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Spindle Cell Neoplasm of Nasal Cavity and Frontal Recess -A Rare Case Report.

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ABSTRACT-

Spindle cell carcinoma is a unique variant of squamous cell carcinoma. Spindle cell carcinoma confined to nasal cavity is extremely rare hence we present a case of nasal cavity spindle cell carcinoma. A 17year old male presented with intermittent epistaxis from right nasal cavity and right nasal blockage. On physical examination patient had polypoidal mass the in right nasal cavity. The patient underwent wide local excision of nasal mass from right nasal cavity and frontal recess and the tissue was sent for histopathology which was reported as spindle cell neoplasm with prominent peritheliomatous pattern. Three months after surgery patient continues to have no evidence of disease. No definite treatment protocol exists for this unique entity, but we believe that this tumor should primarily be treated with aggressive, wide local excision.

KEYWORDS - nasal cavity, frontal recess, spindle cell carcinoma

I. INTRODUCTION -

Spindle cell carcinoma (SpCC) is a unique and challenging variant of squamous cell carcinoma (SCC). In the upper aerodigestive tract, similar to conventional SCC, SpCC occurs most commonly in the larynx.[1] Less common primary sites include the hypopharynx, oropharynx, sinuses, and nasal cavity. SpCC confined solely to the nasal cavity is extremely rare, with only one case having been previously reported.[2] Herein we present a case of SpCC confined to the nasal cavity.

II. CASE REPORT-

A 17yr old, male presented to our ENT OPD with complaints of intermittent epistaxis from right nasal cavity since 2 weeks and right nasal cavity blockage since 2 months. History revealed that the swelling was insidiousin onset and gradually progressed to present size. There was no history of purulent rhinorrhoea, anosmia, facial pain, proptosis, dysphagia odynophagia, change in voice, vision lossor weight loss. There was no palpable lymphadenopathy.

On diagnostic nasal rigid endoscopy (Fig no.1) the patient had polypoidal, well circumscribed mass in right nasal cavity which was bleeding on touch and was extending upto the vestibule of the nose. Due to pressure effect of the lesion there was deviated nasal septum on left side. The lesion was vascular in nature so biopsy was not taken.

Visual acuity was checked and it was 6/6.

Intraoral examination revealed fair oral hygiene and full complement of teeth with no extension of swelling.

Computed tomography scan of paranasal sinuses (Plain + contrast) (Fig no.2)reported as well defined moderately enhancing lesion in the right nasal cavity. The lesion measures 4.2 x 1.5 x4.6 cm (AP x TR x CC). There are few soft calcific foci within it. The lesion is extending into right frontal sinus with subtle extension into left frontal sinus. There is bowing of nasal septum to left. The right osteomeatal unit and frontoethmoidal recess are blocked by the lesion. There is no extension of lesion into nasopharynx. No intracranial or intraorbital extension of the lesion. There is no obvious lesion in sphenopalatine foramen and pterygopalatine fossa. Differential diagnosis includes- 1.vascular tumour like haemangioma /angiomatous polyp 2.inverted papilloma 3.juvenile nasopharyngeal angiofibroma.

He underwent preoperative laboratory tests and consent for surgery and associated complication was taken. The case was planned for surgery under general anaesthesia.The patient subsequently underwent endoscopic excision of right nasal massby Lothrop II procedure under general anaesthesia. Grossly the mass was well-circumscribed, arising from frontal recess into right nasal cavity up to the vestibule.(Fig no.5)

The mass was sent for histopathological examination (Fig no.3) which was reported as spindle cell neoplasm with peritheliomatous pattern**.**Sections show tissue predominantly lined by respiratory epithelium. The subepithelial tissue

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shows a tumour composed of ovoid to spindle cells having oval nuclei and scanty cytoplasm arranged predominantly in peritheliomatous arrangement. Areas of dilated vascular spaces with haemorrhage and hemosiderophages are noted. In areas the tumour in hypercellular with fascicular arrangement and focal rosette like formation. Ultrastructural examination revealed intermediate filaments in the spindle cells without desmosomes or other junctional complexes. The cells was immunoreactive (Fig no.4) for cytokeratin 7(CK7), vimentin and pan keratin AE1/AE3. The tumour was negative for melanoma markers (HMB-45, Melan-A, S100) , smooth muscle actin(SMA),CD34 and myoepithelial markers (CD10, Calponin , p63 and GFAP). The diagnostic impression was spindle cell neoplasm.

