SSR Institute of International Journal of Life Sciences
ISSN (O): 2581-8740 | ISSN (P): 2581-8732
Anjankar *et al*., 2024
DOI: 10.21276/SSR-IIJLS.2024.10.1.13

Case Report

open access

Rapidly Progressive Severe Proptosis in Esthesioneuroblastoma–A Case Report

Shailendra Anjankar[1]*, Charandeep Singh Gandhoke[1], Binoy Kumar Singh[1], Abbas Moaiyadi[2], Mayank Tripathi[2], Pavan Lanjewar[2]

[1]Associate Professor, Department of Neurosurgery, AIIMS Raipur, Chhattisgarh, India
[2]Senior Resident, Department of Neurosurgery, AIIMS Raipur, Chhattisgarh, India

***Address for Correspondence:** Dr. Shailendra Anjankar, Associate Professor, Department of Neurosurgery, AIIMS Raipur, Chhattisgarh, India
E-mail: anjankar.gmc@gmail.com

Received: 06 Aug 2023/ Revised: 14 Oct 2023/ Accepted: 14 Dec 2023

ABSTRACT

Background: Esthesioneuroblastoma (ENB), a rare malignancy originating from olfactory mucosa cells, was first identified by Berger. Clinical manifestations include nasal obstruction, proptosis, and cranial nerve palsies. Despite a bimodal incidence curve, delayed diagnosis is expected due to vague symptoms, emphasizing the need for a comprehensive understanding of its varied presentations.

Methods: We have presented an illustrative case of an 18-year-old male with ENB, highlighting sudden-onset severe symptoms. Thorough clinical examination and imaging studies (MRI, CT) informed a surgical approach, revealing an infiltrative solid mass with substantial bony destruction. Bifrontal craniotomy and meticulous tumor excision were performed, offering insights into the tumor's aggressive features.

Results: Imaging showcased an invasive mass with significant bony destruction. Surgical intervention achieved near-total removal, enabling detailed histopathological examination. Classified as ENB (Hayms grade IV), the tumor exhibited aggressive vascularization and infiltration of surrounding tissues.

Conclusion: Given its diverse and severe symptoms, this case emphasizes the necessity of a multidisciplinary approach for ENB. Timely diagnosis and intervention are crucial, highlighting the significance of combining clinical evaluation, advanced imaging, and surgical expertise. Understanding ENB's clinical behavior and optimizing treatment strategies are paramount for improving patient outcomes.

Key-words: Blindness, Esthesioneuroblastoma, Malignant tumor, Olfactory neuroblastoma, Proptosis

INTRODUCTION

Berger was the initial discoverer of esthesioneuroblastoma, an uncommon malignant tumor originating from bipolar sensory receptor cells in the olfactory mucosa. Stemming from their neural crest progenitor status, these cells contribute to the development of various elements of the olfactory sense. [1,2]

How to cite this article
Anjankar S, Singh C, Singh BK, Moaiyadi A, Tripathi M, et al. Rapidly Progressive Severe Proptosis in Esthesioneuroblastoma–A Case Report. SSR Inst Int J Life Sci., 2024; 10(1): 3537-3541.

Access this article online
https://iijls.com/

The most common initial indication is unilateral nasal obstruction, often accompanied by epistaxis. As the tumor locally extends, additional clinical signs may include proptosis, neck edema, cranial nerve palsies, and symptoms suggestive of metastasis. [3,4] Illustrated here is the case of a young man who, within days of onset, experienced malignant proptosis leading to vision loss.

ENB constitutes up to 5% of malignant nasal cavity tumors, with Berger et al. initially describing the condition in 1924. The incidence curve for ENB exhibits a bimodal distribution, demonstrating an approximately equal distribution among genders. The initial peak is observed in the second decade of life, while the second peak occurs in the sixth decade [5-7]. Patients frequently report nonspecific symptoms like headaches, epistaxis, nasal blockage, and pain.

DOI: 10.21276/SSR-IIJLS.2024.10.1.13

Due to the ambiguous nature of the initial symptoms, diagnosis is often delayed, and patients may have a protracted medical history before receiving a definitive diagnosis [8,9]. Proptosis, epiphora, and decreased visual acuity are commonly associated with ocular malignancies [10,11]. In this context, we present a case of ENB in a sixty-year-old male patient, evident through proptosis and nasal obstruction as revealed on computed tomography scans.

