Two cases of spindle cell carcinoma with a histopathological review of biphasic tumours of head and neck

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Abstract

Spindle cell carcinoma (SPCC) is a very rare and unique biphasic variant of squamous cell carcinoma (SCC) which is a very rare malignancy of head and neck region. Spindle cell tumours are challenging feature of head and neck pathology because of different types of spindle cell lesions occurring which can be reactive to benign to malignant and very aggressive. We present here two rare cases of SPCC one involving the nasal cavity in a 38 year female and other a very aggressive maxillary sinus tumour in a 60 year male and discuss its differential diagnosis with an emphasis on biphasic tumours of head and neck.

Key words: Spindle cell carcinoma, Head and neck, Biphasic tumours

Introduction

Spindle cell carcinoma (SPCC) is a very rare and unique biphasic variant of squamous cell carcinoma (SCC) which can be seen to involve any part of the body including head and neck region. It has an epithelial origin but with a mesenchymal appearance hence called spindle cell carcinoma or carcinosarcoma. Nasal cavity and sinuses are very rare sites of involvement [1, 2]. Spindle cell tumours are challenging feature of head and neck pathology because of different types of spindle cell lesions occurring which can be reactive to malignant and very aggressive. This makes accurate diagnosis difficult.

We present here two rare cases of SPCC one involving the nasal cavity in a 38 year female and other a very aggressive maxillary sinus tumour in a 60 year male and discuss its differential diagnosis with an emphasis on biphasic tumours of head and neck.

Case Reports

Case-1: A 38 year old female presented with history of intermittent epistaxis from left nasal cavity from one year with gradually increasing nasal obstruction from 6 months. There was no history of smoking or alcohol addiction. On examination, the patient had an ulcerated, polypoid, irregular mass obstructing the complete left nasal vestibule. There was no lymphadenopathy. The patient was weak and anaemic (Haemoglobin- 9 gm/dl) Computerized tomography (CT) scan of the head and neck showed a heterogeneously enhancing mass of the left nasal cavity. Biopsy of the mass revealed a malignant spindle cell proliferation (Figure 1) with tumour cells showing hyperchromatic nuclei and prominent nucleoli and eosinophilic cytoplasm. (Figure 2) Immunohistochemistry showed tumour cell to be pan-cytokeratin (CK), vimentin positive. (Figure 3) Smooth muscle actin (SMA), desmin, HMB 45, S100, calponin, CD 34, myogenin were negative. A diagnosis of Spindle Cell Carcinoma (SPCC) of nasal cavity was made. The patient was referred to higher centre for surgical resection and management where tumour was removed by wide local excision. The patient did well post surgery and received chemo radiotherapy. She is currently under follow up from 1 year.

Case-2: A 60 year male was suffering from intermittent epistaxis from 6 months. Then he started to complain of left nasal obstruction from 4 months and slowly developed diplopia. He also complained of anorexia, dysphagia and weight loss in past 1 month. Patient had the addiction of using snuff and smoking. On examination, a diffuse swelling was seen
below the left eye and cheek measuring 5x4cm extending to the hard palate with slight proptosis of left eye. Patient was very weak and anaemic (Hemoglobin 8 gm/dl). On rhinoscopy, patient had a large pinkish fleshy mass in left nasal cavity. There was no lymphadenopathy. CT scan and MRI of head and neck showed a large heterogenous mass in the maxillary sinus and with destruction of all its walls and extension to the left orbital cavity with involvement of the medial and inferior rectus muscles. The mass had also extended into the nasal cavity and superiorly to the cribriform plate, posteriorly to the left pterygopalatine fossa and the infra-temporal fossa.

There was destruction of the inferior wall of the sphenoid sinus and inferiorly, the mass had destroyed the left side hard palate, extending into the oral cavity (Figure 4). Biopsy of the nasal mass showed malignant spindle cell proliferation in a hemorrhagic background with spindle cells showing hyperchromatic nuclei and prominent nucleoli and eosinophilic cytoplasm. Abundant blood vessels with plump endothelial cells and neutrophils were seen mimicking granulation tissue (Figure 5).