Patient was adviced to follow up every monthly. Six months after surgery on nasal endoscopy patient had healthy nasal mucosa with well opened sinuses.No post-operative adjuvant therapy was given and the patient continues to have no evidence of disease.

III. DISCUSSION-

SpCC is a variant of SCC that histologically exhibits spindled or pleomorphic tumour cells. SpCC has similar demographics to that of conventional SCC. It occurs most often in the fifth and sixth decades, is strongly associated with smoking and alcohol use, and shows a strong male preponderance.[3] Risk factors for SpCC include smoking, alcohol consumption and previous radiotherapy. The larynx (glottis) is the most common primary site, followed by the oral cavity (tongue, floor of mouth, gingivae). Less common sites include the hypopharynx, oropharynx, sinuses, and nasal cavity.[4–6] SpCC has a unique, macroscopic growth pattern.[1] In the larynx, greater than 90% of the tumours present as polypoid, ulcerated, exophytic masses.[3,5,7]

Histologically, SpCC are often biphasic, showing areas of conventional SCC mixed with areas of spindle and/or pleomorphic tumor cells. However, it has been reported that in up to 28% of cases, a classical SCC component cannot be identified.[8–9] While the clonal proliferation of atypical spindle cells may simulate a sarcoma of mesenchymal origin, recentstudies have shown head and neck sarcomatoid carcinomas to arise from transformation of more conventional epithelial components.[10–13] While recent molecular studies have lent credence to this argument, clinically this can be shown through the use of immunohistochemistry - most of these tumors stain with both epithelial and mesenchymal markers. The most common epithelial stain, AE1/AE3 (pancytokeratin), is positive in anywhere from 26% to 62% of cases.[3,5,14] Some cases have also been shown to express other common epithelial markers including EMA and p63.[3,5,6,14] In regards to mesenchymal-type markers, almost 100% of cases are positive for vimentin.[3,5] Some SpCC lesions have also been shown to express markers of muscle differentiation including SMA and muscle specific actin.[3] Ultrastructurally, the presence of cytokeratin type intermediate filaments, cadherins, desmosomes, tight junctions or adherens junctions also supports an epithelial origin of these tumors.[10] One previous case has been reported by Ahluwalia et al.[2] in 1996. The patient was a 40-year-old Hindu male farmer who presented with recurrent epistaxis for one year. Nasal endoscopy showed a grey-white mass attached to the left side of the nasal septum. A biopsy showed a variable histologic pattern, with areas of spindled cells and conventional SCC. Immunohistochemistry showed positivity for pancytokeratin and vimentin, confirming the diagnosis of SpCC. The patient underwent wide local excision of the mass and adjuvant radiotherapy. Two months following therapy completion he developed ipsilateral nodal metastases that were positive for SCC, which were treated with a modified radical neck dissection. At the time of the report, the patient showed no evidence of disease.

Since the inverted papilloma can potentially transformed into SCC,it is likely that it can transform into SpCC, as it is a variant of SCC. It is believed that the chronicity of inverted papilloma is associated with the induction of SpCC transformation.[15]

Another case of a nasal cavity SpCC, this time extending into the maxillary sinus, has been recently reported.[16] The patient was a 75-year-old man who presented with nasal obstruction. Nasal endoscopy showed a 3×3 cm polypoid mass, extending from the nasal septum into the maxillary sinus. A biopsy showed malignant spindle cells with hyperchromatic nuclei. Immunohistochemistry showed positivity for pancytokeratins, high molecular weight cytokeratin, CK 5/6, CK18, CK19, p63, and vimentin. Thus, a diagnosis of SpCC was made. The patient underwent resection of the right maxilla followed by chemotherapy (5-fluorouracil) and radiation at another hospital. At the time of the report, the patient was alive without metastasis 5 years after initial presentation.

IV. CONCLUSION-

On the basis of histopathology and immunohistochemistry we diagnosed the case as

spindle cell neoplasm of right nasal cavity and frontal recess. No definite treatment protocol exists for this unique entity, but we believe that this tumour should primarily be treated with aggressive, wide local excision. Adjuvant radiation and/or chemotherapy have also been usedanecdotally.

