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Atypical Meningioma with Perineural Spread Along Hypoglossal Nerve

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A 50-year-old female presented with acute onset bilateral limb weakness, episodes of severe occipital headache with multiple episodes of loss of consciousness with a background history of left side neck pain, and occipital headache in the past 4 years. There was no significant past medical/surgical, social, or family history. On examination, there was left-sided deviation of the tongue with left-sided face weakness. Bilateral upper limb has power was 2/5 and lower limb power was 3/5 without any bowel and bladder incontinence.

On imaging, magnetic resonance imaging (MRI) revealed a large well-defined lobulated extra-axial left petroclival dura-based mass. The mass was seen extending along the prepontine, left cerebellopontine, and cerebellomedullary cisterns and inferiorly into the spinal canal through the foramen magnum (►**Fig. 1A**–**D**). The mass was isointense on T1-weighted (T1) imaging and on T2-weighted (T2) imaging. There was no significant diffusion restriction in diffusion-weighted imaging or blooming in gradient images. The tumor displayed intense, homogeneous post-contrast enhancement. The mass showed broad base along the tentorium cerebelli and clivus. Laterally the lesion was seen widening and eroding the hypoglossal canal and extending into the left parapharyngeal space. The enhancing component was also seen mildly protruding along the left internal acoustic meatus. Medially the lesion encased the terminal bilateral vertebral arteries and basilar artery with maintained flow voids. It caused mass effect with displacement of pons and medulla to the right and over the left cerebellar hemisphere and middle cerebellar peduncle (MCP). There was compression of outlet of fourth ventricle, with upstream tetraventricular hydrocephalus. Denervation atrophy of the left sided tongue muscles was noted (►**Fig. 1E** and **F**).

Computed tomography of the head and neck it showed internal soft calcification within the intracranial component of the mass. There was widening and erosion of the hypoglossal canal with hyperostosis of the surrounding bone (►**Fig. 1G**).

Tissue samples were obtained from the surgical specimen. This sample exhibited meningothelial tumor with syncytial and whorling architecture and dense intratumoral areas of hyalinization. There were numerous densely thickened hyalinized blood vessels admixed with areas of cellularity and foci of psammomatous type of dystrophic calcification were seen. No significant mitosis was identified. On immunohistochemistry, few tumor cells showed positivity for p16 protein; Monoclonal Antibody to Ki-67 (MIB-1) labelling index was approximately 1 to 2%. Based upon these features, a definitive diagnosis of transitional meningioma (histologically grade 1) was made (►**Fig. 2**).

The tumor was deemed too extensive for complete resection. The surgical team had performed retromastoid craniotomy to debulk the tumor and internal cerebrospinal fluid shunt diversion was done. Immediate postoperative scans revealed postoperative changes with residual tumor. At the last clinical review, the patient was 3 months' post-operation and had benefitted from alleviation of her headache and neck pain. All other clinical features were stable. Follow-up MRI scans have shown a stable appearance of the residual tumor.

Meningiomas are nonglial neoplasms arising from the meningothelial cells of the arachnoid layer. These tumors do not typically show perineural spread (PNS).[1–4] Review of the literature revealed few previous case reports of meningiomas spreading in this manner along trigeminal and facial

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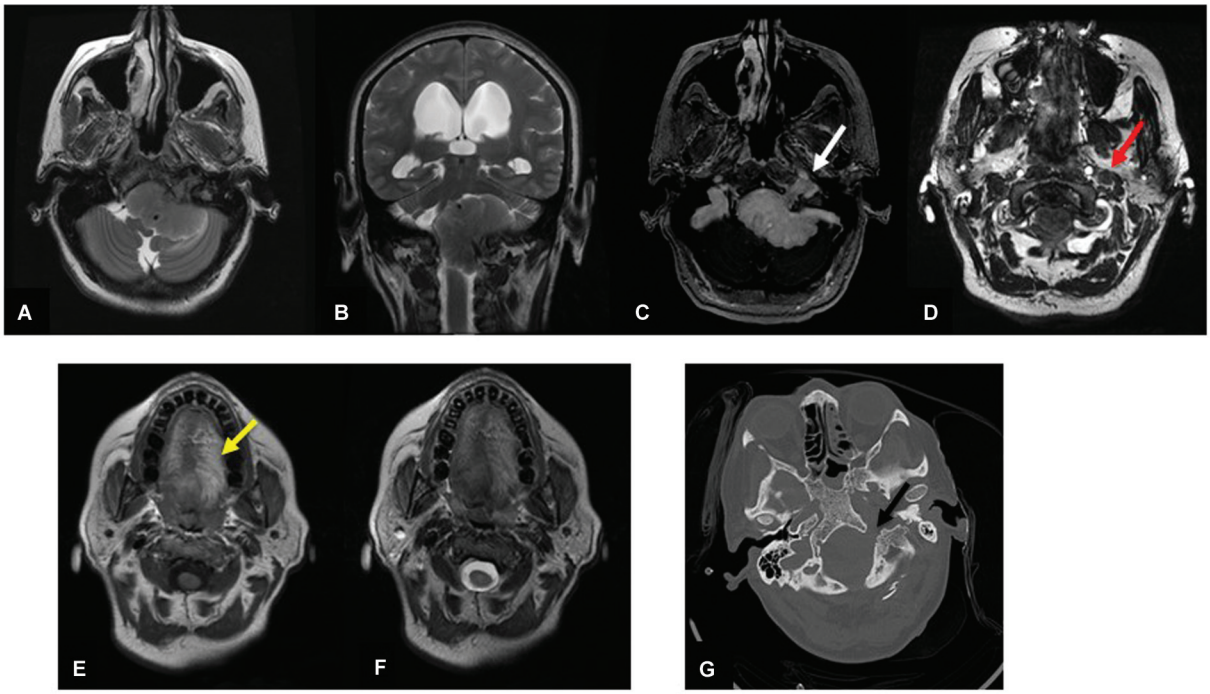

