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Primary neuroendocrine tumour of petrous part of temporal bone - A rare entity

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Abstract

Skull based neuroendocrine Tumors are rare Tumors with only a few case reports in literature. Here we present an unusual case of primary neuroendocrine tumor of petrous part of temporal bone in a 62 year old male which was proven on surgical biopsy. The primary features of this rare tumor along with differential diagnosis are discussed with brief review of the literature.

Keywords: Neuroendocrine, skull base, Dural based space occupying lesion at petrous ridge.

Introduction

The petrous apex is a complex region of the central skull base that is surrounded by a number of important vascular and neural structures and can be home to a wide range of disease processes. Lesions arising in or spreading to the petrous apex cause varied and occasionally severe clinical sequelae which typically result from mass effect or direct invasion of the cranial nerves, brainstem, or internal carotid artery. Because the petrous apex is not amenable to direct examination, cross sectional imaging with computed tomography and magnetic resonance imaging plays an important role in diagnosis and characterization of lesions occurring there Petrous apex lesions can be classified on the basis of their origin into the following categories: developmental lesions, inflammatory lesions, benign tumours, malignant tumours, vascular lesions, osseous dysplasia. Primary skull base neuroendocrine tumours are rarely mentioned in the literature. We report a rare case of a pathologically proven neuroendocrine tumour of skull base, in the middle cranial fossa.

Extensive metastatic workup revealed no other primary lesion, indicating this to be the primary site of involvement.

Case report

A 62 year old male presented with history of headache, right sided ear discharge and deviation of the angle of the mouth to the left side, of 1 year duration. On examination, there was right lower motor neuron seventh cranial nerve palsy and right aural serosanguinous discharge. For further assessment, the patient was referred for computed tomography and magnetic resonance imaging of the brain including the skull base.

A number of tumours and tumour-like non- neoplastic lesions with different cell types on histology occur in the skull base. A wide variety of disease and lesion appearance often complicates the process of radiological diagnosis.

The main role of radiographic imaging is in the detection and characterization of skull base lesions, with the aim of delineating and mapping the extent of involvement and for the preservation of adjacent critical organs.

Evaluation of the skull base anatomy and surgical planning by using image guidance are of great help to surgeons.

HRCT of the right temporal bone with contrast demonstrated nodular soft tissue polypoidal thickening involving right middle ear cavity in epitympanum, mesotympanum and hypotympanum with extension into the mastoid cavity, through the aditus ad antrum. There was significant post-contrast enhancement on arterial phase with washout on venous and delayed phase.

On plain scan, the lesion was mildly hyperdense. On arterial phase, the lesion was hyper enhancing with wash out on venous phase.

Encasement of the ossicular chain was seen with erosion of long process of incus and non-visualisation of the in cudo-stapedial joint. Minimal extension of this soft tissue density in external ear with opacification of mastoid air cells was seen.

Stapes with both crus was seen.

Incidentally noted, was a large mass involving right temporal lobe. Reactive changes in adjacent bone with significant oedema was seen in the adjacent neuroparenchyma.

Malleo- incus joint was normal.

The visualized cochlea and semi-circular canals were normal. The internal auditory meatus was normal. The visualized tegmen tympanic and the sigmoid sinus plate were intact. There was thickening of right facial nerve in its entire course.

These HRCT findings were in favour of a right middle ear cavity mass.

No abnormality was seen in the left temporal bone.

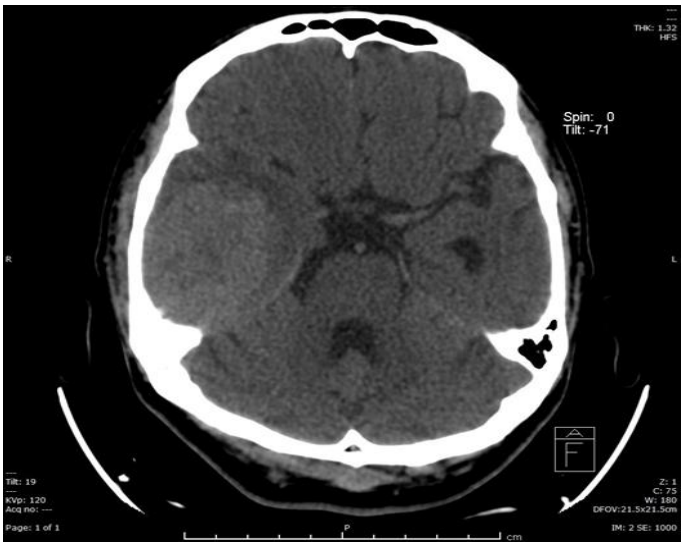

Figure 1:

Plain axial CT of brain shows well defined hyperdense extra-axial lesion in the right temporal convexity, with adjacent mass effect.

