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Case Report

Pathology Section

Extraosseous Ectopic Meningioma on Temporoparietal Region Mimicking a Soft Tissue Tumour: A Case Report

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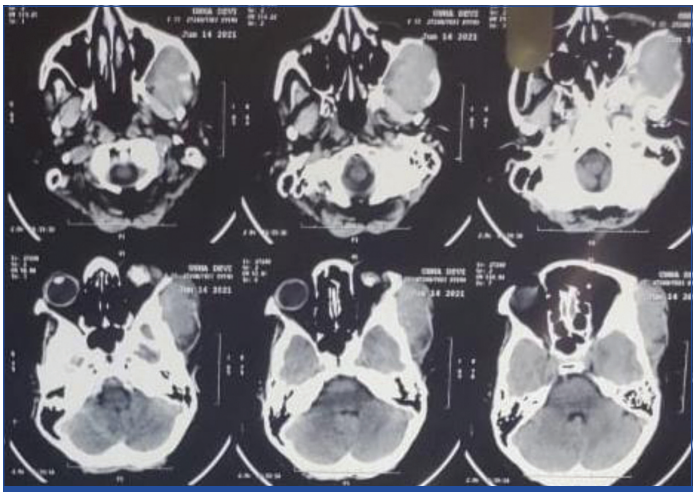
ABSTRACT

Ectopic meningiomas are very rare tumour entities that account for approximately 1-2% of all meningiomas and appear extracranially mostly in the head and neck region. Usually they create diagnostic confusion in an ectopic site with other soft tissue neoplasms, mostly peripheral nerve sheath tumours, myogenic tumours and vascular tumours, more so, with morphological variants like fibroblastic, angiomatous, etc. that may not show the meningothelial cells conspicuously. Here authors report one such case of a 55-year-old female, presenting with a slowly growing non tender left temporal mass since childhood that was thought to be a myogenic tumour on Computed Tomography (CT) scan but on microscopy was a transitional meningioma with mixed fibroblastic and meningothelial pattern. Hence, ectopic meningioma is one of the imperative diagnosis that a pathologist should keep in mind while dealing with soft tissue neoplasms of the head and neck region.

Keywords: Calvarial meningioma, Epithelial membrane antigen, Intranuclear inclusions, Meningeal rests

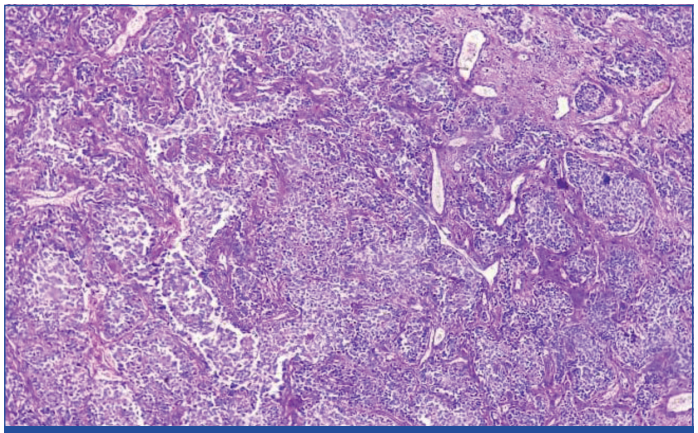
CASE REPORT

A 55-year-old woman presented to the Department of Neurosurgery with chief complaint of a slowly growing mass behind the helix of left ear since childhood and gradually progressing upwards and forwards on the lateral part of left-side of forehead. Physical examination revealed a swelling of 4×3 cm in size which was firm in consistency, mobile and non tender (clinical image not available). Further systemic examination revealed no derangement in the neurological function of the patient. All haematological investigations were unremarkable too. There was no any other significant medical history in the past or present. The patient underwent a Computed Tomography (CT) scan which disclosed a soft tissue lesion on the temporoparietal region with no intracranial extension [Table/Fig-1]. A provisional diagnosis of a benign soft tissue tumour of either vascular or myogenic origin was suggested. For confirmation, the tumour was excised and the tissue was sent to us for histopathological examination.

