Computed Tomography and Magnetic Resonance Imaging of Preoperative Cases of Esthesioneuroblastoma (Olfactory Neuroblastoma): A Case Series

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ABSTRACT
Esthesioneuroblastoma is an uncommon aggressive tumour arising from the olfactory epithelium of superior nasal cavity, frequently invading the base of cranium, cranial vault and orbit. Contrast enhanced Computed Tomography (CT) and contrast enhanced Magnetic Resonance Imaging (MRI) were done for three patients which showed modified Kadish stage C tumour. Biopsy was done to confirm the diagnosis and patients were subjected to surgery and radiotherapy. The purpose of this case series was to report the radiological features of esthesioneuroblastoma. This is a report on three cases of different age groups who presented tumour with intracranial extension which were histopathologically proven as esthesioneuroblastoma.

INTRODUCTION
Esthesioneuroblastoma is also known as olfactory neuroblastoma (neuroectodermal tumour), these are uncommon malignant tumours originating in the olfactory epithelium in the superior nasal cavity of olfactory recess. Approximately, the incidence of esthesioneuroblastoma is 4 per 10,00,000 cases. A wide range of age groups can present with esthesioneuroblastoma and frequently in a bimodal distribution, occurring most frequently in the second and sixth decades. There is no racial predilection to esthesioneuroblastoma [1]. Commonly, males are seen affected more than females with esthesioneuroblastoma. The male to female ratio is approximately 1.2:1 [2-5].

Most of the patients are locally diagnosed with advanced stages of the disease and hence, requires multimodality treatment in form of surgery, chemotherapy or and radiotherapy.

CASE SERIES

Case 1
A young female in her third decade (27 years) presented with a mass in right nasal cavity, nasal bleeding and anosmia for a year. She underwent a lot of medications, but showed no improvement, recently she felt difficulty in breathing and severe frontal headache which were unresponsive to pain killers. On examination, a huge fleshy polypoidal mass was visible in right nasal cavity with non preservation of normal anatomy of nasal cavity and bleed with touch. Examination of the left nasal cavity, mouth, ears, neck and cranial nerves were within normal limited.

MRI with T1 axial and coronal sections, T2/FLAIR (Fluid Atenuated Inversion Recovery) axial sections and T1 post-gadolinium (contrast) axial and coronal sections were achieved. An isointense expansile mass lesion was noted in right nasal cavity with destruction of ethmoid air cells on T1WI [Table/Fig-1a] causing widening and destruction of right cribriform plate. FLAIR sequences showed significant perilesional oedema (hyperintensities), causing buckling and mass effect over the bilateral basi-frontal neuroparenchyma [Table/Fig-1b]. Postcontrast the lesion shows adjacent peritumoural cyst [Table/Fig-1c] with heterogeneous pattern of enhancement [Table/Fig-1d]. On Histopathological Examination (HPE) tumour cells were arranged in lobular pattern and large islands of small cells [Table/Fig-2].

Case 2
A 60-year-old male came to Ear, Nose and Throat (ENT) clinic and presented with history of headache, anosmia, episodic epistaxis, epiphora, left sided nasal blockage for a year and an episode of involuntary movements of both upper and lower...
On non contrast CT [Table/Fig-4b] there was a hypodense mass lesion with few cystic areas in frontal lobe in midline. On post-contrast (iohexol) CT, a large heterogeneously enhancing mass (Hounsfield unit 55 to 68) in left nasal cavity and sinuses [Table/Fig-5a,b] have been seen extending intracranially in left frontal region crossing midline with adjacent peritumoural cyst ([Table/Fig-5c] arrowhead) with enhancing walls and significant peri-tumoural oedema [Table/Fig-5d,e]. No evidence of intra-lesional calcifications/haemorrhage/ intra-orbital extensions.

