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LEPTOMENINGEAL SPREAD IN AN ESTHESIONEUROBLASTOMA

Radiology

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ABSTRACT

Esthesioneuroblastoma is a rare neoplasm of neuroectodermal origin, arising from the olfactory epithelium in the roof of the nasal cavity. The incidence curve for this disease has a bimodal shape with the first peak in the 2nd decade and 2nd peak in 6th decade (1). Extension to the brain can occur less frequently than local recurrence, but leptomeningeal metastases without brain involvement have rarely been mentioned. We report a case with leptomeningeal metastases in an esthesioneuroblastoma.

KEYWORDS:

Esthesioneuroblastoma, Olfactory Neuroblastoma, Leptomeningeal Metastasis, Nasal Cavity Tumors.

INTRODUCTION:

Olfactory neuroblastoma, or esthesioneuroblastoma (ENB), is an uncommon malignant tumor of neural crest origin, arising from the olfactory epithelium of the superior nasal cavity. It accounts for ~ 2 to 6% of the intranasal tumors, with an incidence of ~ 0.4 per million. (2, 3, 4) The age at presentation is bimodal in distribution in the second and sixth decades of life and without a gender predilection.(1) Sinusal, orbital and intracranial extensions are common (5). Leptomeningeal spread of the disease is a very rare site of involvement. (6) We, hereby present MRI findings in a case of leptomeningeal spread of an esthesioneuroblastoma.

CASE REPORT:

A 20 yr old boy presented with difficulty in breathing and a left nasal cavity mass causing blockage, that has gradually increased over few months. Tissue sampling of the mass was done and histopathological diagnosis of esthesioneuroblastoma was made. MRI of the neck and paranasal sinuses with post contrast screening of brain was performed. Magnetic resonance imaging (MRI) findings (Fig 1-3): showed a large heterogeneously enhancing destructive, expansile mass in the left nasal cavity with orbital, maxillary sinus, nasopharyngeal, cavernous sinus. It showed predominant T1 /T2 isointense signal and few peripheral cysts on T2 weighted images. There is an extraaxial lesion with similar morphology in the right anterior temporal fossa with moderate edema in the adjacent anterior temporal lobe (Fig 3, 4). Linear and nodular meningeal enhancement was noted in the left parietal hemisphere on the screening post contrast sequence.(Fig 5).It was stage C as per KADISH staging system(7)and T4N0M1 as per DULGUEROV et CALCATERRA (1992 criteria.(8) The patient underwent chemoradiation for reduction of the tumor bulk before resection of the tumor. Further spine screening for meningeal metastasis was negative.

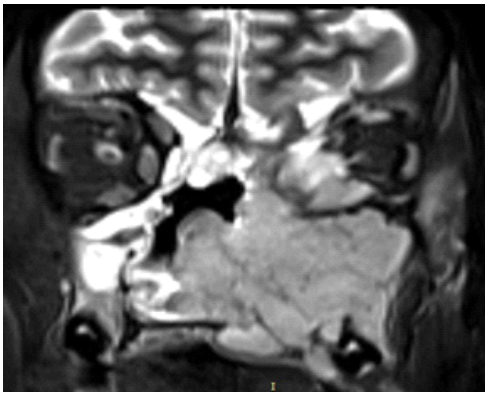

Fig 1- Coronal T2W MRI showing well defined isointense mass in left nasal cavity extending into maxillary sinus and orbit.

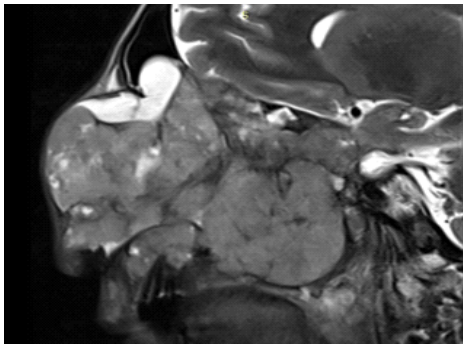

Fig 2-Sagittal T2 MRI showing well defined isointense mass in left nasal cavity extending into sphenoid sinus, without brain parenchymal extension into the frontal lobe.

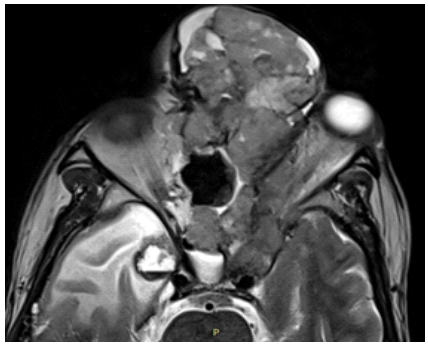

Fig 3-Axial T2 MRI showing extension into the left cavernous sinus and also the right temporal meningeal deposit with resultant anterior temporal lobe parenchymal edema.

