Volume : 12; Issue1; pp 41-44
DOI: http://doi.org/10.4038/cjo.v12i1.5353

Epithelioid Hemangioendothelioma arising from nasal cavity: A Rare Case

Hettiarachchi KS[1], Thuduvage VS[2], Perera RNA[3], Dissanayake HTRW[4], Wickramarathna KBM[5], Widanage WKDM[6], Domingoarachchi D[7]

[1] - ENT Medical officer, University Hospital of Kotelawala Defense University, Sri Lanka.
[2] - Consultant ENT Surgeon, University Hospital of Kotelawala Defense University, Sri Lanka.
[3] - ENT Medical officer, University Hospital of Kotelawala Defense University, Sri Lanka.
[4] - ENT Medical officer, University Hospital of Kotelawala Defense University, Sri Lanka.
[5] - ENT Medical officer, University Hospital of Kotelawala Defense University, Sri Lanka.
[6] - ENT Medical officer, University Hospital of Kotelawala Defense University, Sri Lanka.
[7] - Consultant ENT Surgeon , Apeksha Hospital , Sri Lanka

Abstract

Introduction

Epithelial Hemangioendothelioma is an uncommon malignancy of soft tissues, mostly occur in females in their 4th and 5th decades of life. It is very rare to occur in nasal cavity and prone to have local recurrences and distant metastasis. Here we present a 37 year old male patient with epithelioid hemangioendothelioma in nasal cavity with extension to the surrounding structures.

Case report

Patient presented to University Hospital - Kotelawala Defense University with a history of right side nasal obstruction, facial pain and headache. Imaging studies showed large focal lesion with extension to the surrounding structures and impending intracranial extension. Endoscopy and neuronavigator guided tumor excision done and histopathology report came as epithelioid hemangioendothelioma.

Conclusion

Radical local excision is the key important step in the management of Epithelioid hemangioendothelioma.

Key words: Epithelial Hemangioendothelioma, nasal cavity, Radical local excision, nasal Malignancy

Funding: None

Competing interest: None

Correspondence: Dr KS Hettiarachchi (kanishkasajeevi@gmail.com)

Accepted Date: 24.01.2023 Published Date: 19.03.2023

p-ISSN : 2012 - 855x, e-ISSN: 2576-2040
Ceylon Journal of Otolaryngology

Introduction

Epithelioid hemangiosarcoma is first described by Weiss and Enzinger in 1982[1]. It is an uncommon tumor, mostly occurring in soft tissues of extremities and lungs and rare to occur in head and neck area[2]. Reported cases of epithelioid hemangiomas in nasal cavity are rare in the literature.

Here we present a case of epithelioid hemangioendothelioma in nasal cavity with extension to the maxillary sinus, ethmoidal air cells, extra conal compartment, chaonal space and impending intracranial extension. Endoscopy and Neuronavigator assisted excision of the lesion was performed and achieved a favorable outcome.

Case report

A 37 year old male patient, presented to the ENT unit of University Hospital of Kotelawala Defense University (UHKDU), with a history of right side nasal obstruction for 2 months duration. It was associated with right side eye pain, facial pain and headache. There was no history of symptoms suggestive of upper respiratory tract infection or epistaxis. Anterior rhinoscopy showed a red colored growth in right nostril. His right eye's adduction and depression were impaired, but the vision was normal.

A contrast enhanced CT scan of the nose, paranasal sinuses, orbits and brain was requested and it showed avidly and heterogeneously enhancing large solid focal lesion (71×58×41mm) filling the right nasal cavity with marked extension in the adjacent anatomical structures, laterally into the right maxillary sinus via eroded axillary osteum, superiorly into right ethmoidal air cells abutting the cribriform plate of the ethmoid bone, superolateral extension into extra conal compartment of the right orbit abutting the inferior rectus muscle and medial rectus muscle, posteriorly into the choanal space in both sides and nasopharynx abutting the right pre vertebral muscle (longus colli). Furthermore, it showed an impending intracranial extension through the cribriform plate of ethmoid bone. An MRI of brain, nose and paranasal sinuses was requested and found to have a mass lesion filling up the right nasal cavity with extension in to the R/maxillary sinus, R/frontal sinus, nasopharynx and extra conal compartment of R/orbit with associated intermediate type aggressive bone remodeling/destruction and interval growth compared to previous CT study. MRI brain was normal except for erosions at the cribriform plate and crista galli.

