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Basaloid Squamous Cell Carcinoma of the Sinonasal Tract

Abstract: Basaloid squamous cell carcinoma (BSCC) is a rare variant of squamous cell carcinoma and is known for its aggressive behaviour. It occurs predominantly in the upper aerodigestive tract and presentation in the nasal cavity and paranasal sinus is extremely rare. We present here a case of BSCC in a 36 year female arising in the paranasal sinus with orbital and intracranial extension. Clinical and pathological findings including immunohistochemistry is presented along with brief discussion of literature.

Keywords: basaloid squamous cell carcinoma, squamous cell carcinoma, sinonasal tract.

Introduction: Basaloid squamous cell carcinoma (BSCC) is a rare and high grade variant of squamous cell carcinoma, that preferentially arises in the upper aerodigestive tract, i.e. the base of tongue, tonsil, supraglottic larynx and hypopharynx. The first case was described by Wain et al in 1986 in the upper aerodigestive tract.¹ There have been a few reported cases of sinonasal tract BSCCs. According to Lu et al, less than 30 cases of sinonasal BSCC have been reported.² We report here a case of BSCC arising in paranasal sinus in a 36-year-old woman who presented with unilateral nasal obstruction, proptosis and epistaxis.

Case presentation: A 36 year old woman presented with a four month history of epistaxis, left side nasal obstruction, proptosis and decreased vision in left eye. Past medical history was unremarkable. There was no history of smoking or alcohol consumption.

Nasal examination revealed a mass in the upper portion of left nasal cavity and paranasal sinus. All the other laboratory data was within normal limits. Fundus examination of revealed dilated pupil, clear media, clear disc margins, temporal disc pallor and background vessels were normal.

The computed tomography (CT) scan of head revealed a hypo dense mass lesion showing enhancement on contrast in the left nasal cavity involving ethmoid air cells with expansion and deviation of medial wall of orbit (figure 1). Opacification of bilateral frontal sinus and extension into sphenoid air cells was also seen (figure 2).

Magnetic resonance imaging (MRI) of brain revealed soft tissue mass of mixed intensity on T2W, hypo intense on T1W and heterogenous enhancement on bilateral ethmoid cells with few cystic areas eroding lamina papyracea and cribriform plate.

Probable diagnosis of carcinoma ethmoid air cells and esthesioneuroblastoma were made.

An endoscopic biopsy was performed under local anesthesia. Microscopic examination revealed nests of tumor cells arranged in cords, trabeculae and lobules. There were two types of cell population. The predominant cell population was that of basaloid cell type having hyperchromatic nuclei, inconspicuous nucleoli and scanty cytoplasm. These lobules also display peripheral nuclear palisading (figure 3). The second population was squamous cell type with more cytoplasm, squamous whorls, individual cell keratinization, and intercellular bridges. Atypical mitosis and areas of haemorrhage were also present.

On immunohistochemical (IHC) staining, the tumor cells were positive for High molecular weight cytokeratin 34 β E12, EMA, p63 and negative for vimentin, chromogranin, Bcl2 (figure 4). The above histologic and immunohistochemical findings were consistent with basaloid squamous cell carcinoma.

Discussion: BSCC is a rare and aggressive variant of squamous cell carcinoma that was first identified as a separate histopathologic entity by Wain et al in 1986.¹ It occurs most commonly in upper aerodigestive tract including the oral cavity, larynx, hypopharynx, pyriform sinus, tonsils and the base of tongue. Other less frequently affected sites are nose, paranasal sinus, gingiva, external ear, submandibular region, esophagus, lung, anus, vulva, vagina and the uterine cervix.^{3,4} Although this type of tumor is most commonly found in the head and neck region, occurrence in the nasal cavity or in the paranasal sinus is extremely rare with less than 30 cases having been reported in the current literature.²

It is predominantly found in males in older age group. Tobacco and alcohol abuse are associated etiological factors, however the role of viral infection is unexplored.³ Unilateral nasal obstruction is the most common reported clinical symptom of nasal or paranasal BSCC.⁵

Clinically, it is an aggressive tumor characterized by a high incidence of cervical lymph node metastasis (64%) and distant spread (44%), with 38% mortality and median survival of 17 months.³ A case-control study by Soriano et al found a six times higher risk of distant metastasis as compared to usual type of SCC.⁶ Most BSCCs are diagnosed at advanced clinical stages and they have an unfavourable prognosis because of the poor overall patient survival rates.

