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Case Report

Basaloid Nasopharyngeal Carcinoma: An Entity That Remains Oblivious

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Abstract

Introduction: Basaloid squamous cell carcinoma of the nasopharynx is a rare entity with only scarce cases reported in the English Literature. It is a histological variant of squamous cell carcinoma with aggressive nature and comprises of both basal cell carcinoma and squamous cell carcinoma.

Case presentation: Herein, we report on a middle-aged male who presented with left-sided spontaneous epistaxis and aural fullness with no neck node which turned out to be basaloid cell carcinoma of nasopharynx.

Discussion and conclusion: We highlight high clinical suspicion of rare variant of nasopharyngeal carcinoma although no palpable node was evident upon presentation.

Keywords: Basaloid squamous cell carcinoma; nasopharyngeal carcinoma; high–grade

Introduction

Nasopharyngeal carcinoma (NPC) is a rare neoplasm with remarkable difference in incidence throughout the world. It makes up only 0.7% of all cancers globally, however high incidence prevalence as high as 20–40 per 100 thousand persons in endemic regions such as Southeast Asia and North Africa were discovered. The World Health Organization (WHO) has classified NPC into 3 different histological types: 1) keratinizing squamous cell carcinoma; 2) non keratinizing squamous cell carcinoma, which is further subdivided into differentiated and undifferentiated subtypes; and 3) basaloid squamous cell carcinoma. Basaloid squamous cell carcinoma was first described in 1986 by Wain et al. It was initially reported histologically as high–grade variant of squamous cell carcinoma which typically arises in the upper aerodigestive tract. It is considered an aggressive subtype, with patients often presenting in the advanced stages or with metastases. Occurrence of nasopharyngeal basaloid squamous cell carcinoma (NBSCC) is considered extremely rare with only limited literature exists on the epidemiology and survival characteristics. Only six cases have been reported till date in the English literature.

Case Presentation

A 49–year–old Chinese gentleman, who is an active smoker, presented with two–month history of left–sided spontaneous painless epistaxis with left aural fullness. Patient claims epistaxis is minimal amount with unknown trigger and resolves spontaneously. Aural fullness on the other hand is persistent although no reduced hearing is noted. There was no associated otalgia, otorrhea, tinnitus or vertigo. No recurrent rhinorrhea, sneezing, facial tenderness or headache were mentioned. He denied any constitutional or obstructive symptoms or any family history of malignancy.

On examination, patient was well–built, comfortable under room air. Otoscopic examination revealed no abnormality. Rigid nassoendoscopy showed fungating mass occupying left nasopharynx, obliterating left fossa of
Rosenmuller not crossing midline with all other subsites of nasal cavity intact (Figure 1). Biopsy was taken under local anesthesia. Oral cavity and oropharynx examinations was normal. Neck examination revealed no palpable neck nodes. All cranial nerves and systemic examination were intact. Pure tone audiometry revealed normal hearing bilaterally with type A tympanometry bilaterally.

Histopathological examination of the nasopharynx biopsy demonstrated basaloid cells disposed in sheets, lobules and anastomosing trabeculae, desmoplastic to myxoid stroma. The tumour cells have round to ovoid nuclei with prominent nucleoli and expresses HMWCK 5/6 and diffusely express p63 which is consistent with basaloid type of nasopharyngeal carcinoma (Figure 2). Contract enhanced computed tomography showed lesion in left nasopharynx extending into ipsilateral parapharyngeal space and sphenoid sinus with no bony erosion. Multiple lung nodules were noted (Figure 3). Hence, patient was diagnosed as Basaloid type of NPC T3N0M1. Patient was counselled and referred to National Cancer Institute for chemoradioteraphy. He was planned for palliative chemotherapy with 5–Flourouracil & Cisplatin regime for 6 weeks. However, patient defaulted follow–up after 1 cycle.

Discussion

NPC typically arises from epithelial cells in nasopharynx, consisting of stratified squamous and pseudostratified ciliated columnar epithelium. NPC can locally remain in nasopharynx, or may extend to lateral nasopharyngeal walls, posterosuperiorly to base of skull, palate and nasal cavity, or inferiorly to oropharynx. The tumour may metastasize to cervical lymph nodes, which is presented in 60 – 90% of cases. Other symptoms include epistaxis, otitis media, hearing loss, trismus, pain, cranial nerve palsies and soft palate paresis.