Fig. 1 A 50-year-old female with skull base meningioma with perineural spread. Findings: T2 axial and coronal (**A** and **B**), post-contrast T1 fat-saturated axial (**C**), and FIESTA axial sections (**D**) showing (**A**) broad-based extra-axial homogeneous left petroclival tumor with mass effect on adjacent structures. (**B**) Extension of the tumor into the spinal canal through the foramen magnum (**C**) homogenously enhancing tumor with thickening and enhancement of the left hypoglossal nerve with perineural enhancement (white arrow). (**D**) Thickened left hypoglossal nerve in the parapharyngeal space (red arrow). (**E** and **F**) T1 and T2 axial images demonstrating fatty atrophy and degeneration of the left half of the tongue (yellow arrow). (**G**) Axial bone window computed tomography section demonstrating erosion and enlargement of left hypoglossal canal (black arrow).

nerves. To our knowledge, this is the first reported case of a World Health Organization Grade I meningioma demonstrating perineural spread (PNS) along the hypoglossal nerve.

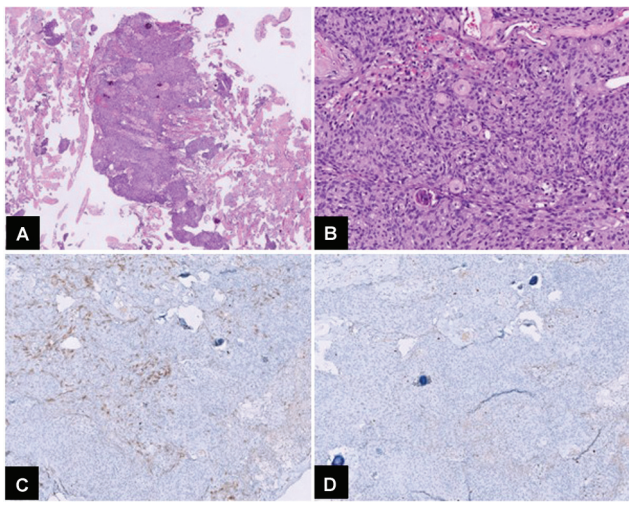

Fig. 2 (**A**) Hematoxylin and eosin (H and E) stain (5x) showing meningothelial tumor composed of cells in syncytial and whorling architecture along with fibrovascular tissue fragments with perivascular hyalinization. (**B**) H and E stain (20x) showing whorls of meningothelial cells with foci of psammomatous calcification. (**C**) P16 (10x) showing patchy and focal staining of p16 seen in the tumor cells. (**D**) MIB1 (10x) showing occasional tumor cell nuclei showing nuclear labeling for MIB1 (1–2%).

Atypical meningiomas represent 4.7 to 7.2% of all meningiomas occurring 10 years earlier on average and are more common in men.[5] PNS refers to involvement of larger, named nerves often accompanied by distant tumor spread along the nerves away from the primary site. Nemzek et al described that in case of PNS of a meningioma, the nerves are used as a scaffold to extend beyond the primary location, rather than any direct invasion of perineurium or endoneurium.[6,7] On imaging, PNS is evident and associated with clinical manifestations directly related to the involved nerve.[6–8] PNS has prognostic implication for clinical management and treatment planning. These patients are often treated with palliative radiation or with surgery performed on a case-by-case basis with the goal of tumor debulking to improve the efficacy of adjuvant therapy and to reduce symptoms.[9]

Differential diagnosis of atypical meningiomas with PNS includes ascending extra cranial tumors, descending intracranial tumors, and tumors arising from the cranial nerves. Nasopharyngeal squamous cell carcinoma and adenoid cystic carcinomas of the salivary gland show PNS with bony erosion and large necrotic nodal masses. On post-contrast sequences, these tumors show mild-to-moderate homogenous enhancement with PNS along the mandibular or maxillary division of trigeminal nerve.[10,11] Tumors arising from the cranial nerves like schwannoma and neurofibroma show smooth enlargement of the neural foramen with variable heterogeneous post-contrast enhancement. Descending intracranial tumors/inflammatory conditions like metastatic

disease and neurosarcoidosis show nodular enhancing deposits with thickening of the nerve.[12]

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Conflict of Interest
None declared.

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