CASE REPORT

Olfactory neuroblastoma: A rare sinonasal malignancy

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Abstract

Olfactory neuroblastoma is a rare malignant tumour arising from the olfactory nerve and extending into the nasal cavity. In this case report, the case of a 42-year-old male is presented. The patient had a two-month history of progressive nasal blockage and episodes of epistaxis. No complaint of anosmia or facial pain was reported. All the necessary examinations were performed. Upon investigation, the CT scan and MRI showed a polypoid mass involving the right maxillary sinus, eroding the medial wall and expanding into the osteo-meatal complex. The diagnosis of olfactory neuroblastoma was confirmed through histopathological examination and further validated by immunohistochemistry as it was positive for synaptophysin, chromogranin, gamma enolase, and neurofilament. On staging, the tumour was Kadish B. The mass was excised by lateral rhinotomy. The patient was kept on radiotherapy and was free from recurrence upon follow-up 10 months later. It was concluded that based on the analysis of findings related to olfactory neuroblastomas, clinicians should contemplate the possibility of an ONB when radiographic images depict a dumbbell-shaped mass within the nasal cavity, accompanied by peritumoural cysts. Using a multimodal treatment approach is advisable.

Keywords: Olfactory neuroblastoma, Esthesioneuroblastoma, Pseudo-rosettes, Lateral rhinotomy, Nasal cavity, synaptophysin positive.

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Introduction

Olfactory neuroblastoma (ONB), also known as Aesthesioneuroblastoma, is a malignant tumour of neural crest origin arising from the olfactory neuroepithelium of the roof of the nasal cavity and paranasal sinuses, representing 3% of sinonasal malignancies. According to the National Cancer Institute (NCI), ONB is seen only in one out of 2.5 million people yearly.[1] It was first described by Berger and Richard in 1924. This condition exhibits a bimodal age distribution, with peak prevalence occurring between 10-20 and 50-60 years. These lesions may go unnoticed because the presenting symptoms mimic those of benign tumours of the nasal cavity. Clinical features of ONB involve unilateral nasal obstruction, epistaxis, anosmia, facial or orbital pain, proptosis, epiphora, diplopia, and blurred vision.

Lymph node metastases in the neck can occur in 10-15% of the cases. These are often an incidental finding during septoplasty or polypectomy.[2] As olfactory neuroblastomas are so uncommon, few evidence-based strategies for optimum management have been reported, although diagnostic and treatment modalities have improved over the past two decades. It is a vascular tumour, and biopsy should not be attempted immediately unless imaging studies have been done. Pathology involves the formation of rosettes and pseudo-rosettes. CT scans and MRIs are necessary to find the extent of the diseases so that grading and staging can be done. There are several treatment options for olfactory neuroblastoma, ranging from minimally invasive surgical approaches[3] to more extensive craniofacial resection procedures, often in combination with radiotherapy or chemotherapy.

The case presented is of a 42 year old male complaining of epistaxis and showing a sessile, polypoid mass in the right nasal cavity, diagnosed as olfactory neuroblastoma grade 1, and was successfully treated by excision.

Case Report

A 42-year-old male presented to the otolaryngology department of Ghurki Trust and Teaching Hospital, Lahore, on August 6, 2022, with complaints of episodes of epistaxis off and on for two months and nasal blockage for the past one year. There was no associated complaint of anosmia or facial pain. A nasal examination was performed with the help of a rigid endoscope, which revealed a sessile, polypoid mass in the right nasal cavity. The mass was fixed lobulated with a non-friable texture. His vision in both eyes was unimpaired, and he had full, unrestricted eye movements.

The patient's CT scan and MRI showed the presence of a polypoid mass involving the right maxillary sinus, eroding the medial wall and expanding into the osteo-meatal complex (Figure 1). Partial extension was also seen in the

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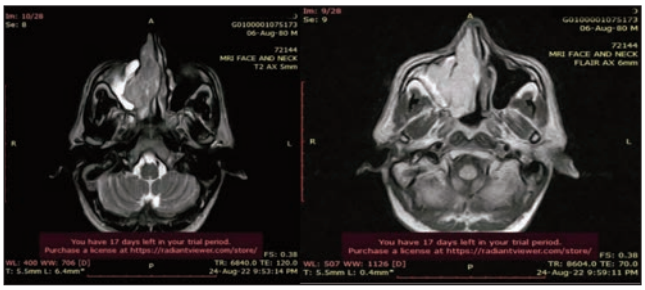

Figure-1: The contrast enhanced MRI (coronal view) showing a soft tissue mass in the right nasal cavity.