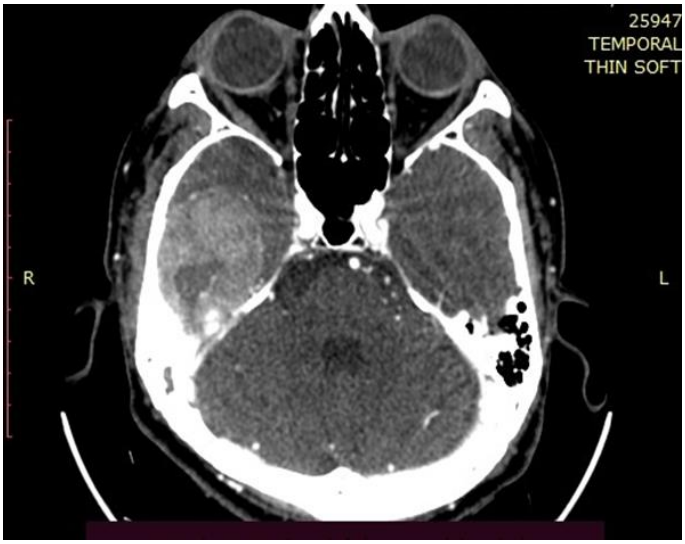

Figure 2:

Axial Post contrast CT image shows extra axial lesion in right temporal convexity showing heterogenous enhancement on post contrast scan.

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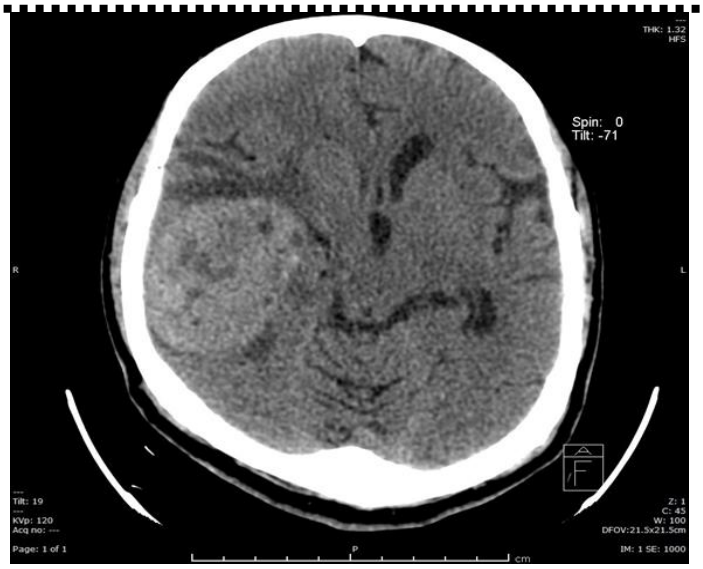

Figure 3:

This image shows heterogenous enhancement on post contrast scan with mass effect, midline shift and indentation on mid brain.

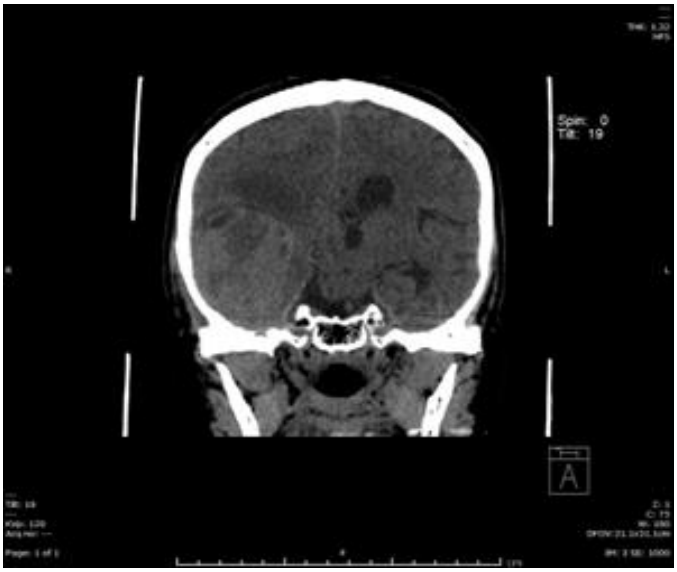

Figure 4:

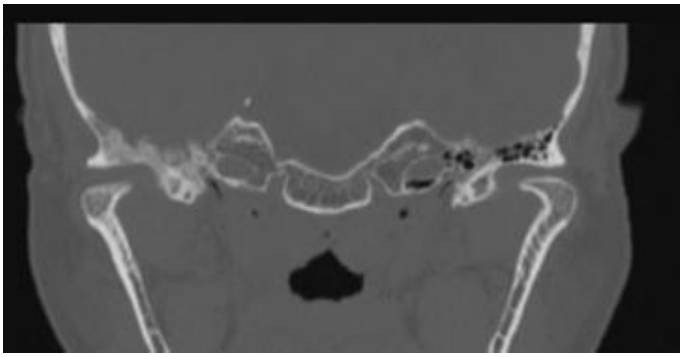

Figure 5:

Coronal reformatted soft tissue and bone window shows erosion of right petrous temporal bone.

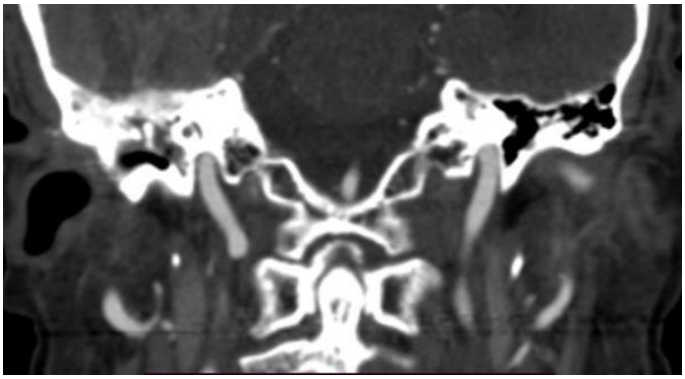

Figure 6:

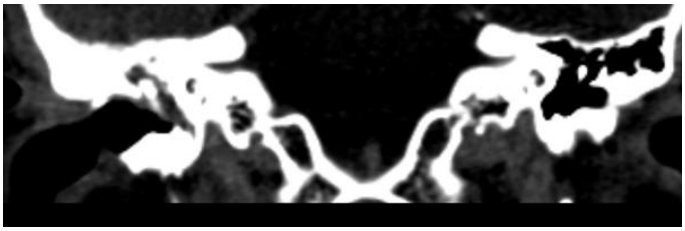

Figure 7:

Reformatted Coronal post contrast image reveals Encasement of the ossicular chain by hetrogenously enhancing soft tissue with erosion of long process of incus and non-visualisation of the incudo-stapedial joint.

MRI brain showed a moderate size large rounded lesion in the right parietotemporal region. The lesion was isointense on T1W ,hyperintense on T2W and showed heterogeneous post-contrast enhancement with central non enhancing cystic/ necrotic areas. The lesion showed patchy areas of restricted diffusion with areas of old haemorrhage within.

The lesion showed a broad based towards the right parietal and temporal region. There was moderate subcortical oedema in the right parietotemporal region with mass effect on the adjacent parenchyma with early midline shift and there was mild prominence of the left lateral ventricle with indentation on the brainstem.

The right temporal bone showed areas of sclerosis with soft tissue in the right middle ear cavity.

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Possibility of an extra-axial lesion, likely meningioma in the right parietotemporal region was considered.

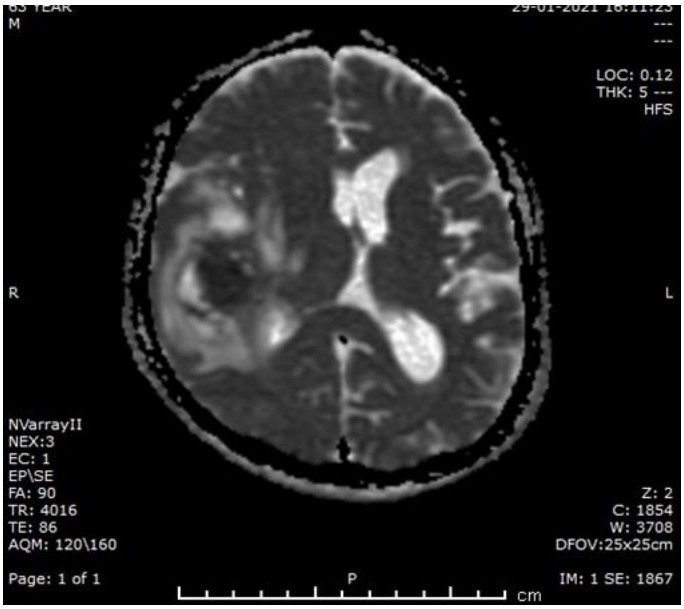

Figure 8:

ADC – shows patchy areas of reduced value on ADC map.