Immunohistochemistry showed tumour cell to be pan-cytokeratin (CK), vimentin, smooth muscle actin (SMA) positive. However desmin, HMB 45, S100, calponin, CD31, CD 34, myogenin, CD 99 were negative. A diagnosis of SPCC of maxillary sinus was made.

Squamous cell component was not seen in both the cases.
The patient was referred to higher centre for further management but expired before being posted for the surgery.

**Discussion**

SPCC is a unique biphasic variant of SCC that histologically exhibits spindled tumour cells. Many alternate terms like carcinosarcoma, pseudo sarcoma, pleomorphic carcinoma, and metaplastic carcinoma had been used for it. It is a rare biphasic tumour which can involve any organ including head and neck region. Many of the characteristic features of this tumour are similar to the conventional SCC [1, 2].

SPCC is more common in elderly people in 5th to 6th decades with male preponderance (Male: female-7:1). It also has a strong association with smoking and alcoholism [1,2,3]

In head and neck, larynx (glottis) is the most common primary site, followed by the oral cavity (tongue, floor of mouth, gingiva). Less common sites include the hypopharynx, oropharynx, sinuses and nasal cavity. They usually present as polyoidal, fleshy, exophytic masses with average size of 2cm [1,2]. Clinically they present as locally advanced stage, most tumours being T3 or T4 at presentation. It is an aggressive tumour if not treated in time [4].

Most significant are the histological features of this tumour, characterized by a biphasic pattern of SCC and generally much larger component of malignant spindled cells, reminiscent of a sarcoma. The squamous component may be scant or even absent on light microscopy. The spindled cells may be bland and regular or may be markedly pleomorphic with multinucleated giant tumor cells with wide variety of architectural patterns including fascicular, storeiform, lacelike, or myxoid and rarely sarcomatous differentiation, such as osteosarcomatous, chondrosarcomatous, or rhabdomyosarcomatous may be seen. Mitotic activity can vary. The tumour can show abundant small vessels with plump endothelial cells and numerous inflammatory cells, particularly neutrophils and can closely mimic exuberant granulation tissue. This makes the diagnosis very difficult and requires immunohistochemical (IHC) and ultra structural studies.

Differential diagnosis of SPCC is therefore extremely exhaustive as many tumours of spindle cell morphology resemble it. Non neoplastic lesions like exuberant granulation tissue, nodular fasciitis, sinonasal polyp with stromal atypia, borderline tumours like inflammatory myofibroblastic tumour and malignant tumours like spindle cell myoepithelioma or carcinoma, solitary fibrous tumor, Kaposi’s sarcoma, synovial sarcoma, angiosarcoma, fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumour (MPNST), undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma), spindle cell melanoma, spindle cell variant of rhabdomyosarcoma, high-grade transformation of dermatofibrosarcoma protuberance.

Non neoplastic and benign low grade tumours can be differentiated by clinical history, absence of atypical mitoses, cellularity, nuclear features and organization while malignant tumours often require further immunohistochemistry with desmin, muscle specific actin, MYO-D, HMB 45, S100, calponin, CD 10, CD 31, CD 34, myogenin, CD 99 etc for confirmation [1,5].

SPCC stain with both epithelial and mesenchymal markers. AE1/AE3 (pancytokeratin) is positive in anywhere from 26% to 62% of cases. Epithelial
Spindle cell carcinoma is a very rare and unique biphasic variant of squamous cell carcinoma. Diagnosis is challenging in head and neck because of different types of spindle cell lesions and requires immuno-histochmical and ultrastructural studies. Their behaviour is similar to that of poorly differentiated carcinomas. Tumour is primarily treated with aggressive, wide local excision. Adjuvant radiation and/or chemotherapy should be used in case of incomplete removal or positive surgical margins [12]. While other studies suggest that postoperative chemoradiation therapy improves patient prognosis and should be considered as the standard therapeutic modality [13].