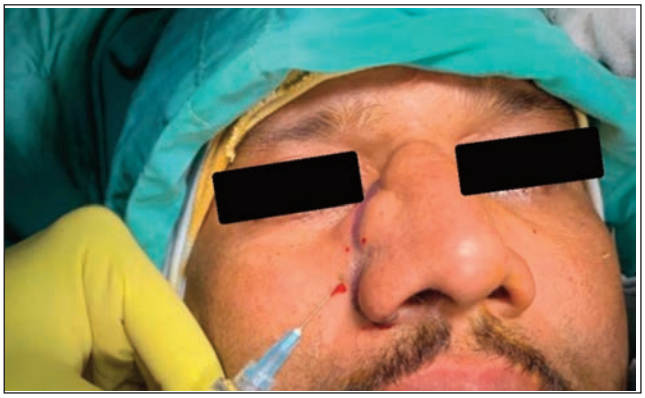

Figure-2: This shows the pre-op condition of the patient. The tumour mass can be appreciated as a bulge on the lateral side of nose.

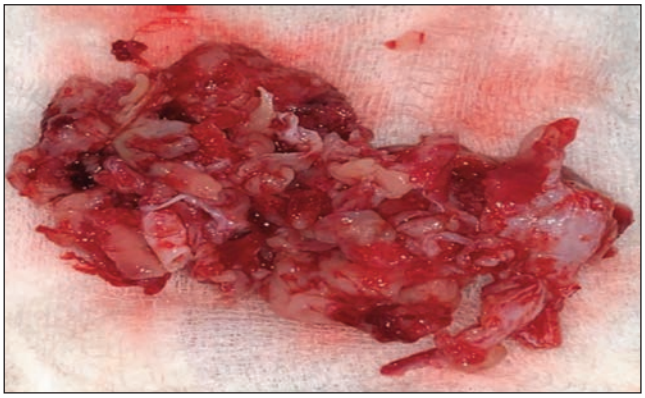

Figure-3: The excised mass after lateral rhinotomy.

left nasal cavity. The disease also extended into the right sphenopalatine foramen. The cribriform plate and lamina papyracea were intact, indicating that there was no intracranial or intra-orbital involvement. Also, there was no involvement of cervical lymph nodes.

Involvement of the posterior aspect of right ethmoid air cells was seen, indicating a high probability of malignancy. The tumour was graded to be Kadish B, according to the Kaddish and Dulguerov classification system.[4] Immunohistochemistry report of the biopsy of the mass confirmed the diagnosis of olfactory neuroblastoma grade 1 as it was positive for synaptophysin, chromogranin, gamma enolase, and neurofilament.

After the confirmation of the diagnosis, treatment options were explained to the patient in detail and informed consent was taken prior to the surgery. The opted treatment was complete surgical excision of the mass followed by radiotherapy. Figure 2 shows the pre-op condition of the patient. A lateral rhinotomy incision was made just above the nasomaxillary groove extending from the inner canthus to the alar groove. The complete flap was raised. Pale growth was seen in the nasal cavity involving the maxillary and ethmoid sinus. It was excised, and the remaining sinuses were evaluated and cleared for any residual tumours in the cavity. Figure 3 shows the excised tumour. The patient was kept under observation for a week. He was then referred to the oncology department for radiotherapy. The first post-op follow-up was scheduled after six weeks, then every three months for two years, and every six months in the coming years. Upon recent follow-up, no recurrence was observed.

Discussion

Olfactory neuroblastomas originate from the olfactory neuroepithelium, which stretches from the upper part of the nasal cavity to the region around the superior turbinate and a segment of the nasal septum. From this point, they can extend into the cribriform plate of the ethmoid sinus. While most cases documented in the literature pertain to adults, a case of this condition occurring in a child as young as two years old has also been reported.[5]

Patients with olfactory neuroblastoma usually present with the symptoms of nasal blockage and epistaxis so it needs to be differentiated from lymphoma, melanoma, plasmacytoma, rhabdomyosarcoma, undifferentiated carcinoma, and neuroendocrine carcinoma.