Microscopically, BSCC is characterised by nesting, lobular and trabecular pattern comprising of small, crowded cells with hyperchromatic nuclei and scant cytoplasm. The lobules of basaloid cells often display peripheral nuclear palisading, comedonecrosis, high mitotic activity and small cystic spaces.³ The second major characteristic of the BSCC is the presence of squamous component that includes at least one of following features: adjacent foci of conventional squamous cell carcinoma, dysplasia or carcinoma in situ of the overlying mucosa.⁷

The differential diagnosis of BSCC includes adenoid cystic carcinoma (ACC), small-cell neuroendocrine carcinoma (SCNC), olfactory neuroblastoma, basal cell carcinoma (BCC), squamous cell carcinoma and adenosquamous carcinoma.⁴ Immunohistochemical studies are useful for differential diagnosis.

Both BSCC and ACC may have PAS positive microcystic spaces. However, ACC has less pleomorphism, mitosis and necrosis than BSCC.⁸ On IHC, BSCC were negative for vimentin

and S100.⁹ Emanuel P et al found that BSCC displayed diffuse p63 positivity, with staining of nearly 100% of tumor cells as compared with ACC with compartmentalized pattern within tumor nests.¹⁰

BSCC, SCNC and olfactory neuroblastoma may show sheets of small blue cells and rosette like structures. However, SCNC shows characteristic nuclear moulding and crushing artefact, and is rarely connected to surface mucosa.⁸ On IHC, neuroendocrine markers as synaptophysin and chromogranin are positive for small cell neuroendocrine carcinoma and olfactory neuroblastoma, while they are negative for BSCC.

Both BSCC and BCC are made up of nests of basaloid cells with peripheral nuclear palisading, hyperchromatic nuclei and scant cytoplasm. However, in BCC retraction spaces are usually observed between the islands and stroma. BSCC commonly have abrupt foci of squamous differentiation within the nests.⁸ On IHC, BCCs are negative for EMA, and are positive for BCL2 and Ber-EP4. The cells in BSCC are positive for 34 β E12 and EMA and focally positive for CEA.¹¹

Morice WG and Ferreiro JA observed more than 95% of BSCC cases were immunoreactive with anti-high-molecular-weight cytokeratin antibody 34 β E12, whereas no reactivity was seen in cases of SCC.¹²

Treatment of choice is complete surgical excision supplemented by radiotherapy/adjuvant chemotherapy. A standard chemotherapy regimen for BSCC has not been established. A greater number of patients must be studied to determine the effectiveness of chemotherapy for BSCC of the head and neck.¹³

Conclusion: Sinonasal BSCC is a histologically distinct variant of squamous cell carcinoma with aggressive biological behaviour. Due to its biphasic nature, BSCC may not be recognized if the minor squamoid component is overlooked or not sampled, especially in small biopsies. BSCC needs to be differentiated from adenoid cystic carcinoma and small cell neuroendocrine carcinoma because of different management regimes.

References:

1. Wain SL, Kier R, Vollmer RT, Bossen EH. Basaloid squamous carcinoma of the tongue hypopharynx and Larynx: report of 10 cases. *Hum Pathol* 1986;17:1158-66.
2. Lu SY, Eng HL, Huang CC, Chien CY, Lui CC, Lin JW. Basaloid squamous cell carcinoma of the sinonasal tract: report of two cases. *Otolaryngol Head Neck Surg* 2006;134:883-5.
3. Oikawa K, Tabuchi K, Nomura M, Okuba H, Wada T, Iijima T et al. Basaloid squamous cell carcinoma of the maxillary sinus: a report of two cases of the head and neck. *Auris Nasus Larynx* 2007;34:119-123.
4. Ide F, Shimoyama T, Horie N, Kusama K. Basaloid squamous cell carcinoma of the oral mucosa: a new case and review of 45 cases in the literature. *Oral Oncol* 2002;38:120-4.
5. Wieneke JA, Thompson LD, Wenig BM. Basaloid squamous cell carcinoma of the sinonasal tract. *Cancer* 1999;85:841-54.
6. Soriano E, Faure C, Lantuejoul S, Reyt E, Bolla M, Brambilla E et al. Course and prognosis of basaloid squamous cell carcinoma of the head and neck: a casecontrol study of 62 patients. *Eur J Cancer* 2008;44:244-50.
7. Barnes L, Ferlito A, Altavilla G, MacMillan C, Rinaldo A, Doglioni C. Basaloid squamous cell carcinoma of the head and neck: clinicopathological features and differential diagnosis. *Ann Otol Rhinol Laryngol* 1996;105:75-82.
8. Vasudev P, Tadross OB and Radhi J. Basaloid squamous cell carcinoma: two case reports. *Cases Journal* 2009;2:9351.
9. Madur BP, Jambhekar NA. Basaloid squamous carcinoma simulating adenoid cystic carcinoma: diagnostic dilemma. *Oral Oncol Extra* 2006;42:227-30.
10. Emanuel P, Wang B, Wu M, Burstein DE. p63 immunohistochemistry in the distinction of adenoid cystic carcinoma from basaloid squamous cell carcinoma. *Mod Pathol* 2005;18:645-50.
11. Beer TW, Shepherd P, Theaker JM: Ber EP4 and epithelial membrane antigen aid distinction of basal cell, squamous cell and basosquamous carcinomas of the skin. *Histopathology* 2000;37:218-22
12. Morice WG, Ferreiro JA. Distinction of basaloid squamous cell carcinoma from adenoid cystic and small cell undifferentiated carcinoma by immunohistochemistry. *Hum Pathol* 1998;29:609-12.
13. Stanciulescu L, Vermesan O, Grintescu I, Enache V, Creanga C, Slavescu D et al. A rare case of basaloid squamous cell carcinoma of the maxilla. *Rom J Morphol Embryol* 2012;53:1081-85.

