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Case Report

Amelanotic mucosal melanoma of the left sinonasal cavity: a case report

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ABSTRACT

Mucosal melanoma is rare type of melanoma accounted only 0.8-3.7% of melanoma. It defined as melanoma which occurs in mucous membrane, oral and nasal cavities, conjunctiva, genital sites, and rarely other mucosae. Head and neck melanoma accounted 50% among other sites. Recent studies, amelanotic subtype account 13.2% of all sino-nasal melanoma. Nasal cavity melanoma usually occurs on lateral nasal area especially in middle turbinate and inferior turbinate. Among other location, sino-nasal originated primary lesion was more deadly compared to other. We reported a case of amelanotic melanoma. 56 years old Asian Female presented with mass in left nose. The patient also complaint of recurring epistaxis, facial pain, epiphora, and blurred vision. After several follow up investigation, a diagnosis of amelanotic melanoma with orbit involvement was made. Subsequently patient underwent surgery for tumor removal. Although tumor has been removed, patient had to continue with adjuvant therapy for following month. Meticulous identifying and confirmation is an important factor which provide better understanding for the clinician in diagnosing.

Keywords: Mucosal melanoma, Amelanotic melanoma, Head and neck, Sino-nasal

INTRODUCTION

Mucosal melanoma is rare type of melanoma accounted only 0.8-3.7 % of melanoma.[1] It defined as melanoma which occurs in mucous membrane, oral and nasal cavities, conjunctiva, genital sites, and rarely other mucosae.[2] Head and neck melanoma accounted 50% among other sites.[3]

Recent studies, amelanotic subtype account 13.2% of all sino-nasal melanoma. Nasal cavity melanoma usually occurs on lateral nasal area especially in middle turbinate and inferior turbinate.[4]

Among other location, sino-nasal originated primary lesion was more deadly compared to other.[5]

CASE REPORT

This was a case report of an unusual cases of amelanotic melanoma that presented to the outpatient clinic with advanced stage and non-specific clinical presentations. The rare and complexity of the medical and family history and clinical symptoms makes the diagnosis difficult, associating with delayed diagnosis and treatment. Here we study the clinical presentations, risk factor, and the diagnostic challenges in the diagnostic of amelanotic melanoma.

Case presentation

A 56-year-old Asian female presented to otolaryngology division clinic with nasal mass in left nose in July 2021. The mass firstly noticed around 5 months ago, then enlarge

and occupied nasal cavity. The patient also reported recurring episodes of epistaxis, facial pain, epiphora, and blurred vision. She had no history of allergic rhinitis or chronic sinusitis, coagulopathy, illicit drug use, rheumatologic or any other systemic disease. There is also no history of recent infection. On physical examination, nasal dorsum was symmetric to no noticeable deformities. The nasal ala were symmetry and external valves were patent. There was no tenderness on maxillary and frontal sinus during palpable examination. No noticeable deformities were found in the septum wall. The right nasal cavity was patent with no noticeable abnormalities. In left nasal cavity was completely obstructed with mass with pink pale and moist mucous; the mucosa along lateral wall were inflamed with dried blood and crusting (Figure 1). For symptomatic patient given pain management only.

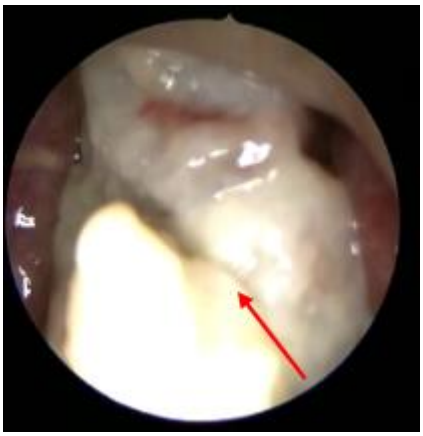

Figure 1: Endoscopic.

Before coming to our clinic patient already undergo biopsy with result of round blue cell tumor squamous cell carcinoma and non-Hodgkin lymphoma with immunohistochemistry suggest chronic infection with granulation. The patient then undergo another biopsy and immunohistochemistry for confirmation in August 2021. The biopsy result of non-Hodgkin lymphoma with large cell type (Figure 2). Then proceed with immunohistochemistry of LCA/CD45, CD3, CD20, Ki-67, Vimentin, S100, Synaptophysin, HMB45, EMA, and Melan-A with positive results in Vimentin, S100, HMB45, Melan-A. This finding result in the diagnosis of amelanotic melanoma.