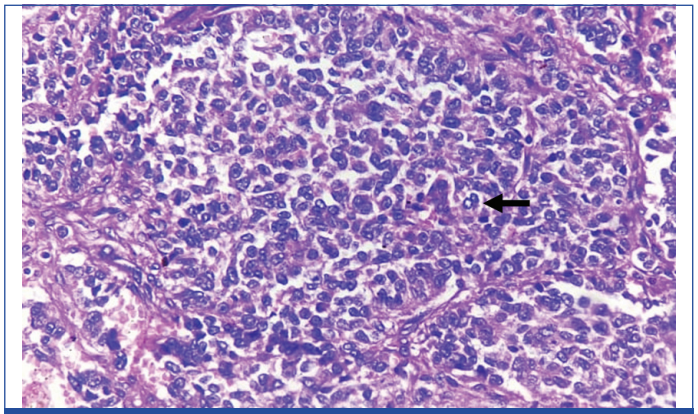


[Table/Fig-1]: CT scan image was suggestive of a soft tissue neoplasm with homogenous contrast enhancement and no intracranial extension predominantly on the temporal region extending posterosuperiorly on the parietal region.

Grossly, it was received in multiple pieces that were firm and greyish white. On microscopy, it was composed of monomorphic neoplastic epithelioid cells arranged mainly in sheets and lobules with numerous proliferating capillaries [Table/Fig-2]. These cells were of uniform size, had central nuclei and prominent intranuclear inclusions with moderate amount of eosinophilic cytoplasm (meningothelial cells) [Table/Fig-3]. At some places the cells were seen acquiring a spindled morphology running in intersecting fascicles [Table/Fig-4]. In addition, few psammoma bodies were seen ([Table/Fig-4] Inset). There was no significant atypia, necrosis or mitosis. The findings were suggesting a meningioma, however, in view of patient's age

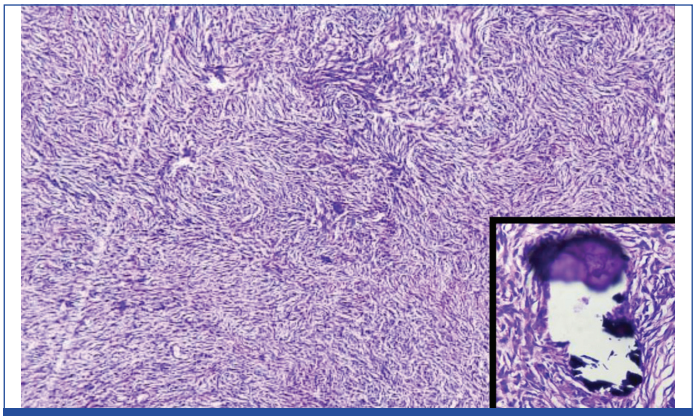


[Table/Fig-2]: Sections showing nests and lobules of tumour cells separated by fibro collagenous septa, few staghorn blood vessels can also be seen (Meningothelial pattern) (H&E, 10X).

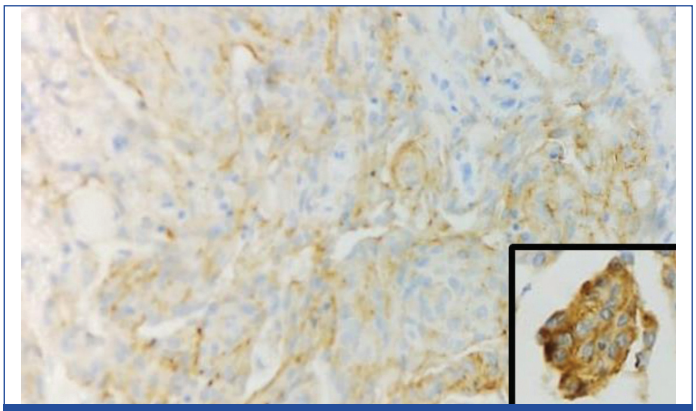


[Table/Fig-3]: Meningothelial cells in syncytium showing high nuclear: cytoplasmic ratio, vesicular chromatin with prominent nucleoli, at some places nuclear inclusions can also be seen (arrow) (H&E, 40X).

