Esthesioneuroblastoma - An Unusual Case Report

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ABSTRACT

Introduction: Esthesioneuroblastoma is an extremely rare malignant tumour of the olfactory epithelium. Originally described first by Berger et al in 1924.

Case report: We present a case of 70 year old female who presented with the chief complain of swelling on the nose associated with difficulty in breathing and nasal obstruction since one and half years. Histopathological specimen was received in the Dept. of Histopathology in 10% formalin solution. After routine processing regular staining with Haematoxylin and Eosin followed by confirmation with Immunohistochemistry.

Conclusion: Olfactory neuroblastoma (ONB) is an rare malignant neuroectodermal nasal tumour that constitutes about 2% of all sinonasal tract tumours without any specific age, sex or racial predilection. It is a slow growing locally invasive tumor with low propensity to metastasis. Metastasis occurs mostly to lungs and bones by hematogenous as well as lymphatic route.

Keywords: Esthesioneuroblastom

INTRODUCTION

Olfactory neuroblastoma is an extremely rare malignant tumour of the olfactory epithelium. Originally described in 1924 by Berger[1] et al in the french literature 'esthesioneuroepitheliome olfactif' with the highest series of cases being reported by Jackson et al in 1984 while highest number of cases was reviewed by Skolnic[3] et al in 1966. Lesser than 1000 cases have been reported in world literature with maximum number of cases in the last 2-3 decades because of the advent of newer advanced diagnostic techniques and the easier availability of immunohistochemistry. Olfactory neuroblastoma exhibits a wide range of morphological diversity ranging an indolent growth to highly aggressive neoplasm with propensity of rapid metastasis.

CASE REPORT

We present a case of 70 year old female who presented to the out patient department with the chief complain of swelling on the nose associated with difficulty in breathing and nasal obstruction since one and half years. On examination, thick blood stained foul smelling discharge was seen coming out of the nasal cavity which on cleaning revealed pinkish grey mass filling the right nasal cavity. The mass bled on manipulation. Haematological and biochemical investigations were within normal range.

Radiological investigations: Computed tomography of the nasal cavity and paranasal sinuses showed mildly enhanced rounded well defined mass lesion measuring 1.7x1.7x1.6 cm in the right nasal cavity which appears to arise from the nasal septum by a broad base causing fullness of the nasal cavity. On post contrast scan, mild enhancement of the lesion was noted. Biopsy was adviced.

Gross: Received multiple pink soft to friable tissue bits all aggregating 2.5 cms in 10 % formalin solution.

Microscopy: Haematoxylin and Eosin stained sections studied revealed homogenously small cells arranged in sheets, clusters as well as singly scattered. Individual tumor cells show uniform round to oval nuclei with inconspicuous nucleoli and scanty cytoplasm. Pseudorosette (Homer wright) formation characterized by a loose arrangement with presence of fibrillary material within the lumen.

The histopathological diagnosis of Olfactory Neuroblastoma (Esthesioneuroblastoma) was given. Immunohistochemistry was advised for confirmation of diagnosis (figure 1-3).

DISCUSSION

The tumour was first reported in the english literature by Schall and Lineback[2] at the Massachusetts Eye and Ear Infirmary in 1951, when they reported three cases of ' Primary intranasal neuroblastoma'). Olfactory neuroblastoma (ONB) is an rare malignant neuroectodermal nasal tumour that constitutes about 2% of all sinonasal tract tumours without any specific age, sex or racial predilection.These tumours have been designated previously as esthesioneuroblastoma, esthesioneuroepithelioma, esthesioneuroma, esthesioneurocytoma and olfactory placode tumour. Olfactory neuroblastoma (esthesioneuroblastoma) arises from olfactory neuroepithelium which extends from the superior turbinate and portion of nasal septum upto the roof of the nose.[2] It can also arise from the cells of the sphenopalatine ganglion and jacobson's organ. The olfactory neuroepithelium is somewhat unique as it can be present as low as the medial aspect of middle turbinate. The epithelium is pseudostratified and consists of bipolar neurons, sustentacular cells and basal cells.

