Primary Mucosal Melanoma of the Nasopharynx : A Case Report

Case Report

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ABSTRACT

Background: Mucosal melanoma is an extremely rare pathology. Nasopharyngeal localization is exceptional. Management is challenging with advances in treatment approaches, however prognosis remains poor.

Case presentation: We report a 60-year-old man presented with chronic unilateral nasal obstruction and homolateral hearing loss. Physical examination found nasopharyngeal mass with homolateral nasal fossa extension, and latero-cervical lymph node. CT scan confirmed nasopharyngeal tumor lesion with presence of right jugulo-carotid lymph node. Histopathological examination, on a biopsy piece, concluded in nasopharyngeal melanoma. Concomitant radiochemotherapy was performed. After one year follow-up, the patient died from brain metastasis.

Conclusions: Nasopharyngeal mucosal melanoma is an uncommon entity with unknown etiology. Poor prognosis correlated with frequent metastasis requires early diagnosis and appropriate management.

Key Words: Head and neck neoplasms, mucosal melanoma, nasopharyngeal neoplasms.

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BACKGROUND

Mucosal melanoma is an uncommon form of cancer[1,2,3,4]. Nasopharyngeal localization is exceptional in this pathology[4,5]. Delay in diagnosis and management issues make the prognosis generally very poor[1]. The authors report a case of nasopharyngeal mucosal melanoma in a senegalese male.

CASE PRESENTATION:

A 60 years old man, presented with a right nasal obstruction, evolving for three months, associated with recurrent right sided epistaxis, ipsilateral hypoacousia and tinnitus. Physical examination showed one single right laterocervical lymph node measuring 1,5 cm in its greatest dimension.

Nasal endoscopy revealed a black coloured lesion in the right lateral wall of the nasopharynx, extended to the homolateral nasal fossa (Fig. 1).

Otoscopic examination demonstrated a blackish right retro-tympanic lesion (Fig. 2) without effusion, retraction or perforation.

No skin lesion was found.

Moderate right-sided conductive hearing loss was noted on audiometry.

CT scan showed a tumor process occupying the right lateral wall of the nasopharynx with ipsilateral laterocervical lymph node (Fig. 3 and 4).

Histological examination on a biopsy piece described a tumor proliferation made of round and spindle cells with eosinophilic cytoplasm encrusted by brownish melanin pigments (Fig. 5). Pathology report demonstrated nasopharyngeal malignant mucosal melanoma.

Staging assessment found no metastatic extension. The lesion was classified T2N1M0 (AJCC 7th edition classification).

Concomitant radio-chemotherapy was administrated.

Patient firstly did well. But, fifteen months later, he was found with a right side zone II N3 node involving internal carotid artery, followed by a right hemi-corporal deficit related to multiple brain metastasis. Due to poor general condition and non-resectable N3 metastatic node, he received palliative chemotherapy and supportive care. The patient died four months later.

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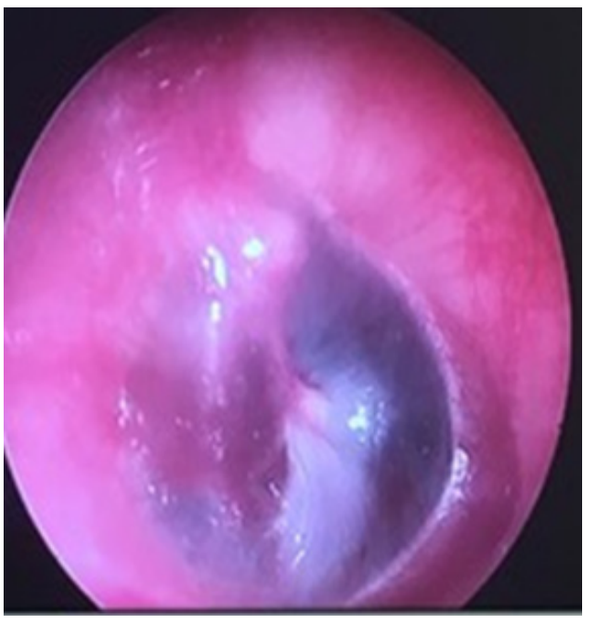

Fig. 1: Otoscopic view of retro-tympanic black lesion

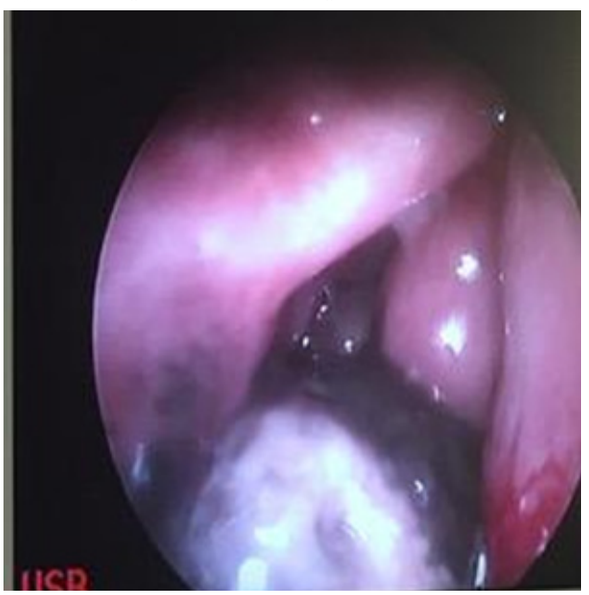

Fig. 2: Endoscopic view of a blackish polyploid mass in the right nasal fossa.