**Conclusion**

Spindle cell carcinoma is a very rare and unique biphasic variant of squamous cell carcinoma. Diagnosis is challenging in head and neck because of different types of spindle cell lesions and requires immuno-histochmical and ultrastructural studies. Their behaviour is similar to that of poorly differentiated carcinomas. Tumour is primarily treated with aggressive, wide local excision, adjuvant radiation and/or chemotherapy used in case of incomplete removal or positive surgical margins.

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**References**

1. Viswanathan S, Rahman K, Pallavi S, Sachin J, Patil A, Chaturvedi P, D'Cruz A, Agarwal J, Kane SV. Sarcomatoid (spindle cell) carcinoma of the head and neck mucosal region: a clinic pathologic review of 103 cases from a tertiary referral cancer centre. Head Neck Pathol. 2010 Dec; 4 (4): 265-75. doi: 10.1007/s12105-010-0204-4. Epub 2010 Aug 22.
2. Leventon GS, Evans HL. Sarcomatoid squamous cell carcinoma of the mucous membranes of the head and neck: a clinicopathologic study of 20 cases. Cancer. 1981 Aug 15;48(4):994-1003.

3. 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 Feb;26(2):153-70.

4. Howard SN, Bond WR, Hong IS, Foss RD. Right maxillary sinus sarcomatoid carcinoma (sarcomatoid/spindle cell carcinoma). Otolaryngol Head Neck Surg. 2007;137(2):355-57.doi:10.1016/j.otohns. 2007.03.002.

5. Lewis JS Jr. Spindle cell lesions--neoplastic or non-neoplastic?: spindle cell carcinoma and other atypical spindle cell lesions of the head and neck. Head Neck Pathol. 2008 Jun;2(2):103-10. doi: 10.1007/s12105-008-0055-4. Epub 2008 May 28.

6. Lewis JE, Olsen KD, Sebo TJ.Spindle cell carcinoma of the larynx: review of 26 cases including DNA content and immunohistochemistry. Hum Pathol. 1997 Jun;28(6):664-73.

7. Ellis GL, Langloss JM, Heffner DK, Hyams VJ. Spindle-cell carcinoma of the aerodigestive tract. An immunohistochemical analysis of 21 cases. Am J Surg Pathol. 1987 May;11(5):335-42.

8. Choi HR, Sturgis EM, Rosenthal DI, Luna MA, Batsakis JG, El-Naggar AK. Sarcomatoid carcinoma of the head and neck: molecular evidence for the evolution and progression from conventional squamous cell carcinoma. Am J Surg Pathol. 2003; 27(9): 1216-20.

9. Fisher C. Immunohistochemistry in diagnosis of soft tissue tumours. Histopathology. 2011 Jun;58(7):1001-12. doi: 10.1111/j.1365-2559.2010.03707.x. Epub 2010 Dec 10.

10. Furuta Y, Nojima T, Terakura N, Fukuda S, Inuyama Y. A rare case of carcinosarcoma of the maxillary sinus with osteosarcomatous differentiation. Auris Nasus Larynx. 2001; 28(S): 127–29.

11. Gupta R, Singh S, Hedau S, Nigam S, Das BC, Singh I, Mandal AK. Spindle cell carcinoma of head and neck: an immunohistochemical and molecular approach to its pathogenesis. J Clin Pathol. 2007 May; 60 (5):472-5. Epub 2006 May 26.

12. Iqbal MS, Paleri V, Brown J, Greystoke A, Dobrowsky W, Kelly C, Kovarik J. Spindle cell carcinoma of the head and neck region: treatment and outcomes of 15 patients. Ecancer medical science. 2015 Nov 18;9:594. doi: 10.3332/ ecancer. 2015. 594. eCollection 2015.

13. Alem HB, AlNoury MK. Management of spindle cell carcinoma of the maxillary sinus: a case report and literature review. Am J Case Rep. 2014 Oct 24;15:454-8. doi: 10.12659/AJCR.891007.

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