Legends:

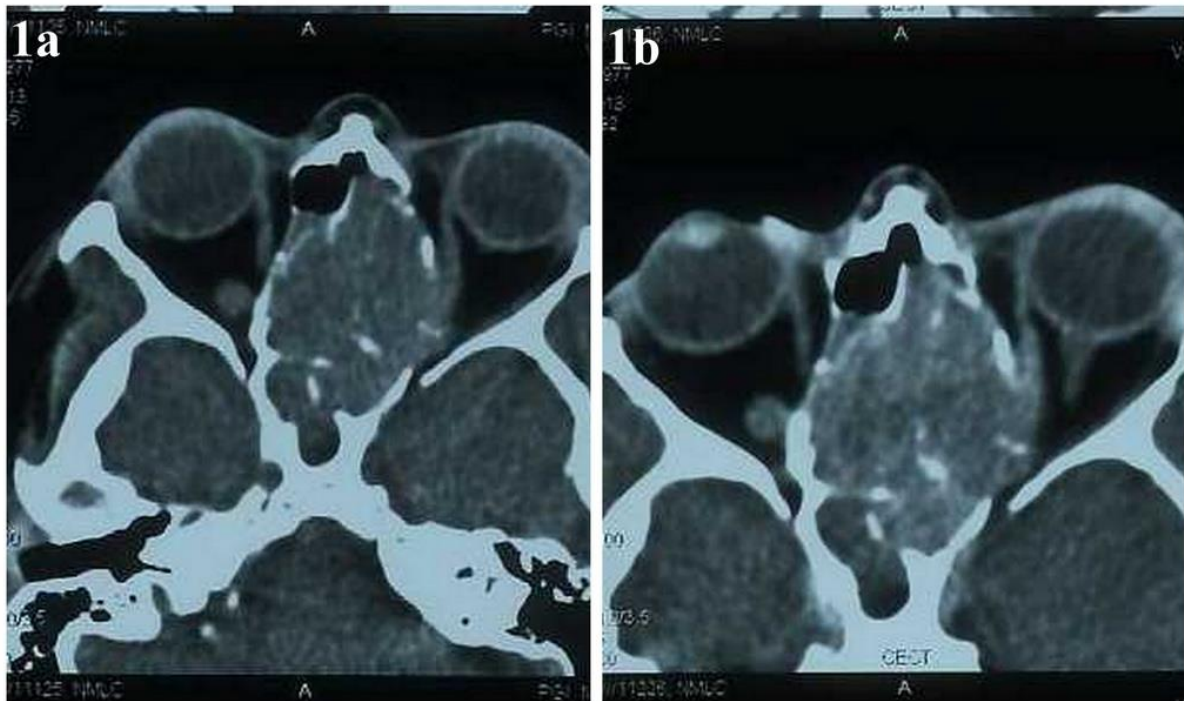


Figure 1: NCCT (1a) and CECT (1b) axial sections show a hypodense mass lesion showing enhancement on contrast in the left nasal cavity involving the ethmoidal air cells and extending posteriorly into the sphenoid sinus. The mass lesion is displacing the medial wall of left orbit and a polypoidal enhancing projection is also seen extending into right retro-orbital space.



Figure 2: Coronal CECT shows intracranial extension of the lesion through cribriform plate involving the bilateral frontal lobes.