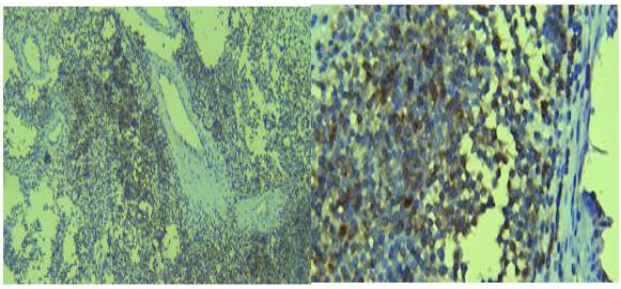

Figure 2: Melan A stain at 100 (left) and 400 magnification (right).

Mucosal melanoma is very uncommon condition and malignant disease. Because of its low incidence and hard to diagnose, there is no sufficient guideline for its treatment and diagnose. Multiple modalities treatment such as radical removal, radiotherapy and chemotherapy does make a difference but still doesn't effectively affect the patient survival. Ultimately, MM is an important differential in patient when nasal mass, and recurring episodes of epistaxis were presented.

Subsequently after physical examination patient also underwent CT scan which revealed the lesion already affected maxillary, ethmoidales, and sphenoid sinuses, with no metastasis on lymph nodes. The mass also expanded to the left eye orbit (Figure 3). Hence, we referred patient to eye department. After examination, they concluded proptosis of left eye with papilla edema and pre-papil atrophic.

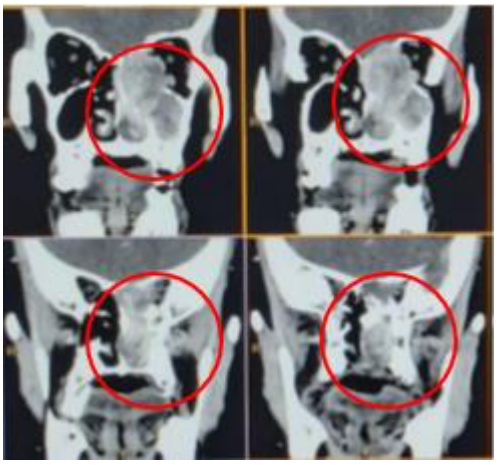

Figure 3: Axial CT-scan.

In December patient undergo surgery for mass removal and orbitotomy and exenteration from eye department. Following the surgery patient to be scheduled for adjuvant radiotherapy and chemotherapy.

DISCUSSION

Classic presentation of MM lesions are dark pigmented, polypoid lesions with irregular borders. Nasal obstruction, mucosal bleeding, epistaxis, facial pain, rhinorrhea and even parathesias are usually presented in classic cases.[6] In recent systematic review by Pontes et all on sino-nasal melanoma; epistaxis, nasal obstruction, and blurry vision were most common observed in patient with MM in sino-nasal. Same symptoms were observed in this patient.[4] Blurry symptoms in our patient due to mass already expanded to left orbit, but we cannot conclude any neural involvement.

The diagnosis of mucosal melanoma can be challenging. In our patient alone, numerous immunohistochemistry was used to ensure our diagnosis. But vimentin, S-100, HMB-45, and Melan-A were the most common factor leading to

diagnosis of amelanotic melanoma.[4] In the latest systematic review, stage I and II were more likely to be observed.[4] Contrary to the previous findings that patient usually already at stage III and stage IV when first diagnosed.[7] These findings associated with the presence of recurring epistaxis and nasal obstruction which help specialist to further investigation.[8]