Generally, biopsy of primary lesion remains as first line to achieve diagnosis. In primary setting, treatment of NPC is usually via definitive radiation in stage I/II and concurrent chemoradiation with or without induction/adjuvant chemotherapy in stage III/IV. Basaloid squamous cell carcinoma (BSCC) is a histologically distinctive variant of squamous cell carcinoma comprising basal cell carcinoma and squamous cell carcinoma.

NBSCC is an extremely rare entity with only limited cases reported in English literature. Recently, a study by Unsal et al. revealed only six overall incidences per 100 million patients in the United States. Strong similarities to with keratinised and non–keratinised type of NPC were demonstrated involving ethnic and geographical
distribution in which, white males were more affected compared to Asians or Pacific Islanders. Age distribution between 40 to 79 years were noted in patients with NBSCC.

NBCC are high grade tumours, which consists of poorly differentiated or undifferentiated tumour. Unlike keratinising and non-keratinising type of NPC, NBSCC has low propensity for cervical node metastasis, although it can present at advanced stage notably stage III/IV. (Unsal et al noted that NBSCC has similar survival rate as NPC 1 and 5 years, but poorer prognosis is seen after 10 years).

Close association with alcohol consumptions, smoking and radiation is seen for NBSCC. Wan et al, there was association noted between EBV and NBSCC, however this is not found if the BSCC is elsewhere. This study also suggested that NBSCC is closely related to the undifferentiated type of NPC, in terms of overall presentation, clinical stage and behaviour.

Due to the extreme low incidence, there are still uncertainties in determining the line of differentiation or pathogenesis of NBSCC. Wain et al suggested that totipotent primitive cells, which are located at base of pseudostratified columnar epithelium or in proximal salivary gland ducts of larynx, hypopharynx and tongue, serves as precursor for basaloid squamous cell carcinoma as well as squamous cell carcinoma, adenocarcinoma, oat cell carcinoma and reserve cell carcinoma. Generally, differential diagnosis for NBSCC includes adenoid cystic carcinoma and small cell carcinoma. In our case, histopathology showed basaloid tumour characterised by peripheral palisading of the tumour cells which strongly expressed HMWCK 5/6 and P63 but did not express CD117 and CK7. CD117 and CK 7 are typically expressed in basaloid neoplasm of salivary glandular origin. Review of the literature showed that adenoid cystic carcinoma cells demonstrated S100 protein-positivity while in NBSCC this were nonreactive. In addition, adenoid cystic carcinoma also showed greater reactivity to smooth muscle actin and vimentin compared to BSCC.

The differentiation of NBSCC from squamous cell carcinoma (variants with glandular or pseudo glandular formation) is comparatively less arduous. BSCC showed variable reactivity to neuroendocrine immunohistochemistry such as chromogranin, synaptophysin and neuron specific enolase, which is consistently positive in squamous cell carcinoma. However, BSCC differs from squamous cell carcinoma whereby pattern of cytokeratin expression is different.

Treatment of this entity remains a dilemma till date owing to the meagre number of cases reported. BSCC of the head and neck includes complete surgical excision with post-operative radiotherapy. Having said that, NBSCC is treated with chemoradiotherapy as NPC. In advanced NBSCC, standard of care has not been established yet. Our patient was treated with chemoradiotherapy.

Basaloid squamous cell carcinoma has the lowest survival (<10%) compared to other subtypes of squamous cell carcinoma. Poor prognostic indication for NPC includes advanced clinical stage, cranial nerve involvement, absence of Epstein Barr virus and keratinising histology. Having said that, prognostic marker for NBSCC has not been widely mentioned due to scarce reported cases.

**Conclusion**

NBSCC despite being a rare entity requires attention due to its extreme low incidence. Low propensity for cervical node as the conventional NPC, leads to this entity being missed. Chemoradiotherapy remains gold standard. Prognosis of NBSCC is poor.

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