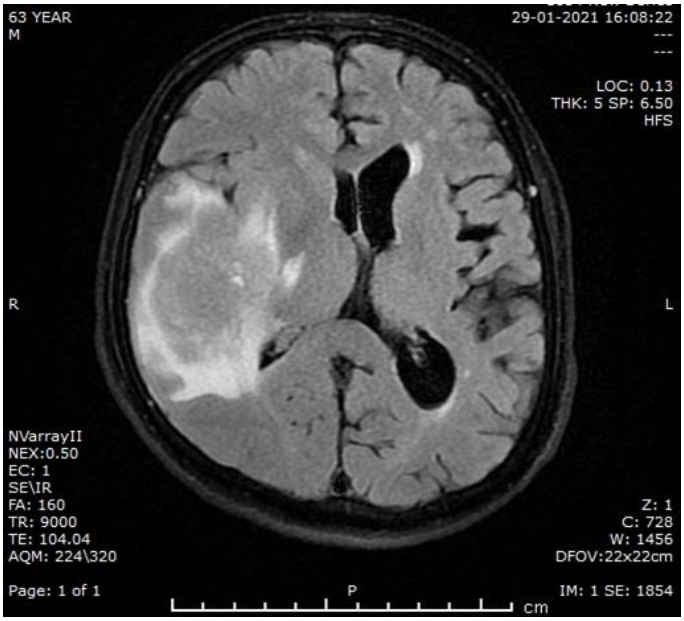

Figure 9:

Axial FLAIR image shows a large, well defined oval extra-axial lesion in the right parieto- temporal convexity, with mass effect, midline shift and oedema in adjacent white matter and causing indentation on the midbrain.

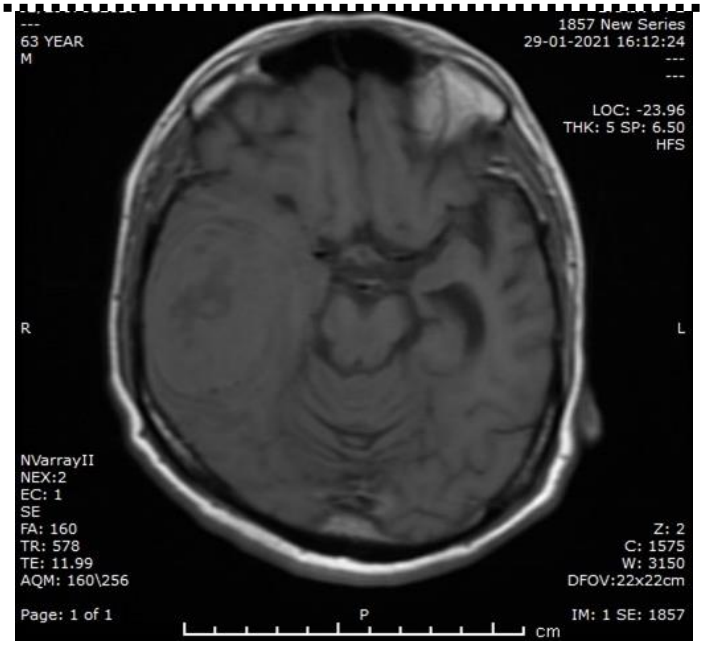

Figure 10:

Axial T1W image shows heterogenous signal intensity in the lesion.

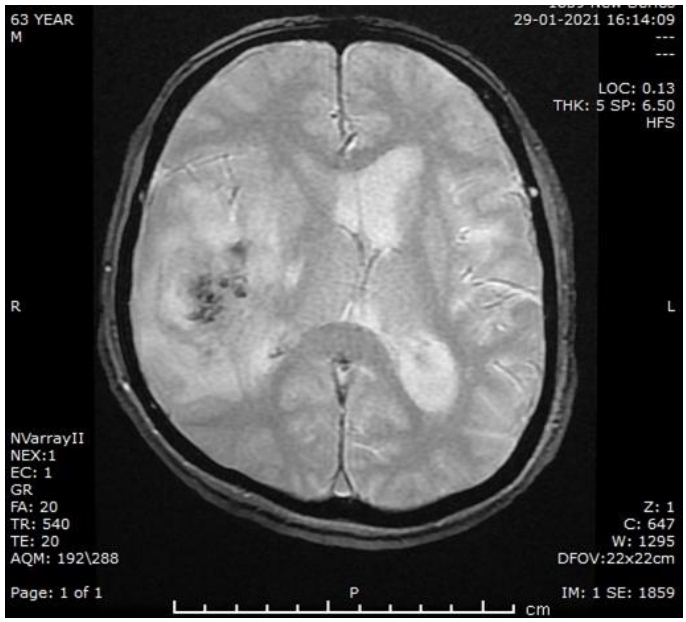

Figure 11:

GRE- Areas of blooming are seen, suggestive of hemosiderin deposition.

