

Case Report

Nasal cavity epithelioid hemangioendothelioma invading the anterior skull base

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Abstract

Background: Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor that frequently occurs in soft tissues. Patients suffer from local recurrence and remote metastasis because of its malignant potential. Here, we present a rare case of EHE that originated from nasal cavity and invaded intracranially through the anterior skull base.

Case Description: This is a 27-year-old woman who presented a local physician with intermittent epistaxis and a facial pain around her nose. Preoperative studies demonstrated that the tumor invaded into anterior skull base and the dura matter. Therefore, we performed combined skull base and transnasal surgery, which achieved complete resection of the tumor. Postoperative course of the patient was uneventful. No recurrence or distant metastasis was observed in the patient for 2 years following the radical resection.

Conclusions: To date, four cases of EHE in the nasal cavity were reported. This is the first case in which EHE demonstrated invasive potentials with intracranial extension. Radical surgical resection plays an important role for better management of invasive paranasal EHE.

Key Words: Anterior skull base, epithelioid hemangioendothelioma, nasal cavity, vascular tumor

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INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is a relatively rare vascular tumor, which was first described by Weiss and Enzinger in 1982.^[1] This unusual vascular tumor is characterized by the proliferation of endothelial cells in “epithelioid” appearance.^[5,11] The tumor possesses histological and clinical features intermediate between those of a benign hemangioma and conventional angiosarcoma.^[11] In the fourth edition of the WHO classification of tumors of the central nervous system, published in 2007, EHE is regarded as a borderline intracranial tumor (ICD-O 9133/1).^[5]

EHE frequently occurs in the soft tissue, liver, breast, lung, and bone but rarely in head and neck.^[2,9,11] Reports regarding EHE originated in the nasal cavity

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are scarce.^[3,6,8,10] Importantly, the reported lesions were all localized inside the nasal cavity. Here, we report a first case of paranasal EHE invading through the orbit and the anterior skull base to the dura matter. Although the tumor showed aggressive clinical characteristics, radical resection using combined anterior skull base and endonasal surgical interventions successfully managed the lesion and provided a favorable outcome.

CASE REPORT

History and presentation

A 27-year-old woman presented to a local physician with an intermittent epistaxis and a facial pain around her nose. Her medical history was unremarkable. Computed tomography (CT) scan demonstrated a heterogeneously enhanced mass lesion centered in the left nasal cavity [Figure 1a].

She was referred to our institute for surgical treatment of a nasal cavity tumor. On admission, she suffered from anosmia and diplopia. CT scan, performed 3 months after the first scan, demonstrated that the enlarged mass lesion breaking through the medial wall of the left orbit and pushed the eye laterally [Figure 1b]. We considered this might be the reason for her diplopia. For tissue diagnosis, the patient underwent endoscopic exploration from the nasal cavity. Because of heavy nasal bleeding, we combined midfacial degloving approach. Although we could control the bleeding, we could not resect the complete tumor.

Pathological study of surgical specimen

Hematoxylin and eosin-stained sections showed neoplastic cells forming small intracellular lumina that included red cells. The endothelium covering blood vessels was hyperplastic. No mitosis and necrosis were observed [Figure 2a]. The Ki-67 labeling index was 28%. Immunohistochemical analysis was positive for factor VIII-related antigen, vimentin, CD34, and CD31 [Figure 2b]. EHE was diagnosed on the basis of the aforementioned findings.

Surgical intervention

Magnetic resonance images indicated that the tumor had invaded the anterior skull base dura matter [Figure 3]. Digital subtraction angiography indicated bilateral ethmoidal arteries as major feeding arteries to the tumor. Because of the aggressive nature of EHE, the cancer board of Tohoku University Hospital agreed radical surgery of the tumor utilizing anterior skull base dissection and endonasal endoscopic exploration.

Before the operation, spinal drainage was introduced to reduce brain retraction. Neurosurgeons conducted bilateral frontal craniotomy and excised the anterior skull base dura where the tumor had invaded. The left anterior

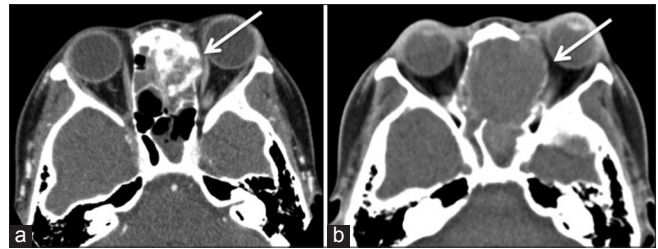


Figure 1: Computed tomography scans at presentation to a local physician (a) and on referral to our institution (b). (a) A heterogeneously enhanced mass was detected in the nasal cavity (arrow). (b) Computed tomography scan without contrast demonstrated rapid progression of the tumor. The tumor invaded into the left orbit (arrow)