Nasal endoscopy shows the presence of exophytic, polypoid, and sessile mass arising from the roof of the nasal cavity. A high-resolution CT scan will confirm the invasion of local bony structures, particularly the cribriform plate, anterior maxillary fossa, and retro-maxillary space. MRI will confirm the extent of soft tissue invasion, particularly the anterior cranial fossa and the orbit. The true nature of the tumour is confirmed upon biopsy. The hallmark of the tumour is the formation of tumour cells into rosettes, pseudo-rosettes, sheets, or clusters separated by a fibrovascular stroma. If low grade, the formation of Homer Wright pseudo-rosettes is seen, and if it is a high-grade tumour, then nuclear pleomorphism but no rosette formations are observed. It often requires special staining for confirmation of the differential diagnosis.

Immunohistochemical studies show that olfactory

neuroblastoma is positive for neuroendocrine markers such as chromogranin, synaptophysin, gamma enolase, and protein gene product 9.5; this eventually gives a definitive diagnosis.

A few tumour staging systems have been devised. The most well-accepted of these is the Kaddish and Dulguerov[11] classification system (stages A through C), which is based on the clinical spread of the tumour; stage A tumours are confined to the nasal cavity, stage B lesions involve the paranasal sinuses, and stage C tumours have intra-orbital and intracranial invasion. The tumours in each of these classifications behave differently with respect to progression and metastasis.

Multimodality therapy proves to be the best treatment option unless the patient is at a very early stage and has limited disease. The treatment involves surgery followed by radiotherapy or chemotherapy. The conventional surgical method for ONB involves anterior craniofacial resection, which incorporates a bifrontal craniotomy in conjunction with a trans-facial lateral rhinotomy. Purely endoscopic approaches are not recommended in cases where the anterior table of the frontal sinus, skin, subcutaneous tissue, nasolacrimal sac, carotid artery, or extensive intra-orbital and intracranial invasion is present. Additionally, contraindications for purely endoscopic approaches encompass lateral extension above the orbit and palatine invasion, in such cases, traditional craniofacial approaches are more appropriate. Irrespective of the chosen approach, the primary goal remains to achieve complete resection with negative margins in all cases.[6] Preoperative irradiation doesn't seem to offer any significant advantages. Following the surgery, there are differing recommendations among authors: some propose radiotherapy solely for advanced tumours, while others argue that it should be given to all patients, regardless of the tumour stage.[7] Currently, the gold standard in managing these tumours involves craniofacial resection combined with radiotherapy.[8] Chemotherapy is reserved for cases where surgical resection is not feasible or for recurrent tumours and distant metastases. Opposed to the standard ongoing practice, a case has been reported with advanced disease (Kadish stage C) in which cure was achieved with chemotherapy combined with adjuvant radiotherapy alone.[9]

Olfactory neuroblastomas possess the capability to spread within the surrounding region. In a comprehensive review by Rinaldo and colleagues involving 320 cases, it was observed that the occurrence of both concurrent and subsequent neck metastases exhibited significant variability, spanning from 5% to 100%, with an average rate of 23.4%.[10] As with the majority of head and neck cancers, the presence of cervical lymph node involvement is an unfavourable sign and heightens the likelihood of distant metastasis, primarily to the bones.[12] In the present case, there was no lymph node involvement and this factor played a significant role in deciding the treatment plan.

Even with intensive treatment, recurrence may occur shortly after the initial therapy or several years later. The long-term survival rate is poor. Proper patient follow-up is necessary for a detailed observation of the patient's condition. Cytogenetic studies have unveiled considerable variability in the chromosomal aberrations observed in ONB tumours. To discern advanced potential therapeutic targets for ONB, it is imperative to conduct comprehensive genomic and epigenomic studies involving large-scale cohorts.[13]

Conclusion

The case of an ONB with typical presenting complaints is documented. Based on the analysis of findings related to olfactory neuroblastomas, clinicians should contemplate the possibility of an ONB diagnosis when radiographic images depict a dumbbell-shaped mass within the nasal cavity, accompanied by peritumoral cysts. For primary ONB located in the nasopharynx, either a combination of surgery and radiotherapy or definitive radiotherapy is advisable. A proper follow-up is always necessary to monitor and manage any recurrence promptly.

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Author Contribution:
AAU: Data Acquisition, interpretation, proofreading.
SZ: Compiling of patient's data and initial handling of the case.
AS: Concept, writing and initial composition.
SI: Writing, references and editing