CASE PRESENTATION

An 18-year-old male presented with a gradual onset, progressive holocranial headache persisting for 2 months. Notably, there has been a protrusion of the right eyeball for the past 15 days and a similar occurrence in the left eye for the last eight days, both sudden in onset and rapidly progressive. The patient reported a history of diminishing vision in both eyes for the past 8 days, leading to complete blindness over the last 5 days. Additionally, there was a history of nasal bleeding occurring intermittently for the past 3 days, coupled with difficulty in opening the mouth. The patient has no co-morbidities and no significant past medical history.

Upon examination, bilateral severe proptosis of the eyes was observed, with the right eye displaying total corneal melting and the left eye exhibiting exposure keratopathy. Furthermore, there was bilateral mild lid edema and severe conjunctival chemosis (Fig. 1).

Fig. 1: Clinical photograph of patient showing severe proptosis

Bilateral compression of the iris, lens, and globe was observed due to pressure posterior to the world. The patient experienced complete vision loss with absent extraocular eye movement bilaterally. Loss of smell from both nostrils and decreased taste sensation was noted, with no other cranial nerve deficits. Swelling over the bilateral parotid region was observed. Limb power and sensation were average, and vital signs were stable. MRI revealed a poorly defined, infiltrative solid cellular mass affecting bilateral ethmoidal cells, with an extra-axial mass measuring 4 x 3.7 x 1.5 centimetres. The mass invaded the medial orbital wall and infiltrated the medial extra coanal spaces bilaterally, exhibiting homogeneous moderate enhancement with gadolinium (Fig. 2).

A brain CT scan revealed bone degradation in the lamina papyreacea, bony nasal septum, anterior sphenoidal sinus wall, bilateral ethmoidal lamella, and anterior cranial fossa floor. Once the required consents were secured, his procedure was arranged, and the prognosis was shared. Using a cranial approach, a bifrontal craniotomy and tumor excision were performed. The tumor was vascular, firm, and gray in appearance. It adhered to the dura and nasal mucosa like glue.

Additionally, it penetrated and destroyed the bone architecture of the front skull base. A malformation in the bottom of the skull would allow the cranial, nasal, and sinus components to be removed almost completely. Using autologous adipose tissue, split calvarial graft from the frontal bone, and pericranial graft, the base of the skull was rebuilt. There was also bilateral tarsoraphy. The patient did not show any new neurological abnormalities and was not receiving oxygen. The tumor's pathology revealed grade IV Hayms embryo-neuroblastoma (Fig. 3).