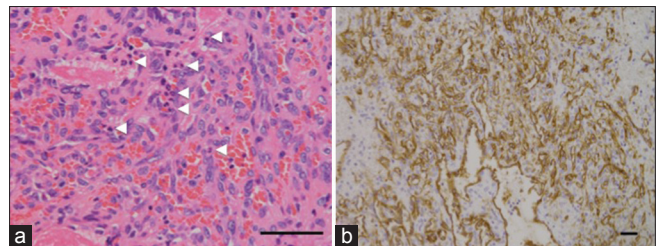


Figure 2: Histological examination of the tumor cells. (a) Neoplastic cells occasionally arranging vascular channels. Mitotic figures are observed (arrowheads). (b) Positive immunohistochemical reaction of the tumor cells for CD31 was observed. Scale bars = 100 μ m

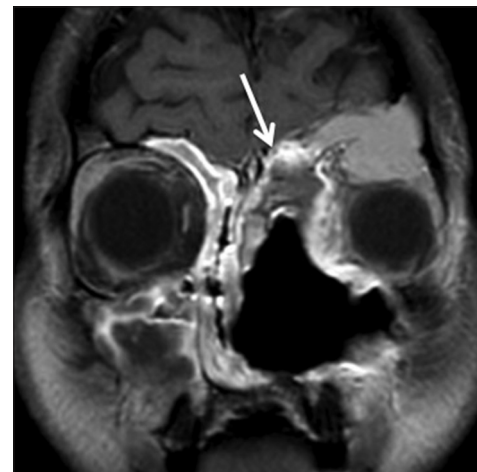


Figure 3: T1-weighted coronal magnetic resonance image with gadolinium enhancement before radical resection of the tumor. Note the tumor originating from the nasal cavity invading the anterior skull base (arrow)

and posterior ethmoidal arteries were coagulated and cut. Following skull base dissections, otolaryngologists achieved complete removal of the tumor through the canine fossa using an endoscope. We confirmed that the tumor was adhesive but had not invaded into the orbit. Intraorbital component was left intact as we had planned preoperatively. Following resection of the tumor and the dura matter, skull base reconstruction was performed using the pericranial flap, fascia lata, and titanium mesh plate.

Table 1: Clinical features of previously published epithelioid hemangioendotheliomas of the nasal cavity

Reference	Age/sex	Degree of resection	Intracranial invasion	Follow-up (months)	Recurrence
Di Girolamo <i>et al.</i> , 2003 ^[3]	23/male	Total	No	12	No
Tseng <i>et al.</i> , 2005 ^[10]	25/male	Total	No	N/A	N/A
Naqvi <i>et al.</i> , 2008 ^[7]	4/male	Subtotal	No	36	Yes*
Patnayak <i>et al.</i> , 2010 ^[9]	40/male	Total	No	9	No
Present case	27/female	Total	Yes	26	No

*Re-operation was performed. There was no recurrence after complete removal of the tumor in the second operation. N/A: Not available

Postoperative course

The patient recovered uneventfully from the radical resection of the tumor. Her diplopia improved. She has been free from local recurrence or distant metastasis for 26 months after the surgery.

DISCUSSION

EHE is an uncommon vascular tumor of the soft tissues characterized by the proliferation of endothelial cells with an epithelioid morphology.^[5,11] Although anatomic locations of EHE were diverse according to previous literatures,^[2,9,11] only four cases of EHE in the nasal cavity have been reported.^[3,6,8,10] Clinical characteristics of the previous and present cases of nasal cavity EHE are summarized in Table 1. All cases were treated surgically. Importantly, no recurrence was observed following complete removal of the tumors. In a case after the subtotal resection, the tumor recurred.^[6] However, the second surgery completely removed the tumor, and the patient had a favorable outcome.

Based on the reported cases, it is difficult to establish therapeutic protocols for paranasal EHE. Because the data are limited, the role of radiotherapy and chemotherapy cannot be completely evaluated. However, as suggested in previous literatures,^[4,7] the most prominent prognostic factors for paranasal EHE could be the degrees of surgical resection. Even when the paranasal EHE invaded the skull base as demonstrated in the presented case, a radical resection was feasible by combining the skull base and transnasal approaches. Parajón and Vaquero^[7] reviewed 34 cases of intracranial EHE and concluded that if complete surgical resection was achieved, no adjuvant radiotherapy was necessary.

A recent study of 49 soft tissue EHEs indicated the higher mitotic activity (>3/50 HPF) as one of the poor prognostic factors.^[2] Our case had a high mitotic activity [Figure 2a] as well as high Ki-67 labeling index suggesting a poor clinical course. However, the patient was free from recurrence or metastasis for 2 years after the surgery. Furthermore, no mortality was recorded in the paranasal EHE in the available follow-up periods [Table 1]. As Bollinger *et al.* reported the prognosis of EHE might vary among locations.^[1] In cases of intracranial EHE, the mortality rate was reported to be 6%.^[7] While the

mortality rate of EHE in the lung was as high as 65% in a 4-year period.^[1] Further study and observations are necessary to disclose clinical characteristics of paranasal EHE.

CONCLUSION

In this case, paranasal EHE invaded the anterior skull base and dura matter. Although the tumor was histologically aggressive, combined anterior skull base and transnasal approaches achieved total removal of the tumor. Radical surgical resection was important to provide a favorable outcome in EHE.

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Conflicts of interest

There are no conflicts of interest.

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