The exact cell of origin is controversial and no clear etiological agents have been documented. The tumor has been reported in all age groups and most of the cases described in literature involved adults. It is a slow growing locally invasive tumor with low propensity to metastasis. Metastasis occurs mostly to lungs and bones by hematogenous as well as lymphatic route. Clinically these tumours presents most commonly with polypoidal mass causing nasal obstruction.

Sign and Symptoms

Most common symptoms of olfactory neuroblastoma are nasal obstruction, recurrent epistaxis and headache. Patients with extensively advance tumours may have orbital symptoms because of pressure effect such as proptosis, diplopia, epiphora and nasal discharge. Olfactory neuroblastoma is often

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Microscopic features	Grade I	Grade II	Grade III	Grade IV
Architecture	Lobular	Lobular	+/- Lobular	+/- Lobular
Pleomorphism	Absent to slight	Present	Prominent	Marked
Neurofibrillary matrix	Prominent	Present	May be present	Present
Rosettes	Homer Wright	Homer Wright	Flexner-wintersteiner	Flexner- wintersteiner
Mitoses	Absent	Present	Prominent	Marked
Necrosis	Absent	Absent	Present	Prominent
Glands	May be present	May be present	May be present	May be present
Calcification	Variable	Variable	Absent	Absent

Table-1: Hyam's[8] grading system

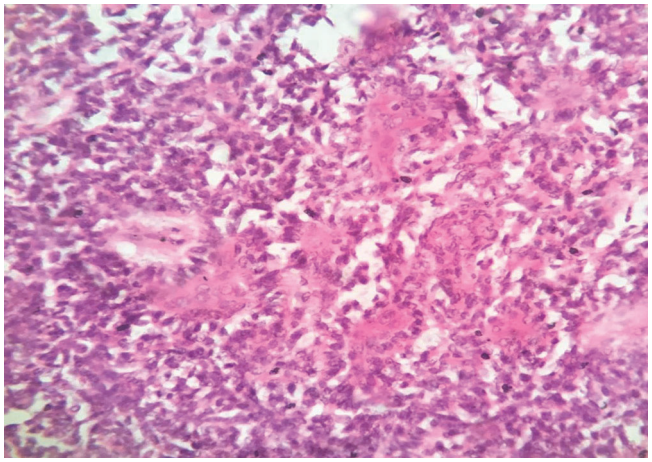

Figure-1: (H and E): Showing formation of Homer- Wright rosettes (pseudorosettes)(40x).

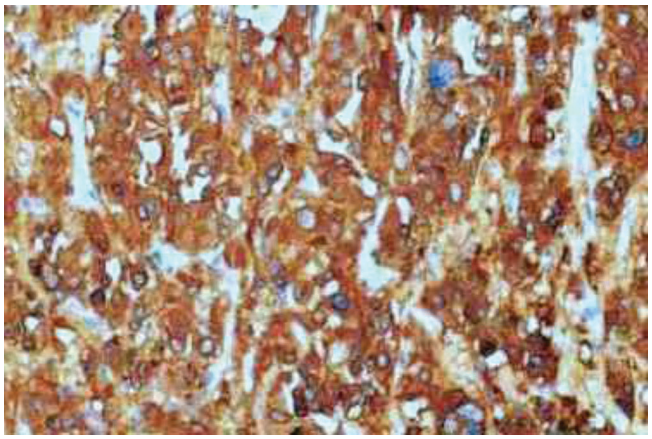

Figure-2: (S-100): Showing diffuse positivity(100x)

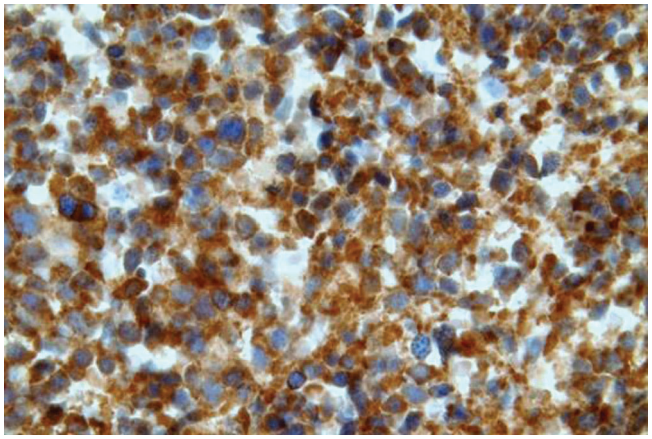

Figure-3: (Synaptophysin): Showing diffuse positivity(100x).

missed during its early stages because of its benign clinical symptoms. Skolnik[3] et al reported in his study of 97 cases of esthesioneuroblastoma that most of the cases reported in Europe and America presented with nasal polypoidal mass causing nasal obstruction and bilateral proptosis.