DOI: 10.21276/SSR-IIJLS.2024.10.1.13

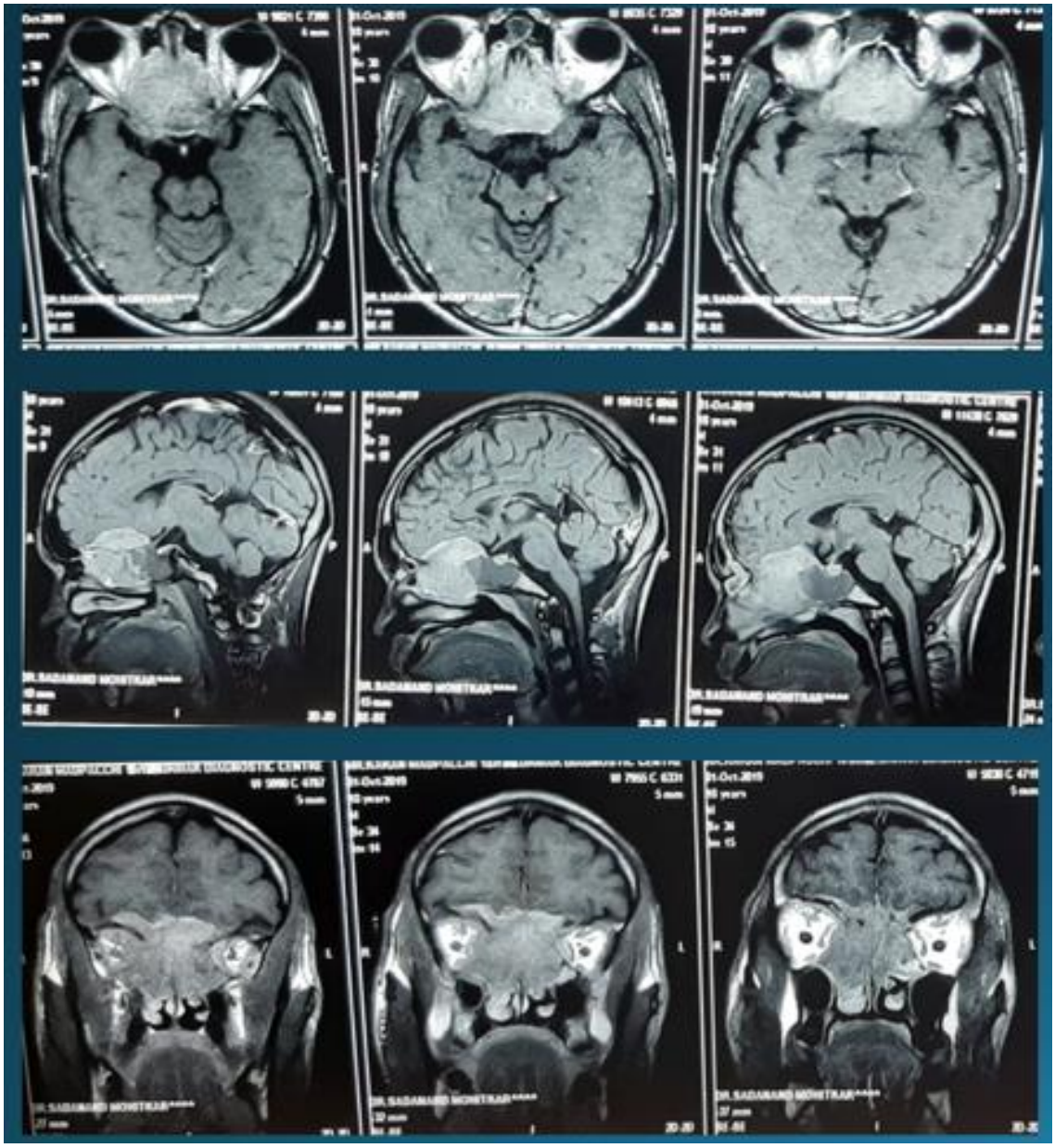

Fig. 2: MRI Brain of patient showing homogeneously enhancing, locally aggressive, ill-defined infiltrative solid cellular mass of size 4x3.7x1.5 cm invading the medial orbital wall and infiltrating bilateral medial extra choanal spaces