CT of para-nasal sinuses and brain was performed with axial, coronal and sagittal reformation which showed an expansile irregular soft tissue density mass lesion (Hounsfield unit 14 to 20) in left nasal cavity involving left maxillary and sphenoid sinuses [Table/Fig-3a] causing obliteration of ostiomeatal complexes and extending superiorly intracranially into basi-frontal region crossing the midline. Destruction and thinning of left ethmoid air cells, left maxillary sinus walls, nasal septum, left inferolateral part of crista galli with widening and erosion of cribriform plate on left side [Table/Fig-3b,c] are extending posteriorly unto nasopharynx [Table/Fig-4a].

On HPE examination, scant vesicular cytoplasm and scattered rosettes along with fibrillary matrix and mitotic activity with no evidence of necrosis was observed [Table/Fig-6] red-arrow.

Case 3
A 35-year-old female came with the complaints of nasal obstruction and increasing swelling over left cheek region and headache for a year. Recently, she developed severe recurrent epistaxis and complete anosmia and severe headache which was not relieved with painkiller. No previous medical treatment or surgery was performed. On examination, a fleshy pinkish mass was visible in left nostril causing completely occupying left nasal cavity and firmed, non tender swelling with ill-defined margins over the left cheek and maxillary region and loss of smell sensation. There was no rise in local temperature in left cheek region. Cold spatula test did not show fogging on left side. Blood profile was within normal limits of complete blood count, liver function test and renal function test.
CT of para-nasal sinuses and brain was performed with axial, coronal and sagittal reformation which showed an ill-defined irregular heterogeneously enhancing mass lesion in left nasal cavity. The maxillary sinus completely obliterates left nasal cavity and extends to anterior cranial fossa through the cribiform plate. The lesion was causing destruction to medial wall of left maxillary sinus, uncinate process, superior, middle, inferior turbinate, nasal septum and superiorly cribiform plate [Table/Fig-7a-c].

The heterogeneous mass lesion shows diffusion restriction on Diffusion Weighted Imaging (DWI) [Table/Fig-8a] with corresponding coronal and sagittal images [Table/Fig-8b,c]. MRI with T1 axial and coronal sections, T2/FLAIR, DWI, Apparent Diffusion Coefficient (ADC), Gradient Echo (GRE) axial sections and T1 post gadolinium (contrast) axial and coronal sections were achieved which showed an irregular heterogeneously enhancing mass lesion in left nasal cavity superiorly extending intracranially to anterior cranial fossa eroding cribiform plate on left side causing obliteration of fronto-ethmoidal recess and retention of secretions in left maxillary, sphenoid and frontal sinuses with downward and lateral displacement of left middle and inferior turbinates [Table/Fig-9,10].

A nasal cotton gauze soaked in a mixture of 5 mL of lidocaine 1% and 1 mL of adrenalin 1:50,000 is inserted in nasal cavity on the same side of lesion at an ear, nose and throat clinic then the biopsy specimen was taken under topical anesthesia. All the above three cases were diagnosed as esthesioneuroblastoma. HPE showed tumour cells arranged in lobular pattern and large islands of small cells, having a hyperchromatic nucleus with inconspicuous nucleolus, [Table/ Fig-11]. Differentials were olfactory neuroepithelioma and sinonasal carcinoma on imaging appears identical to esthesioneuroblastoma; however peritumoural cysts are uncommon in latter.

DISCUSSION
Esthesioneuroblastoma is an unusual (1-5% of all nasal tumours) and destructive malignant neurogenic tumour located in the nasal cavity. Since, its initial description in 1924 by Berger and Luc [6] more than 1,000 cases of esthesioneuroblastoma were reported worldwide [7]. Symptoms are associated with the site of origin and extension/invasion of the tumour. Kadish S et al., 1976 accord tumour extensions as stage A, B, and C [8]. Later, Morita A et al., added stage D tumours as metastases in cervical lymph nodes and distant sites [9]. Biler HF et al., Dulguerov P and Calcaterra T are other two staging methods used [10,11]. The groups A-D are described below-