Investigations

High resolution Computed tomography scan and magnetic resonance imaging can be used as supportive investigations to accurately delineate the extent of the tumor into surounding structures such as cribriform plate, fovea ethmoidalis, anterior cranial fossa and retromaxillary space. Olfactory neuroblastoma does not have a specific radiological feature and appears as a homogenous soft tissue mass with uniform contrast enhancement. Computed tomography images are essential for the accurate staging and to see for erosion of bones. Though confusion may arise when trying to differentiate obstruction of sinus ostia draining leading to accumulation of nasal secretion from tumor tissue.

Magnetic Resonance Imaging is better in delineating intracerebral or intraorbital extensions and appears as isotense or hyper intense to gray matter on T2-weighted images. Endoscopic examination is required for obtaining biopsy specimens and for evaluating the extent of the tumor.

Histological features

The typical histopathological appearance of well differentiated olfactory neuroblastoma exhibits homogenous small cells with uniform round to oval nuclei with rosette formation against a fibrillar background. McCormack[4] et al described three different histopathological types of olfactory neuroblastoma. True rosettes (Flexner-winter Steiner) refers to a ring of columnar cells surrounding a central space while pseudorosettes (Homer wright) are characterized by a loose arrangement with presence of fibrillary material within the lumen.

It is often difficult to differentiate it from malignant melanoma, rhabdomyosarcoma, undifferentiated carcinoma and extramedullary plasmacytoma.[5]

Occasionally special silver stains such as Bodian, Grimelius and Churukian-schenk may be used in highlighting the neurosecretory granules. Immunohistochemistry can be used as confirmatory test in conjuction with histopathological findings. Olfactory neuroblastoma shows positivity for neuroendocrine markers such as- S-100 protein, neuron specific enolase, chromogranin and synaptophysin.

Staging systems

Malignant lesions of the nasal cavity are extremely rare with prevalence of different histopathological types, hence no approved classification and staging system has been accepted

universally.

Kadish[9] et al in 1976 proposed the first well accepted staging classification (Stage A, B and C). Stage A- Disease confined to the nasal cavity, Stage B- Disease extending beyond nasal cavity to one or more paranasal sinuses and Stage C- Disease extending beyond to orbit, base of skull intracranial cavity, lymphnodes or distant metastases.

Biller[6] et al in 1990 proposed a staging system based on the TNM classification. It takes into account the size of the primary tumor and the presence or absence of regional and distal metastasis. T1- Tumor involving the nasal cavity and paranasal sinuses, extending the sphenoid with or without the erosion of the anterior cranial fossa, T2- Tumor extending into the orbit or protruding into the anterior cranial fossa, T3- Tumor involving the brain that is resectable with margins, T4- Unresectable tumor.

Dulgerov[7] et al in 1992 proposed their staging system based on TNM staging and CT and MRI findings. T1- Tumor involving the nasal cavity and sinuses (except sphenoid), T2- Tumor in the nasal cavity and sinuses (including sphenoid) with extension to or erosion of the cribriform plate, T3- Tumor extending into the orbit or into the anterior cranial fossa without dural invasion(extradural), T4- Tumor involving the brain, N0- No cervical lymphnode metastasis, N1- Any form of cervical lymph node metastasis.

Based on the degree of differentiation the olfactory neuroblastoma can be separated into four grades (grade I to IV) (table-1).

CONCLUSION

Our case was graded according to the Hyam's pathological grading as Grade II and staged as Stage A. It has a propensity for rapid metastasis. So in every patient presenting with nasal obstruction and epistaxis and intracranial signs and symptoms in the 2nd to 4rth decade should be evaluated for esthesioneuroblastoma. Because of its close resemblance to round cell tumours like lymphoma, rhabdomyosarcoma, undifferentiated carcinoma and malignant melanoma special immunohistochemical stains has become very essential for proper and accurate diagnosis.

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