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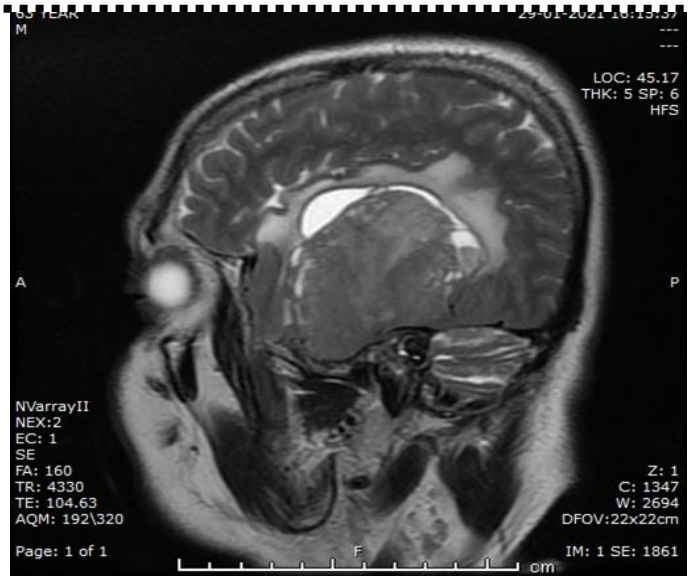

Figure 12:

Sagittal T2 W image through right temporal region showing extra axial lesion in right temporal lobe appearing heterogenous predominately hypointense on T2 W images with mass effect.

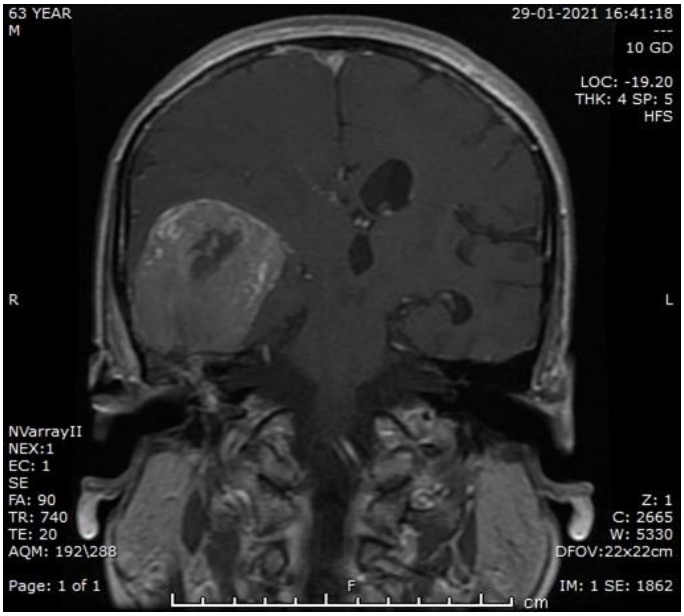

Figure 13:

On post-contrast coronal T1W fat sat image, heterogenous enhancement with lesion extending into the petrous and mastoid temporal bone on the right side.

Events

A craniotomy with excision of the space occupying lesion was performed and was reported as an atypical meningioma by histopathologist. Immunohistochemistry (IHC) showed an AE1/AE3, few tumor cells positive for chromogranin A ,CD 56 and showed weak positivity to synaptophysin , negative for glypican 3, PSA, PAX 8, CK7, TTF1, C kit, MiB labelling index was 5-7%. Possibility of a neuroendocrine tumour, grade II (WHO 2017) was suggested.

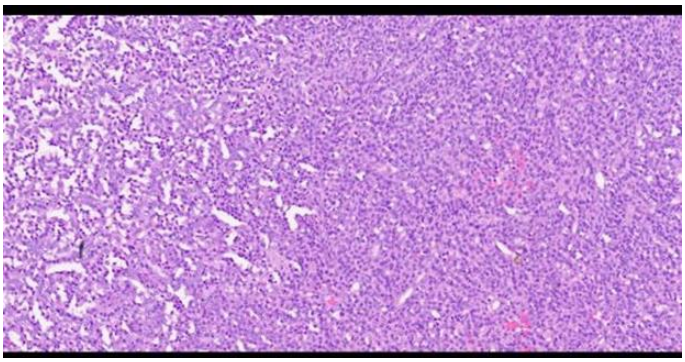

Figure 14:

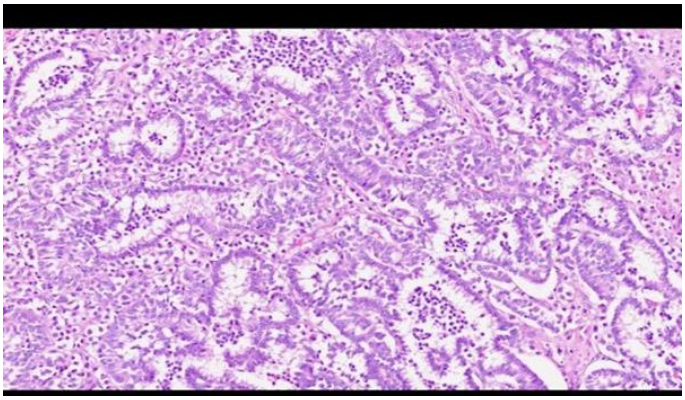

Figure 15:

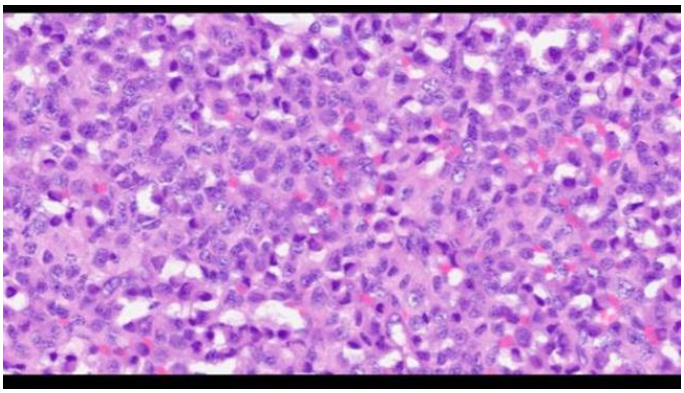

Figure 16:

H & E Stain---These sections show a circumscribed cellular tumour composed of cells arranged as nested aggregates, tubules and papillae. The tumour cells show

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round to oval nuclei with finely clumped chromatin, inconspicuous nucleoli and pale eosinophilic cytoplasm. Mitosis was scarce.

IHC---

The tumour cells expressed EMA, cytokeratin, synaptophysin and chromogranin A. Immunonegative for CK 7, CK 20, TTF1. The MiB -1 labelling index was approximately 5%.

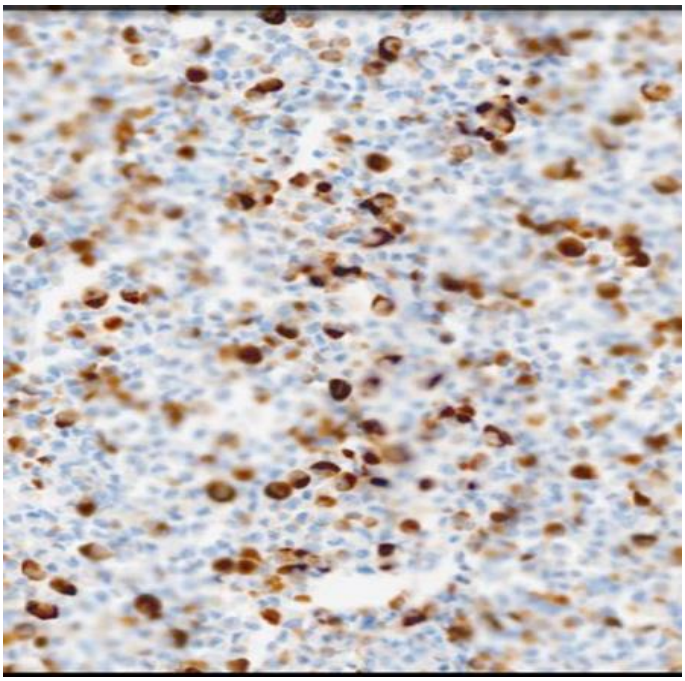

Figure 17:

IHC—CHROMOGRANIN: Scattered positivity in the tumour cells.