Group A: Mass lesion limited to the nasal cavity
Group B: Mass lesion limited to the nasal cavity and paranasal sinuses
Group C: Mass lesion extending beyond the nasal cavity and paranasal sinuses such as intracranial/intraorbital extension
Group D: Cervical nodal metastases

All three patients presented with Stage C in present study according to Kadish S et al., [8] with intracranial extension. The age group in present series showed patients ranged between 21 and 65 years old. There is still a lack of consensus over the incidence in both genders. Some reported that esthesioneuroblastoma affect male and female patients equally [12], some thought that it is slightly more common in males [13-16] or otherwise [17]. In general, the clinical symptoms of esthesioneuroblastoma were not specific and related to tumour sites and invasion [18]. The commonest presenting symptoms were unilateral nasal obstruction and anosmia in index study. Due to its vague symptoms and slow growing nature, esthesioneuroblastoma patients often have a long history of progressive symptoms for months, prior to diagnosis [11]. The average duration from symptom onset to management was 1 year in index study and six months (0–18 months) in the USA, reportedly [13]. This study established that MRI and CT manifestation of esthesioneuroblastoma are imprecise, sharing similar signal intensity and density features and
more commoner and anosmia being the commonest symptom. It suggests that intracranial extension and peritumoural cysts are helpful in choosing appropriate initial operative management. Esthesioneuroblastomas presented as expansible intranasal lesions. CT/MRI can help predict patient management [33]. In these aspects, both MR and CT scanning are helpful in providing necessary information as to surgery. Due to the high aggressive behaviour and recurrence rate [28,32] of esthesioneuroblastoma, early diagnosis, accurate metastases were not seen. This report mainly concentrated on these three cases with direct intracranial extension and categorised them into three types based on the depth of invasion: brain parenchyma (Kadish Group C) (3/3), other types being in nasal cavity (Grade A) or para-nasal sinuses (Grade B). The brain parenchyma type had the primary tumour extend across the cribriform plate, growing through the dura and then infiltrate into the anterior frontal lobe. Even though the bulk of the tumour may lie intracranial, it is still attached to the cribriform plate. Compared with the gray matter, the tumour parenchyma appears T2/FLAIR hyperintense and hypointense on T1-weighted images. FLAIR image exhibits the peritumoural oedema more clearly, but gadolinium-enhanced T1-weighted images visualise the tumoural boundary due to significant enhancement of all involved soft tissues. Peri-tumoural cysts (arrowhead in Table/Fig-5c) along the intracranial tumour margin are portrayed clearly, MR and CT images show that the tumour located at high in the nasal cavity extending through the left cribriform plate and invading the ipsilateral frontal lobe. Peri-tumoural cyst (arrows) along the margin of the intracranial tumour is portrayed clearly. Pre-contrast coronal CT image shows a hyperdense endonasal mass with slight high density to the surrounding soft tissue. Post-contrast coronal CT image shows the tumour enhances significantly and heterogeneously. No intra-lesional calcification can be considered pathognomonic for esthesioneuroblastoma. Presence of intracranial cysts in a sino-nasal mass with CNS extension is highly suggestive of esthesioneuroblastoma [31]. The base-of-skull type was revealed in three cases, where bulging dura mater were demonstrated and thinned or destroyed cribriform complex were discovered during surgery. Due to the high aggressive behaviour and recurrence rate [28,32] of esthesioneuroblastoma, early diagnosis, accurate staging and close follow-ups are essential for optimistic patient management [33]. In these aspects, both MR and CT scanning are helpful in providing necessary information as to the tumour location and extension and proximity to the adjacent structures.

**CONCLUSION(S)**

Esthesioneuroblastomas presented as expansible intranasal lesions that unilaterally widens the olfactory recess. CT/MRI can help predict extent and staging of tumour, guide extent for biopsy, potentially helpful in choosing appropriate initial operative management. This analysis of the case series of esthesioneuroblastomas suggests that intracranial extension and peritumoral cysts are more commoner and anosmia being the commonest symptom.

**REFERENCES**


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