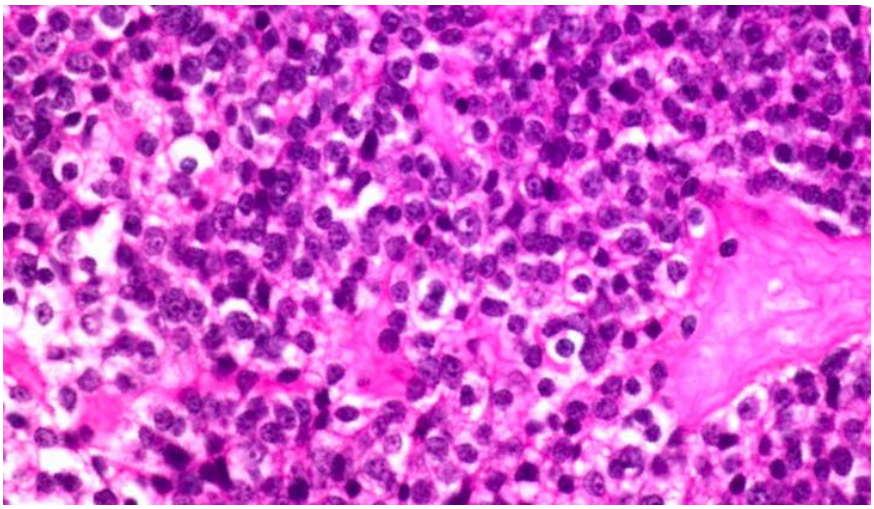

Fig. 3: Histopathology of tumor suggestive of esthesioneurobastoma

DOI: 10.21276/SSR-IIJLS.2024.10.1.13

DISCUSSION

Only 1225 ENB occurrences have been reported in published literature as of this writing. [3-6] Bimodal and Inci are dividing. The tumor ENB has almost equal maxima in the second and sixth decades. Locally, the tumor is aggressive. Despite the aggressive presentation of teenage ENB, multimodal modality therapy produces favourable clinical outcomes. [12,13] Nair *et al.* [14] have documented two cases of ENB; the patient in one of those cases was younger and the clinical symptoms manifested and progressed similarly to our patient. Ansari *et al.* [15] reported a case of ENB presenting with proptosis over an indolent 1.5 years; the case under review, however, is a young newborn whose symptoms develop rapidly and progress to proptosis and blindness in a few days. Mahdi Shahriari et al. reported a case akin to this one involving a blind 21-month-old girl with a limited medical history and bilateral proptosis. [16] This illustrates how the ENB presentation varies according to age group. Since younger patients usually have higher initial Kadish grades when they initially show up, diagnosis durations are generally shorter. According to Kadish's study, ENB was divided into three phases: Group C demonstrated that the tumor had progressed to other regions, such as the orbit, base of the skull, and cerebral compartment. Group A showed that the tumor was exclusively detected in the nasal cavity; Group B demonstrated that it was seen in both the nasal cavity and the paranasal sinuses. [17]

Radiation and surgery demonstrated considerably greater 5-year overall and progress-free survival than alternative treatment modalities. [4-7] Radiation therapy is the most often used treatment for ENB since it has the highest overall survival rate. The most common type of treatment is surgery. [3-5] Cranial therapy may be required if a higher-grade tumor is causing erosion at the base of the skull. In this instance, the endonasal method might be used to remove the intranasal component of the tumor during the second surgical stage. [18-2] Since ENB patients are rare, current therapy recommendations are unclear. According to ENB, the degree of resection, positive lymph nodes, Hyams grade, Kadish stage, and postoperative radiation therapy (RT) with a minimum dose of 54 Gy are the most important prognostic markers influencing the outcome. [17]

CONCLUSIONS

Younger patients with Esthesioneuroblastoma often present with rapidly progressing disease and may manifest late-stage symptoms, including complete blindness. Recognizing ENB as a potential cause of proptosis in the young is crucial. The established mainstay of treatment involves surgery followed by radiotherapy.

However, for future advancements, a larger sample size is imperative. Outcomes should be individually evaluated based on factors such as Hyams grade, positive lymph nodes, Kadish stage, extent of resection, and postoperative radiotherapy. This approach will facilitate nuanced prognostication and enhance the treatment paradigm for this highly malignant tumor.

CONTRIBUTION OF AUTHORS

Research concept- Dr. Shailendra Anjankar
Research design- Dr. Shailendra Anjankar
Supervision- Dr. Shailendra Anjankar
Materials- Dr. Shailendra Anjankar
Data collection- Dr. Shailendra Anjankar
Data analysis and Interpretation- Dr. Shailendra Anjankar, Dr. Charandeep Singh Gandhoke, Dr. Binoy Kumar Singh, Dr Abbas Moaiyadi, Dr. Mayank Tripathi, Dr. Pavan Lanjewar
Literature search- Dr. Shailendra Anjankar, Dr. Charandeep Singh Gandhoke, Dr. Binoy Kumar Singh, Dr Abbas Moaiyadi, Dr. Mayank Tripathi, Dr. Pavan Lanjewar
Writing article- Dr. Shailendra Anjankar, Dr. Charandeep Singh Gandhoke, Dr. Binoy Kumar Singh, Dr Abbas Moaiyadi, Dr. Mayank Tripathi, Dr. Pavan Lanjewar
Critical review-Dr. Shailendra Anjankar, Dr. Charandeep Singh Gandhoke, Dr. Binoy Kumar Singh, Dr Abbas Moaiyadi, Dr. Mayank Tripathi, Dr. Pavan Lanjewar
Article editing-Dr. Shailendra Anjankar, Dr. Charandeep Singh Gandhoke, Dr. Binoy Kumar Singh, Dr Abbas Moaiyadi, Dr. Mayank Tripathi, Dr. Pavan Lanjewar
Final approval- Dr. Shailendra Anjankar

REFERENCES

[1] Dulguerov P, Allal AS, Calcaterra TC. Esthesioneuroblastoma: a meta-analysis and review. The lancet oncology, 2001; 2(11): 683-90.