and site in mind, a differential diagnosis of an amelanotic malignant melanoma was also considered. Immunohistochemical staining revealed diffuse positivity for Epithelial Membrane Antigen (EMA) [Table/Fig-5] and negativity for Human Melanoma Black 45 (HMB45). Hence, this case was finally diagnosed as an ectopic transitional meningioma {World Health Organisation (WHO) grade 1} [1]. No additional therapy was given to the patient and she was doing apparently well when she came for follow-up at three months and six months.



[Table/Fig-4]: Sections from other areas showed spindled tumour cells arranged in compact intersecting fascicles and vague storiform pattern (fibrous pattern) (H&E 10X). Inset- psammoma body (H&E, 40X).



[Table/Fig-5]: Tumour cells showing strong EMA positivity (IHC, 10X). Inset- Tumour cells displaying the classical membrane staining (IHC, 40X).

DISCUSSION

Meningiomas, one of the most common tumour entities in the central nervous system are generally benign and have their origin in the meningocytes of the arachnoid villi. However, in rare cases, ectopic forms of this tumour entity can appear extracranially constituting fewer than 1.6% of all meningiomas [2]. Epidemiologically, ectopic meningiomas are slightly more frequent in females with a ratio of 1:1.2 [3,4]. Primary ectopic meningiomas occur mostly in the head and neck region (orbit of the eye, nose, paranasal sinuses, jaw bones and ear) or paraspinal soft tissues. Other rare sites include foot, skin, retroperitoneum and mediastinum [4,5].

In India, few individual case reports of ectopic meningiomas have been published till date, out of which maximum cases were in the paranasal sinuses, orbit and scalp with few documented in rare sites such as palatine tonsil, parapharyngeal space and lung. As far as temporal region is concerned only a handful of studies have been published suggesting it to be not a very usual site of an ectopic meningiomas [6-8].

According to a recent paper, till date worldwide, 117 cases of primary ectopic meningiomas have been reported from the head and neck region, and 67 cases from other areas of body [5]. Case reports of the head and neck ectopic meningiomas have shown the temporal region involvement to be mostly intraosseous with only one case presenting as soft tissue mass at this site [Table/Fig-6] [3,4,6-12].

Meningiomas are usually clinically indolent in nature and morphologically heterogeneous tumours. The diagnosis of ectopic meningioma is difficult to make on the basis of imaging alone. On Magnetic Resonance Imaging (MRI), meningiomas usually have homogenous contrast enhancement and signal intensity similar to that of other brain lesions [1]. Hence, a definitive diagnosis depends on histopathological and immunohistochemical findings.

Several hypotheses have been proposed for their origin including extradural enclosing of arachnoid cell nests during embryogenesis, ectopic migration and the metaplastic development of arachnoid cells in combination with the peripheral nerves [3]. However, ectopic meningiomas have also been postulated to be mesenchymal tumours that arise from multipotential mesenchymal cells, particularly if no associations to the cranial nerves are apparent. In the head and neck region, this tumour entity is often associated with cranial nerves and, therefore, is considered to be derived from ectopic arachnoid tissue present around these nerves [5].

In 1960, Hoye SJ et al., proposed the classification of ectopic meningiomas into four types:

(1) Intracranial tumours with extracranial extension;
(2) Meningiomas originating in cranial nerve sheaths;
(3) Extracranial tumours without any connection to cranial nerve foramina;
(4) Intracranial benign lesions with extracranial metastases [13].