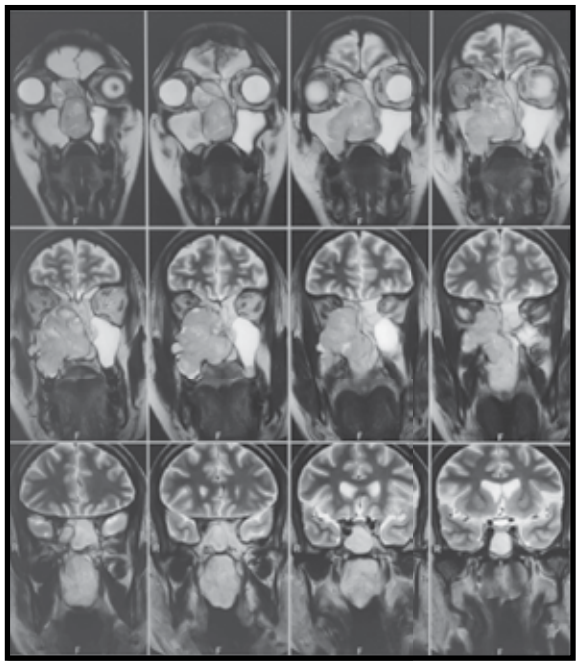

Figure 1.MRI appearance of the mass lesion filling up the right nasal cavity.

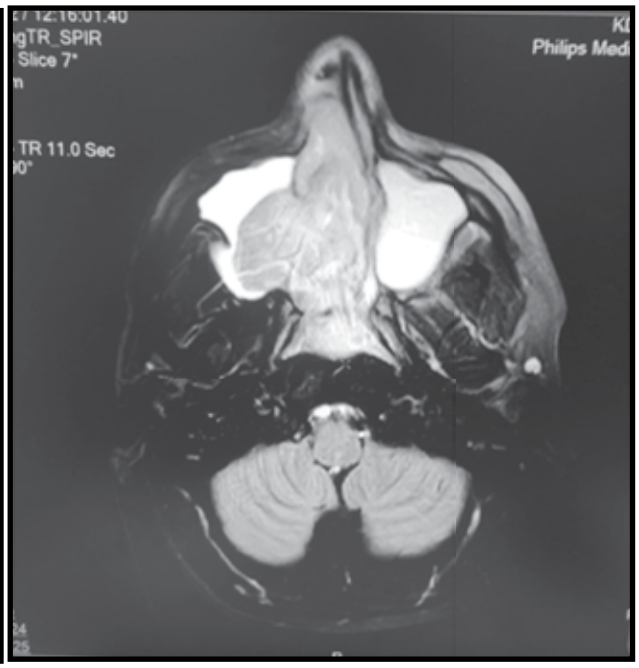

Figure 2.MRI showing mass lesion extension into the maxillary sinus

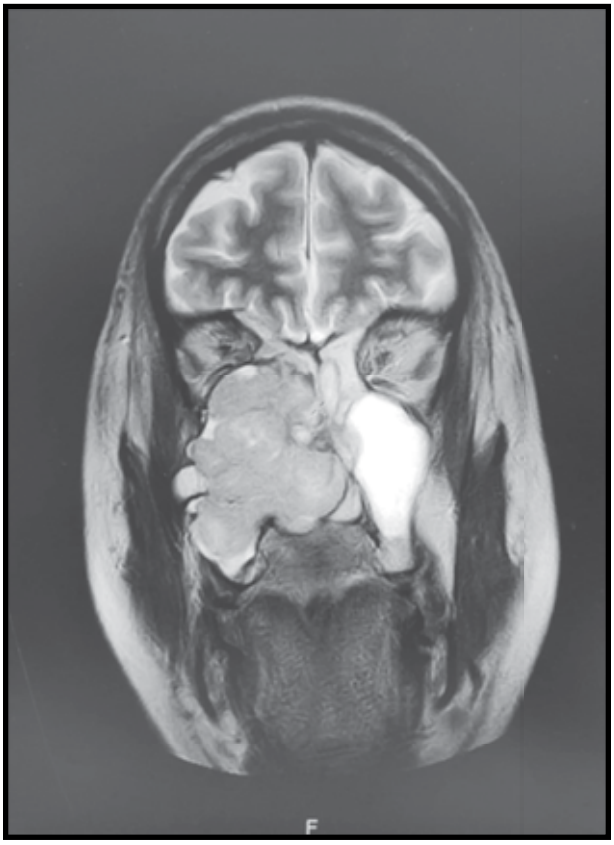

Figure 2.MRI showing mass lesion extension into the maxillary sinus

Humphrey visual field (HVF) 24-2 test was done to assess his vision and it was found to be normal. Blood investigations including full blood count, liver function tests, and renal function tests and clotting profile were also normal.

Endoscopy and Neuronavigator assisted excision of right side nasal lesion performed under general anaesthesia by the Consultant ENT surgeon in collaboration with the Consultant Ophthalmologist and Consultant Neurosurgeon. The histopathology report was compatible with a vascular tumour in favor of an Epithelioid Heamangioendothelioma.