DOI: 10.21276/SSR-IIJLS.2024.10.1.13

[2] Kumar R. Esthesioneuroblastoma: Multimodal management and review of literature. WJCC, 2015; 3(9): 774.

[3] Yang SH, Yen CC, et al. Long-term remission of advanced esthesioneuroblastoma after chemoradio therapy. J Cancer Res Pract., 2012; 28(4): 177–82.

[4] Loy AH, Reibel JF, Read PW, et al. Esthesioneuroblastoma: continued follow-up of a single institution's experience. Arch Otolaryngol Head Neck Surg., 2006; 132: 134–38.

[5] Carey RM, Godovchik J, et al. Patient, disease, and treatment factors associated with overall survival in esthesioneuroblastoma. Int Forum Allergy Rhinol., 2017; pp. 1186-94.

[6] Prakash SP, Park MC, Huang D, Deville C, Cortez S, et al. Esthesioneuroblastoma (Olfactory Neuroblastoma) with Hemorrhage: an unusual presentation. Skull Base, 2006; 16: 169–74.

[7] Kiyota N, Tahara M, Fujii S, et al. Nonplatinum-based chemotherapy with irinotecan plus docetaxel for advanced or metastatic olfactory neuroblastoma. Cancer, 2008; 112: 885–91.

[8] Chan LP, Wang LF, Tai CF, Wu CC, Kuo WR. Huge sphenoid sinus olfactory neuroblastoma: a case report. Kaohsiung J Med Sci., 2009; 25: 87–92.

[9] Sharma S, Sharma MC, Johnson MH, Lou M, Thakara A, et al. Esthesioneuroblastoma- a clinicopathologic study and role of DNA topoisomerase alpha. Pathol Oncol Res., 2007; 13: 123–29.

[10] Li-wei Z, Ming-shan Z, Ji Q, et al. Management of intracranial invasive olfactory neuroblastoma. Chin Med J., 2007; 120: 224–27.

[11] Thompson LDR. Olfactory neuroblastoma. Ear Nose Throat J., 2006; 85: 569–70.

[12] Dulguerov P, Allal AS, Calcaterra TC. Esthesioneuroblastoma: a meta-analysis and review. Lancet Oncol., 2001; 2: 683–90.

[13] Kiyota N, Tahara M, Fujii S, et al. Nonplatinum-based chemotherapy with irinotecan plus docetaxel for advanced or metastatic olfactory neuroblastoma: A retrospective analysis of 12 cases. Cancer, 2008; 112(4): 885-91.

[14] Nair R, Nayal B, Kumar V, Jaiprakash P. Esthesioneuroblastomas: Reservations and recommendations. J Clin Diagn Res., 2016; 10.

[15] Ansari S, Ahmad K, Dhungel K, Gupta MK, Amanullah MF. Esthesioneuroblastoma: one of the causes of proptosis. Head Face Med., 2013; 9(1): 19. doi: 10.1186/1746-160X-9-19.

[16] Shahriari M, Shakibazad N, Moradi M. Esthesioneuroblastoma Presenting with Bilateral Proptosis and Blindness in a Child: A Case Report. J Clin Diagn Res., 2017; 11(8): XD01.

[17] Kairemo KJA, Jekunen AP, Kestila MS, Ramsay HA. Imaging of olfactory neuroblastoma—an analysis of 17 cases. Auris Nasus Larynx, 1998; 25 (2): 173-79. doi: 10.1016/S0385-8146(98)00003-0.

[18] Morita A, Ebersold MJ, Olsen KD, et al. Esthesioneuroblastoma: Prognosis and management. Neuro., 1993; 32: 706–14.

[19] Yuan Y, Ye J, Qiu H, Niu S, Wen B, et al. Exploration of the optimal treatment regimes for Esthesioneuroblastoma: a single center experience in China. J Cancer, 2018; 9(1): 174.

[20] Porter AB, Bernold DM, Giannini C, et al. Retrospective review of adjuvant chemotherapy for esthesioneuroblastoma. J Neurooncol., 2008; 90: 201–04.

[21] Karp EE, Van Gompel JJ, Choby G. Esthesioneuroblastoma (Olfactory Neuroblastoma): Overview and Extent of Surgical Approach and Skull Base Resection. J Neurol Surg Rep., 2022; 83: e80–e82.