CONFLICTOF INTEREST -NIL

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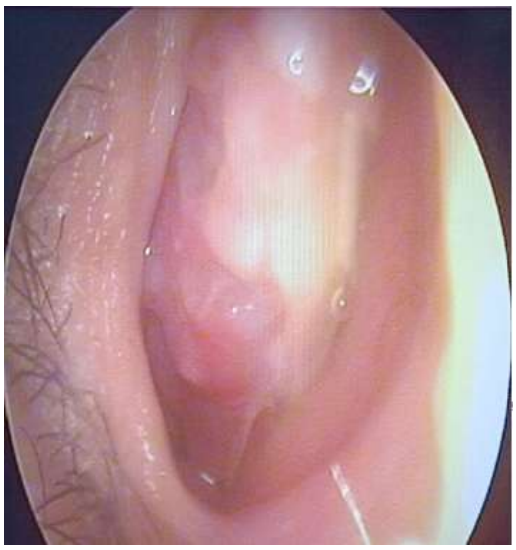

Figure no.1 Diagnostic NasalEndoscopic view of right nasal cavity mass

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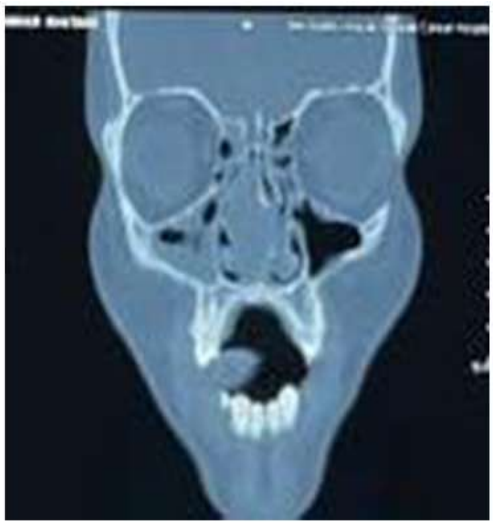

Figure no.2 Computed tomographyscan with contrast

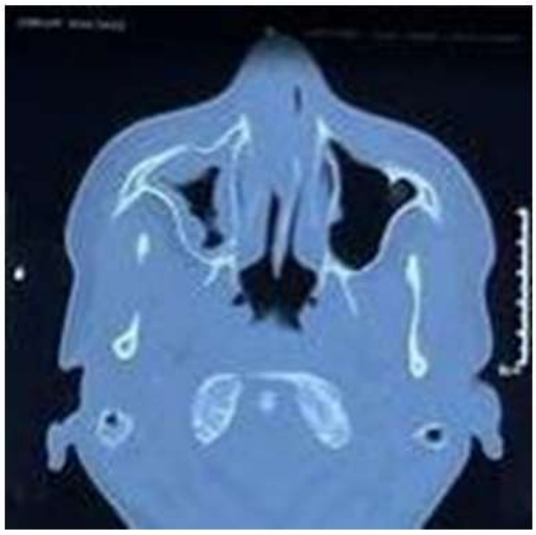

Figure no.3 Histopathological images

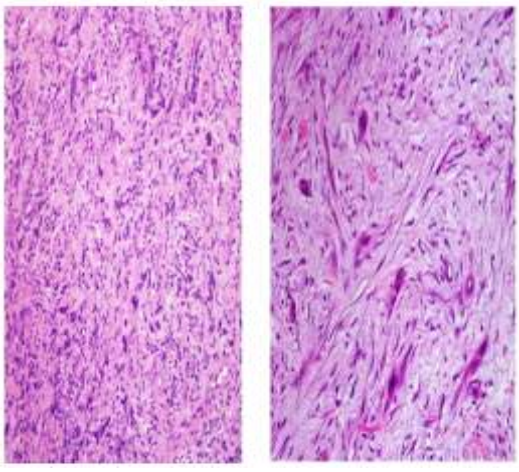

Figure no.4 Immunohistochemistry images

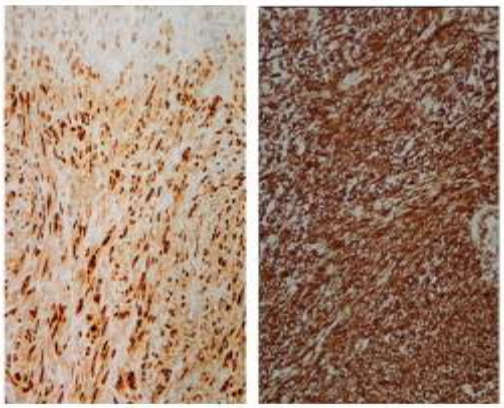

Figure no.5 Intraoperative picture of right nasal cavity-