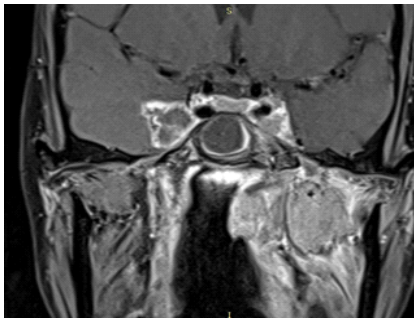

Fig 4-Coronal T1W post contrast fat saturated images showing the left cavernous sinus extension with peripherally enhancing right anterior right temporal meningeal deposit

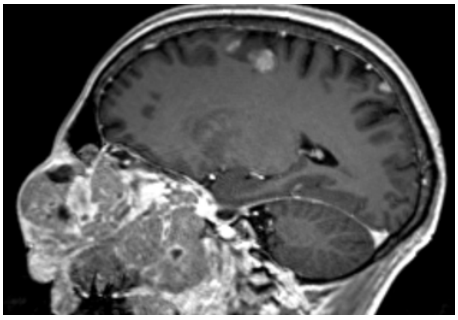

Fig 5-Sagittal T1 Post contrast fat sat MRI showing well defined heterogeneously enhanicng mass in left nasal cavity with multiple nodular deposits in the parietal region (arrows)

DISCUSSION:

Esthesioneuroblastoma is an uncommon malignant tumour that arises from bipolar sensory receptor cells in the olfactory mucosa. These cells originate in the neural crest and differentiate into the olfactory sensory elements. Esthesioneuroblastomas are histologically similar to adrenal or sympathetic ganglionic neuroblastomas and retinoblastomas. [9]

The mass may be relatively slow growing for malignant tumour and cause some expansion and remodeling of bony structures. This neoplasm is locally aggressive and cause metastasis by lymphatic and hematogenous routes. [10] Local recurrence has been reported in upto 57% of patients. A metastatic rate of 20% to 60% is reported with the most common site being the cervical lymph node. Other sites include the parotid glands, skin, lungs, bone, liver, orbit, spinal cord and spinal canal. [5]

Metastasis to the central nervous system is infrequent and usually identified only on post-mortem examination. In the spinal cord 80% of metastasis is in the cauda equine.

The diagnosis and evaluation of staging of esthesioneuroblastoma can be done by CT which provides the best information about the tumour invasion. On T1W MR Images, esthesioneuroblastoma presents as homogenously enhancing tumour with intermediate signal intensity, where as T2W images, the original intensity is increased. MRI can delineate intraorbital and intracerebral extension. The tumour appears hypo to gray matter onT1WI and iso or hyper to gray matter on T2WI. Gadolinium enhanced MR images help to differentiate tumour from obstructed secretions in paranasal sinuses, determining meningeal and extradural spread and to detect perineural spread. [11]

The staging system of KADISH et al. (1976) (7)
Stage A: tumour localised to the nasal cavity
Stage B: spread to sinuses
Stage C: extension over paranasal sinuses
The main drawback of Kadish classification is the language of defining stage C is too broad.
In 1992, Dulgerov and Calcaterra proposed modified TNM staging based on CT and/or MR findings(8).
Modified TNM according to DULGUEROV et CALCATERRA (1992)
T1: tumour localised to the nasal cavity and
paranasal sinuses with a space between tumour
and lamina cribosa
T2: tumour developped in nasal cavity or sinuses
but in contact with cribiform lamina and/or
sphenoid extension
T3: tumour with intracranial extradural and/or
orbital expansion
T4: tumour with intracranial intradural extension
N0: no metastatic cervical nodes
N1: metastatic cervical nodes
M0: no distant mestastases
M1: distant

The role of imaging in these patients is to map the tumour so as to precisely anticipate surgical boundaries. Our patient had rare extension to the leptomeninges without direct brain parenchymal involvement which is suggestive of poor prognosis (12)

CONCLUSION

MRI is important for staging, as it exhibits the extent of soft tissue invasion with involvement of local structures, such as orbits and sinuses. The use of MRI is crucial for discriminating between postobstructive secretion and tumor tissue. In addition lack of contrast uptake of sinusal retention cysts on Gadolinium enhanced MR images can help to differentiate tumour from obstructed secretions in paranasal sinuses, and is useful in determining meningeal and extradural spread and

perineural spread. Enhanced images, particularly in the sagittal plane, were very helpful

in identifying meningeal extension in our case. Thus, olfactory neuroblastomas can be detected, delineated and its characteristics suspected by MRI, but definite diagnosis however is still based on histopathology. We thus present a rare case of esthesioneuroblastoma with leptomeningeal spread and significance of MRI in extent and staging of the same.

DISCLOSURE Conflict of interest: None

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