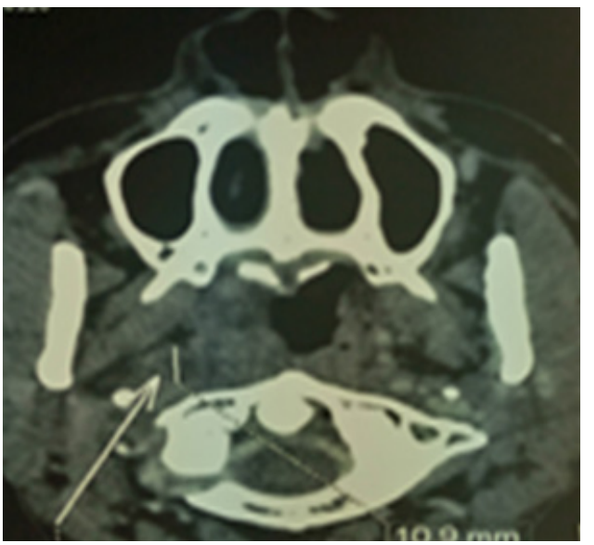

Fig. 3: Axial CT scan image showing soft tissue mass of the right lateral pharyngeal wall with cervical lymph node (Arrow).

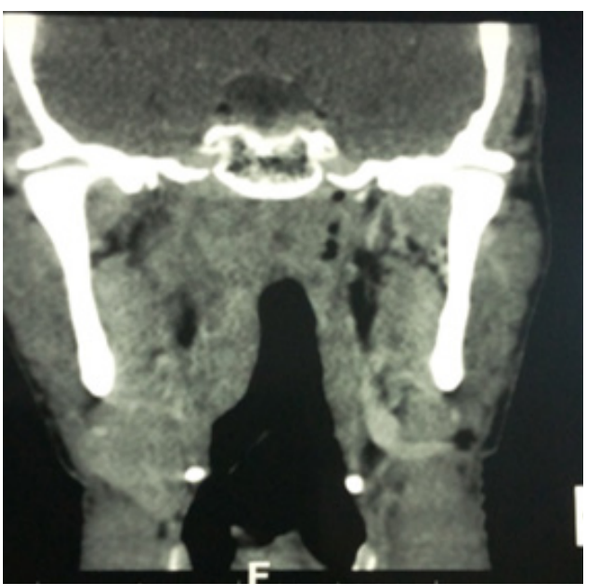

Fig. 4: coronal CT scan image showing soft tissue mass of the right lateral pharyngeal wall

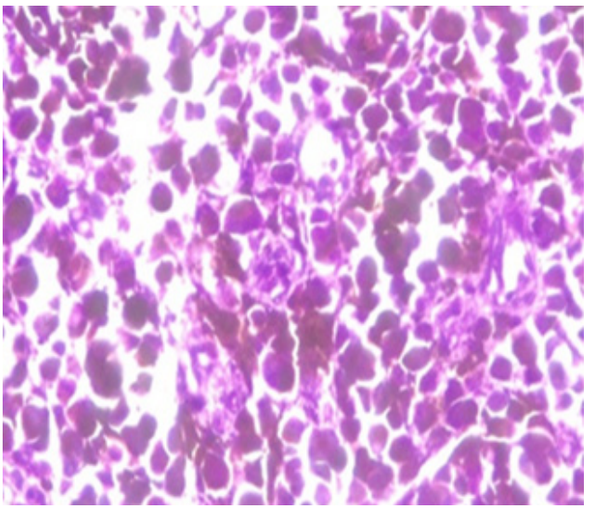

Fig. 5: Primary mucosal melanoma Polygonal tumor cells with eosinophilic cytoplasm and melanin pigments. (HE x 40).

DISCUSSION

Mucosal melanoma is a rare entity[1,2,3,4]. It accounts about 0.2 to 8% of melanomas[2,4] and represents 0.5 to 1% of all head and neck cancers[3]. Nasosinusal and oropharyngeal melanomas are the most common[2]. Nasopharyngeal localization is exceptional[4, 5].

The etiopathogeny of nasopharyngeal mucosal melanoma has not yet been elucidated[2,6]. Several risk factors were implicated, including chronic volatile carcinogens inhalation (formaldehyde) and chronic smoking[2,7].

