:Spindle cell carcinoma of oral cavity: A case report of a rare entity and review of literature:World Journal of Dentistry, 2010; 1(1): 55–58.
11. Thompson LD, Wieneke JA, Miettinen M, Heffner DK. Spindle cell (sarcomatoid) carcinomas of the larynx: a clinicopathologic study of 187 cases. Am J Surg. Pathol. 2002; 26: 153–70.
12. Minami SB, Shinden S, Yamashita T. Spindle cell carcinoma of the palatine tonsil: report of a diagnostic pitfall and literature review. Am J Otolaryngol 2008; 29: 123–5.
13. Jean E Lewis, Kerry D Oslen, Thomas J Sebo. Spindle cell carcinoma of larynx. Review of 26 cases including DNA content and immunohistochemistry. Human Pathology.1997; 28 (6): 664–73.
14. Shibuya Y,Umeda M,Yokoo S,Komori T. Spindle cell squamous cell carcinoma of maxilla-Report of a case with immunohistochemical analysis. J Oral maxillofac surg.2000; 58: 1164–69.
15. Su HH, Chu ST, Hou YY, Chang KP, Chen CJ. Spindle cell carcinoma of the oral cavity and oropharynx: factors affecting outcome.J Chin Med Assoc 2006; 69: 478–83.
16. Randall G, Alonso WA, Ogura JH. Spindle cell carcinoma (pseudosarcoma) of the larynx Arch Otolaryngol 1975; 101: 63–66.