Ectopic meningiomas may exhibit variety of histological patterns similar to their intracranial counterparts. Though the presence of meningothelial cells is quite characteristic to clinch the diagnosis, it

Year of study	Author	Country	Number of cases	Age/sex	Intraosseous/extraosseous	Clinical presenation
1983	Granich MS et al., [9]	America	7	Mean age: 63.3 years, F=6; M=1	Intraosseous	Hearing loss and otorrhoea.
1996	Kuzeyli K et al., [10]	Turkey	1	6 years/M	Intraosseous	Slowly progressive temporal scalp mass.
1997	Muthukumar N [6]	India	1	15 years/F	Intraosseous	Headache and swelling in the right temporal region.
2001	Yadav YR et al., [7]	India	1	16 years/M	Intraosseous	Slowly progressive mass.
2009	Rushing EJ et al., [4]	America	38	Mean age: 50.1 years, F=25; M=13	Intraosseous	Sensorineural or conductive hearing loss, otitis, headaches, dizziness.
2012	Iaconetta G et al., [3]	Italy	1	75 years/F	Extraosseous	Temporal swelling with local pain and progressive proptosis.
2014	Sanei MH et al., [11]	Iran	1	49 years/F	Intraosseous	Pain and mixed unilateral hearing loss in the left ear.
2018	Ravikanth R et al., [8]	India	1	38 years/F	Intraosseous	Left-sided headache, facial pain, and left-sided cheek swelling.
2022	Singh J et al., [12]	America	1	42 years/F	Intraosseous	Right ear hearing loss.
2022	Present case	India	1	55 years/F	Extraosseuos	Soft tissue mass.

[Table/Fig-6]: Data of ectopic meningiomas of the temporal region reported till date worldwide [3,4,6-12].

is the variants like fibroblastic/angiomatous etc., that may not show the meningothelial cells conspicuously, and can create diagnostic confusion in an ectopic site with other soft tissue neoplasms, mostly peripheral nerve sheath tumours, myogenic tumours and vascular tumours. Perineuroma tumour cells and their architectural arrangement may show striking similarities to fibroblastic meningioma, including the formation of prominent (meningothelial-like) cellular whorls, lamellar fascicles and perivascular accentuation. Approximately, 10% of all perineuromas occur at head and neck sites, including subcutaneous tissue and the oral cavity. These cases are at higher risk of being misinterpreted as extracranial meningiomas, particularly if only EMA is used [14].

In the present case, the clinicoradiological findings suggested a benign soft tissue tumour, whereas on microscopy since the typical meningothelial cells were seen, authors did not keep any other mesenchymal tumour in the differential diagnosis. But because she was an elderly female, authors ruled out an amelanotic nodular malignant melanoma developing on a childhood nevi, since it can also present as a lump [15]. Immunohistochemistry (IHC) confirmed the diagnosis of an ectopic transitional meningioma.

The therapy of choice for these is surgical excision. Given the complexity of anatomy of the temporal bone complete en-bloc removal of tumour may sometimes be difficult. Role of radiotherapy and chemotherapy is not well-defined because of the limited number of case reports at this site [12].

Prognosis depends mainly on the grade of meningioma and the adequacy of resection. Grade I and grade II tumours in general carry a good prognosis whereas grade III tumours have adverse prognosis. Periodic follow-up and combined treatment with other medical specialties are important in preventing recurrence and finding other lesions [16].

CONCLUSION(S)

To summarise, ectopic meningiomas are extremely rare lesions encountered in the soft tissues/bones of head and neck region. It is the rarity and uncertainty of its aetiology that poses difficulty in its diagnosis. Radiological imaging is useful in preoperative diagnosis and surgical planning, but the ultimate diagnosis has to be confirmed by histopathological examination which can be rendered only if the pathologist is aware about its existence and does not misdiagnose it as a soft tissue neoplasm.

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