The disease appears in people between 50 to 70 years with a slight male predominance[3,5]. Our patient was 60 years old.

Symptoms are similar as in all nasopharyngeal tumors. The lesion manifests by rhinological and/ or otological signs, which are more evocative when unilateral. Our patient presented with a right nasal obstruction with ipsilateral hearing loss. However, the diagnosis is often delayed compared to symptom onset[1].

Endoscopy reveals a budding or polyploid nasopharyngeal lesion, which is pigmented with varying degrees of coloration ranging from black to white[2,4].

Diagnosis of melanoma is histopathological[3,4].

Microscopically, melanoma is characterized by neoplastic melanocytes with variable phenotypes (rounded, spindle-shaped, globular) arranged in leaf form, in clusters, nests or in layers[2,4].

Immunohistochemical study is positive for melanoma antigens (Vimentin, S100, HMB45, Melan-A)[2,3,4,7]. Immunostaining is negative for cytokeratinine and for epithelial membrane antigen[2,3].

Diagnosis of primary nasopharyngeal melanoma can only be accepted after eliminating other localization, especially cutaneous melanoma[4]. In our patient, no other site was found.

CT scan and MRI complete clinical manifestations, endoscopy findings and histopathology for patient tumor staging[2,3].

Surgery is the gold standard tratment for melanomas[1,2,8,9]; but complete tumor removal with healthy margins remains difficult due to nasopharyngeal complex anatomy, close proximity of vital structures and delayed diagnosis of these lesions at an advanced stage[1,2,6].

Surgery is often limited to neck dissection for cases with local lymph node metastasis[7].

Irradiation alone or irradiation with surgery or chemotherpy have been described[1,2,7]. Irradiation can improve locoregional control, without really improving on survival rates[1,2,3,6,9].

Chemotherapy is not well standardized in the management of mucosal melaomas[2].

Immunotherapy and targeted therapy, which have revolutionized cutaneous melanomas treatment, are now used in the management of unresectable or metastatic mucosal melanomas[2,6,7,9].

However, prognosis remains poor[1,2,3]; 5-year overall survival rate is 10 to 30% with a high rate of local recurrence and metastasis[1,2,3]. Average survival is 24 months, and most patients die from lung or brain metastasis. This is the case with our patient.

CONCLUSION

Primary mucosal melanoma is a rare condition for which etiopathology remains unknown. Nasopharyngeal localization is exceptional. Management is challenging and prognosis remains poor. Early diagnosis and adequate management are required to improve survival rates. Targeted therapies are a promised alternative for advanced stages and metastasis.

CONFLICT OF INTEREST

There are no conflicts of interest.

REFERENCES

1. Azoury SC, Crompton JG, Straughan DM, Klemen ND, Reardon ES, Beresnev TH, *et al*. Unknown primary nasopharyngeal melanoma presenting as severe recurrent epistaxis and hearing loss following treatment and remission of metastatic disease: A case report and literature review. Int J Surg Case Rep 2015;10:232-5.

2. Ascierto PA, Accorona R, Botti G, Farina D, Fossati P, Gatta G, *et al*. Mucosal melanoma of the head and neck. Crit Rev Oncol Hematol 2017;112:136-52.

3. Lietin B, Montalban A, Louvrier C, Kemeny J-L, Mom T, Gilain L. Sinonasal mucosal melanomas. Eur Ann Otorhinolaryngol Head Neck Dis. mai 2010;127(2):70-6.

4. Mekni A, Bouraoui S, Amdouni N, Haifa TG, Kchir N, Zitouna M, *et al*. Une tumeur rhinopharyngée rare. Une Tumeur Rhinopharyngée Rare. Ann Pathol 2004;24(4):375-6.

5. Mardi K. Primary mucosal malignant melanoma of nasopharynx: a rare case report. J Cancer Res Ther 2014;10(2):416-8.

6. Thierauf J, Glück A-M, Plinkert P, Veit JA, Hoffmann TK, Körber A, *et al*. Mucosal melanoma of the cranio-facial region: Surgical challenges and therapeutic options. Auris Nasus Larynx 2019;46(2):252-9.

7. Troussier I, Baglin A-C, Marcy P-Y, Even C, Moya-Plana A, Krengli M, *et al*. [Mucosal melanomas of the head and neck: State of the art and current controversies. Bull Cancer 2015;102(6):559-67.

8. Dhouib H, Mnejja M, Bouayed W, Chakroun A, Boudawara T, Charfeddine I, *et al*. Formes histologiques particulieres du cancer du nasopharynx. J Tun ORL 2007;19:47-51.

9. Moreno MA, Hanna EY. Management of mucosal melanomas of the head and neck: did we make any progress? Curr Opin Otolaryngol Head Neck Surg 2010;18(2):101-6.