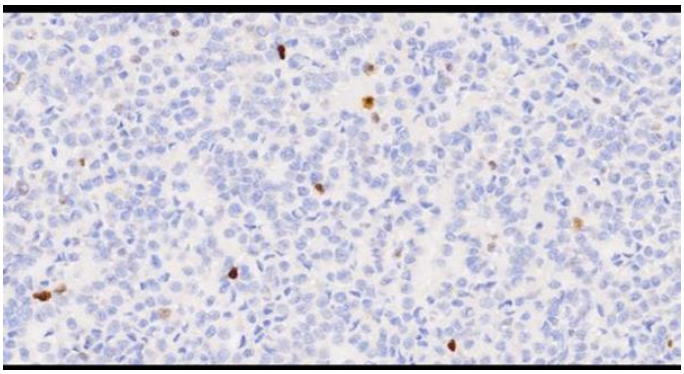

Figure 18:

IHC—Ki 67---5%

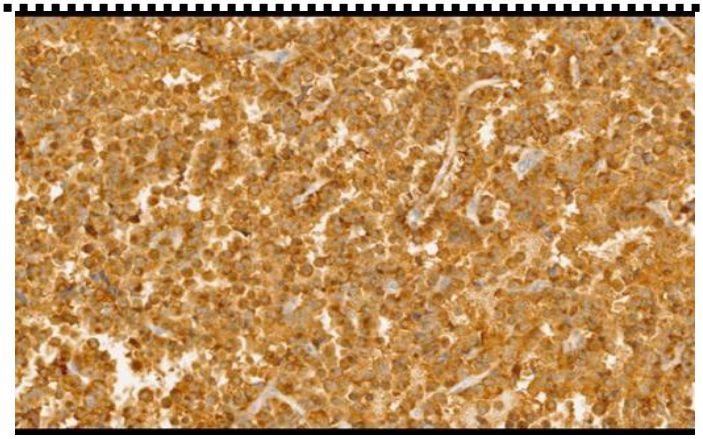

Figure 19:

IHC--SYNAPTOPHYSIN

Diffuse strong positivity in tumour cells

It was reported as a Neuroendocrine tumour (NET) G2, WHO 2017

FDG PET -CT done within a month of the surgery revealed few areas of lytic erosion of tegmen tympani on the right side, closely abutting the inferomedial aspect of the reconstructed flap margins.

Mild low-grade FDG uptake/mucosal soft tissue thickening was noted involving the epitympanum, tympanum and hypotympanum region of the middle cavity. No other lesion was seen.

After the IHC confirmed the lesion to be a neuroendocrine tumour, two subsequent DOTANOC PET-CT scans done over a gap of 2 to 3 months, revealed a gradual increase in DOTANOC uptake in the lesion seen involving the right mastoid and petrous bone at the base of the skull. Loss of pneumatization with diffuse sclerosis was seen in the right mastoid region. Mild increase in hypodensity was noted in the right temporal lobe.

Radiation

Radiation therapy was given. Type of Radiation therapy (IG-IMRT) , Delivered dose (55.8Gy) /Fractions (31 fr)/ I

Partial improvement was seen.

The latest PET- CT scan done showed no active lesion.

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Discussion

Neuroendocrine tumours (NET) originate from the amine precursor uptake and decarboxylation cells (APUD cells) of the diffuse neuroendocrine system. [2, 6]

Primary intracranial NETs are extremely rare entities, with only few reported cases in literature, most of which were located at the skull base and seller region.[8]

Incidence of intracranial neuroendocrine tumour in brain and middle ear is 0.8% and 2% respectively. [1]

Neuroendocrine cells are involved in the coordination of the neurotransmitter- initiated synthesis and release of biologically active substances into the blood.[8]

NETs can be categorised clinically as functional or non-functional. Functional tumours secreting one or more hormones would result in endocrine symptoms [1-2]. Additionally, they can be categorised by their anatomical location and by their grade.

In biochemical measurements, Chromogranin A, may be used as a tumour marker, as it increases in 70-90% of patients with NET. [6]

Final diagnosis of NET depends upon the pathological characteristics of the tumour.

Malignancy is divided into three levels according to the Ki67 labelling index. Ki-67 is a function of the degree of proliferation because it is expressed in the actively dividing cells, with 1-3% low level, 3-20% middle level and above 20% high level. [2]

Primary skull base neuroendocrine tumours are rare entity. Liu et al describe 2 cases of primary intracranial tumours centered in the seller / suprasellar region and the anterior cranial fossa.