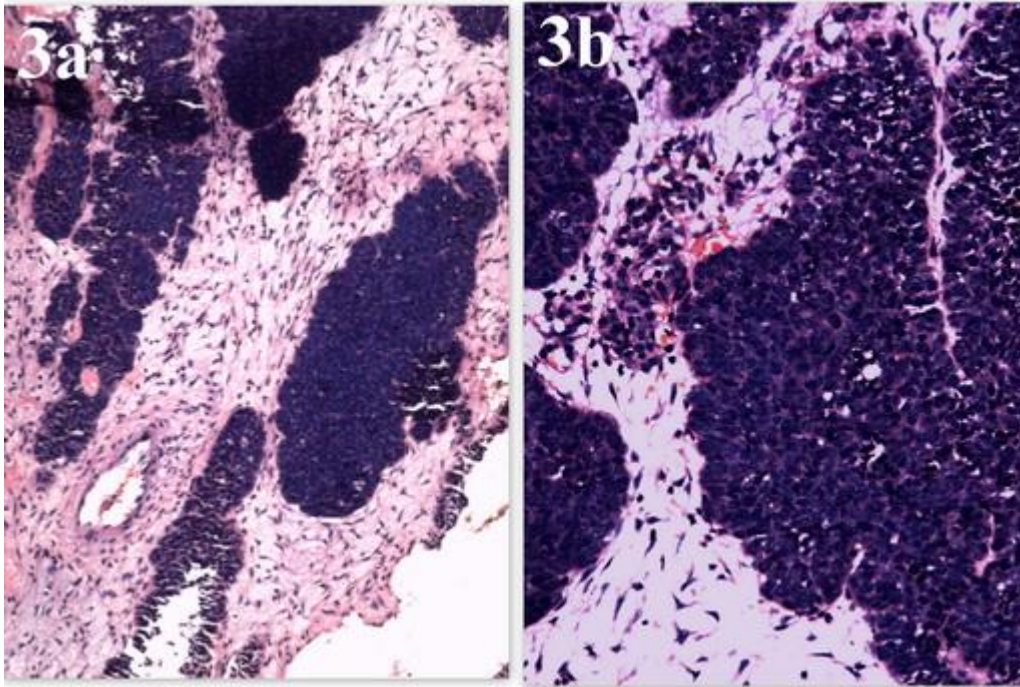


Figure 3: (3a) Nest of tumor cells arranged in cords, trabeculae and lobules.

3b) These lobules display peripheral nuclear palisading. Basaloid cells with hyperchromatic nuclei, inconspicuous nucleoli and scanty cytoplasm.

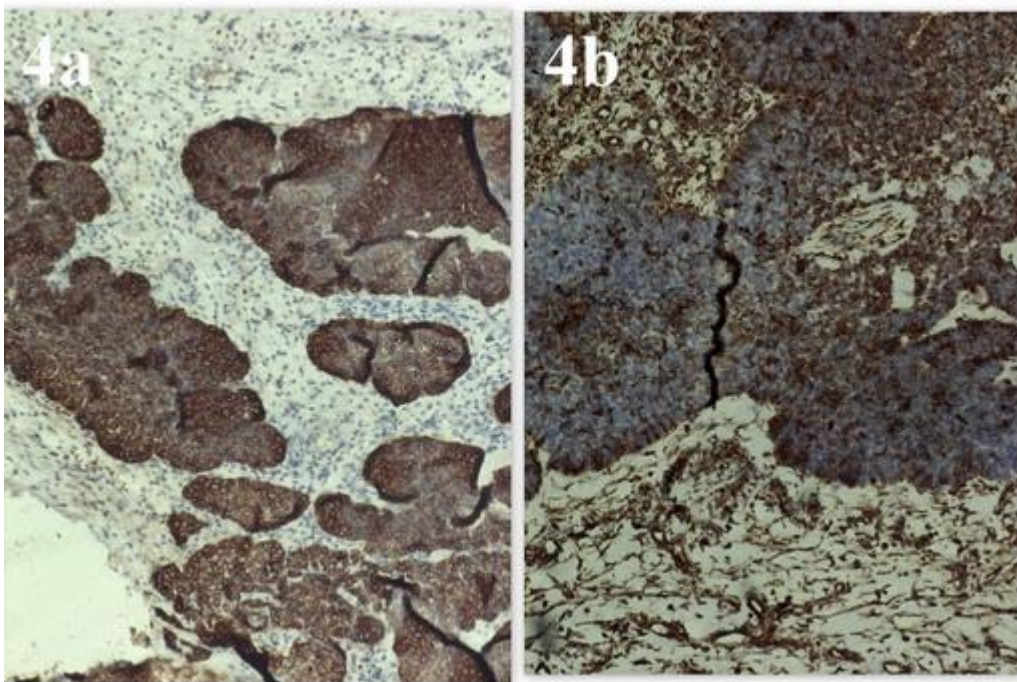


Figure 4: (4a) Tumor cells were diffuse and intense positive with anti-high-molecular-weight cytokeratin antibody 34βE12.

4b) Vimentin focal positive in tumor cells while intense positive in peritumoral stroma.