Patient was referred for oncology team for further management.

Discussion

Epithelioid Haemangioendothelioma (EHE) is a vascular tumor occurring mostly on soft tissues of extremities and lungs[3]. It is characterized by the proliferation of the endothelial cells with an epithelioid morphology[1]. It is generally occurs in adults preferably in 4th and 5th decade of life. Females are more commonly affected than males[4]. Risk factors have not been identified but, possible associations with trauma, therapeutic radiation and hormonal factors have been considered[5]. EHEs are extremely rare to occur in nasal cavity and only few cases are reported in the literature.

Clinical diagnosis is difficult in EHE as the only symptom is progressively enlarging, cutaneous, tender, solitary skin colored mass next to a vessel course[6]. However the cutaneous manifestation may be variable such as solitary, multiple, eruptive, and ulcerative skin lesions[7,8,9,10] . The differential diagnosis for EHE include hemangioma, pyogenic granuloma, angiomyoma, epithelioid sarcoma, hemangioendothelioma and epithelioid angiosarcoma.

The treatment of choice for EHE is radical local excision and lymph node dissection[1]. Definitive diagnosis depends on the histopathologic and immunohistochemical features of the tumor[11].

EHE usually grows around or within the affected vessel and tumor cells radiates out[12]. It has poor expression of marker Ki67 and weak positivity for VEGF which indicates less aggressive nature of the tumor[1]. Due to the extensive vascularity and mitotic activity, positive staining factor VIII differentiate it from malignant tumors[1]. Also immunohistochemically these tumors are positive for cytokeratin (specially CK18), and vascular marker including CK31[13].

After confirming the tumor by immunohistochemical features regular follow-up is advised due to the possibility of recurrence.

Conclusion

Even though the tumor had already invaded the surrounding structures, Endoscopy and Neuronavigator assisted excision of the lesion brought a good outcome. Radical local excision is the key important step in the management of epithelioid hemangioendothelioma.

References

1. *Weiss SW, Enzinger FM. Epithelioid hemangioendothelioma: a vascular tumor often mistaken for a carcinoma. Cancer. 1982;50:970–81.*
2. *Patnayak R, Jena A, Reddy MK, Chowhan AK, Rao LC, Rukhamangadha N. Epithelioid hemangioendothelioma of nasal cavity. J Lab Physicians. 2010 Jul;2(2):111-3. doi: 10.4103/0974-2727.72214. PMID: 21346909; PMCID: PMC3040087.*
3. *Liu YI, Brown SS, Elihu A, Bonham CA, Concepcion W, Longacre TA, et al. Hepatic epithelioid hemangioendothelioma. Dig Dis Sci 2011. Feb;56(2):303-306 10.1007/s10620-010-1470-4*
4. *Antonescu C. Malignant vascular tumors – An update. Mod Pathol 2014;27 Suppl 1:S30-8.*
5. *Enzinger FM, Weiss SW. Hemangioendothelioma: vascular tumors of intermediate malignancy. In: Soft Tissue Tumors, 4th edition. St Louis: CV Mosby, 1995:891–900.*
6. *Ko JS, Billings SD. Diagnostically challenging epithelioid vascular tumors. Surg Pathol Clin 2015;8:331-51.*
7. *Polk P, Webb JM. Isolated cutaneous epithelioid hemangioendothelioma. J Am Acad Dermatol 1997;36:1026-8.*
8. *Kato N, Tamura A, Okushiba M. Multiple cutaneous epithelioid hemangioendothelioma: A case with spindle cells. J Dermatol 1998;25:453-9.*
9. *Kanik AB, Hall JD, Bhawan J. Eruptive epithelioid hemangioendothelioma with spindle cells. Am J Dermatopathol 1995;17:612-7.*
10. *Park SY, Lee JK, Jo S, Huh CH, Cho KH, Na JI. Cutaneous epithelioid hemangioendothelioma presented as an ulcerated areolar mass. J Dermatol 2014;41:112-3.*
11. *Tseng C-C, Tsay S-H, Tsai T-L, Shu C-H. Epithelioid hemangioendothelioma of the nasal cavity. Journal of the Chinese Medical Association. 2005;68(1):45–8.*
12. *Alramthan A, Abdulkader A, Alenezi M. Isolated cutaneous epithelioid hemangioendothelioma of the nose: Case report and comprehensive literature review. Journal of Dermatology and Dermatologic Surgery. 2019;23(2):102.*
13. *Chaouki A, Mkhatri A, Ballage A, Zouhair N, Mahtar M. Extensive epithelioid hemangioendothelioma of the maxillary sinus: A case report. International Journal of Surgery Case Reports. 2019;58:70–3.*