Melanoma already have higher death rate and poorer survival rate.[9] MM have better outcome other than other in term of general type, but in sino-nasal is still inconclusive. Among other location, sino-nasal originated primary lesion was more deadly compared to other.[5] Other prognostic factors such as clinical presentation such as age more than >67.6 years, middle turbinated primary tumor, advanced disease, radiotherapy alone, recurrence, presence of distant metastatic associated with less survival rate.[4] Another finding, elevated lactate dehydrogenase (LDH) level and higher performance score were associated with poorer survival rate.[5] Amelanotic melanoma have higher recurrence index than pigmented melanoma. But recurrence does differ depends on several risk factors, the higher stage tends to occur more, other factor are age more than >67.6 and gender. Although amelanotic melanoma occurrence higher in female, its likely to recurrence in male patient.[8] Chemotherapy can be indicated if there were distant metastasis and in unresectable patients. But the therapeutic effect of chemotherapy on mucosal melanoma in head and neck remain inconclusive.[10] The prognosis of mucosal melanoma remains uncertain leaning to poorer outcome due to its malignant nature. Even with multiple modalities treatment the prognosis and survival rate remain despicable. Hence further research in diagnosis and treatment were needed to achieve better outcomes

CONCLUSION

Amelanotic mucosal melanoma is very uncommon condition and malignant disease. Because of its low incidence and hard to diagnose, there is no sufficient guideline for its treatment and diagnose. Multiple modalities treatment such as radical removal, radiotherapy and chemotherapy does make a difference but still doesn't effectively affect the patient survival. Ultimately, MM is an important differential in patient when nasal mass, and recurring episodes of epistaxis were presented. The diagnosis of mucosal melanoma can be challenging and vimentin, S-100, HMB-45, and melan-A were the most common factor leading to diagnosis of amelanotic melanoma.

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REFERENCES

1. Spencer KR, Mehnert JM. Mucosal Melanoma: Epidemiology, Biology and Treatment. Cancer Treat Res. 2016;167:295-320.
2. Elder DE, Bastian BC, Cree IA, Massi D, Scolyer RA. The 2018 World Health Organization Classification of Cutaneous, Mucosal, and Uveal Melanoma: Detailed Analysis of 9 Distinct Subtypes Defined by Their Evolutionary Pathway. Arch Pathol Lab Med. 2020;144(4):500-22.
3. Breik O, Sim F, Wong T, Nastri A, Iseli TA, Wiesenfeld D. Survival Outcomes of Mucosal Melanoma in the Head and Neck: Case Series and Review of Current Treatment Guidelines. J Oral Maxillofac Surg. 2016;74(9):1859-71.
4. Pontes FSC, de Souza LL, de Abreu MC, Fernandes LA, Rodrigues ALM, do Nascimento DM, et al. Sinonasal melanoma: a systematic review of the prognostic factors. Int J Oral Maxillofac Surg. 2020;49(5):549-57.
5. Hahn HM, Lee KG, Choi W, Cheong SH, Myung KB, Hahn HJ. An updated review of mucosal melanoma: Survival meta-analysis. Mol Clin Oncol. 2019;11(2):116-26.
6. Hasan S, Jamdar SF, Jangra J, Al Beaiji SM. Oral malignant melanoma: An aggressive clinical entity - Report of a rare case with review of literature. J Int Soc Prev Community Dent. 2016;6(2):176-81.
7. Castelhano L, Correia F, Raposo D, Campos A, Melo M, Pereira S, et al. Malignant tumors of the nose and paranasal sinuses: a retrospective review of a Portuguese cancer center's experience. Head and Neck Tumors (HNT). 2021;11:10-7.
8. Dréno M, Georges M, Espitalier F, Ferron C, Charnolé A, Dréno B, Malard O. Sinonasal mucosal melanoma: A 44-case study and literature analysis. Eur Ann Otorhinolaryngol Head Neck Dis. 2017;134(4):237-42.
9. Strazzulla LC, Li X, Zhu K, Okhovat JP, Lee SJ, Kim CC. Clinicopathologic, misdiagnosis, and survival differences between clinically amelanotic melanomas and pigmented melanomas. J Am Acad Dermatol. 2019;80(5):1292-8.
10. Verma R, Lokesh KP, Gupta K, Panda NK. Sinonasal amelanotic malignant melanoma – A diagnostic dilemma. Egyptian J Ear Nose Throat Allied Sci. 2015;16(3):275-8.
11. Xu QQ, Lai YZ, Huang ZL, Zeng ZY, Zhang YN, Ou RY, et al. Clinical outcomes and patterns of failure of head and neck mucosal melanoma treated with multiple treatment modalities. Radiat Oncol. 2021;16(1):138.

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