They concluded that the mechanism , diagnosis, and treatment of NET are still challenging. Surgical resection followed by radiotherapy has demonstrated an effective treatment, but chemotherapy still needs researchers to demonstrate its therapeutic efficacy. [1-2]

DE shales and Huang reported central nervous system (CNS) carcinoid mimicking a meningioma. They concluded that intracranial carcinoid should be included in the differential diagnosis of dural based, extra- axial brain lesions.[3]

Vishwanath Vijay Joshi et al reported an unusual rare case of primary neuroendocrine carcinoma of the skull base, proven on surgical biopsy. They concluded that the diagnosis of NET depends on the histopathological characteristics and immunohistochemistry of the tumour.[1]

Baxi A. J, et al concluded that primary neuroendocrine tumours of skull are highly unusual and rare, metastasis is more common. They account for 1.5% of all patients with carcinoid tumours. [4]

Optimal workup of NETs requires use of a combination of conventional and somatostatin- based imaging techniques. Cross sectional and functional imaging play an important role in diagnosis, lesion characterisation and staging of skull base tumours. On imaging, there are no specific features of neuroendocrine carcinomas. Bone erosion on CT scans, hypo intensity on T1W, hyperintensity on T2W and heterogenous enhancement are general characteristics of these tumours. Common pathologies like glomus tumour, meningioma, schwannoma and metastasis form part of differentials to be considered when evaluating the lesion based on its location ,in the skull base.[1]

As most neuroendocrine tumours express type 2 Somatostatin receptors, scintigraphy is widely used as the primary imaging method for the diagnosis, staging and monitoring of neuroendocrine tumours. FDG PET is used to detect malignancy for a variety of tumour types. Unfortunately, majority of NETs tend to be relatively metabolically inactive and fail to take up the tracer well. However, high grade NETs are more likely to

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demonstrate avid uptake of 18 FDG, giving these scans utility in identifying tumours likely to display more aggressive behaviour.[1]

Finally, the diagnosis of NET depends on the pathological characteristics and immunohistochemistry. Multidisciplinary approach is required for accurate diagnosis, lesion characterisation, localisation, staging and monitoring treatment response.

The mechanism, diagnosis and treatment of Neuroendocrine tumours is still challenging. Surgical resection followed by radiotherapy appears as an effective treatment option. Chemotherapy still needs research to demonstrate its therapeutic efficacy. [2]

Conclusion

Primary skull base neuroendocrine tumours are extremely rare and are mostly metastatic from the gastrointestinal tract or the respiratory tract. The radiological appearances are also not specific, showing similar findings in meningiomas, schwannomas, glomus tumor or metastasis. Hence, it is essential to do a full body imaging, using a combination of anatomic and functional imaging to rule out a primary lesion away from the CNS. In this regard, scintigraphy has demonstrated its usefulness for the location of radiologically occult tumours. [7]

Pathological diagnosis is the gold standard. Synaptophysin and Chromogranin A are sensitive but not specific markers, and several immunohistochemical markers must be combined to arrive at a diagnosis. Their malignant potential depends upon the Ki 67 index.

Treatment should be conducted in a multidisciplinary manner with multiple modalities if required, including surgery, radiotherapy and chemotherapy. Assessing skull bone erosion by CT before radiotherapy and performing radiological follow up examinations are recommended for a more favourable clinical course.[8]

Declaration of interest

None declared under financial ,general and Institutional competing interests.

References

1. Vishwanath Vijay Joshi, Pranav Thombare, Mitusha Verma, Neemish Kamat, D Patkar: Primary skull base neuroendocrine carcinoma--case report and review of literature. Radiol Case rep, 2020 Jul; 15 (7): 1071-77.

2. Liu h; Wang H; Qi X; Yu C; Primary intracranial neuroendocrine tumour : two case reports. W J Surg Oncol 2016; 14:138

3. DE shales E.M.; Adamo M.A. Qian J; DiRisio D.A. A carcinoid tumour A carcinoid tumour mimicking an isolated intracranial meningioma J. Neurosurg. 2004; 101:858-860.

4. Baxi A.J., Chintapalli K., Katkar A., Restropo C.S.,---Multimodality imaging findings in carcinoid tumours; a head to toe spectrum. Radiographics.2017;37:516-536

5. Deshaies EM, Adamo MA, Qian J, DiRisio DA. A carcinoid tumor mimicking an isolated intracranial meningioma. J Neurosurg 2004;101:858–60.

6. Hai long Liu, Mingshan Zhang, Xuan Wang et al; Primary intracranial neuroendocrine tumour with ectopic adrenocorticotropic hormone syndrome—A rare and complicated case report and literature review; Molecular and Clinical Oncology 5; 99-102, 2016.

7. Idoya Zazpe, Daniel D Frutos, Carlos Beaumont et al; Primary neuroendocrine tumour of the sphenoid sinus : Three case reports; J Neurol Surg B Skull Base 2020, 81 (S 01) : S1-S272.

8. Atsushi Kambe, Sadao Nakajima, Masami chi Kurosaki; Primary intracranial neuroendocrine tumour of the skull base complicated with tension pneumocephalus after radiotherapy; NMC Case Report Journal; the Japan neurosurgical society.

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