Introduction

Tumors of the trochlear nerve are rare; even schwannomas, the most common pathology, comprise only ~30 cases in the literature.\(^1\)\(^2\) Other pathologies include meningioma especially of the cavernous sinus,\(^3\) hemangioblastoma,\(^4\) cavernous angioma,\(^5\) and neurofibroma.\(^6\) Here we present an isolated trochlear nerve meningioma demonstrating infiltration of the tumor into the nerve tissue, which represents the first case of its kind to be reported. A brief review of cases in which meningioma infiltrated cranial nerves and the implications for tumor grading are discussed.

Case Report

A 52-year-old male with no medical history presented with blurred and double vision. He first developed dizziness and blurred vision 10 years prior during a soccer game. From that point, he noticed mild double vision with physical exertion. As the diplopia progressed, he started wearing prism glasses 5 years ago but still had symptoms when fatigued. He underwent left inferior rectus recession surgery and had resolution for ~1 year, but his double vision eventually returned. At presentation, he wore nonprescription prism glasses that resolved the diplopia but required replacement with stronger prisms every year. On physical exam, he demonstrated a mild, intermittent head tilt to the right.

No obvious restriction of his extraocular movements was noted, but the patient reported worsening diplopia on right inferior gaze. Contrast-enhanced magnetic resonance imaging scan of the brain showed a 4 mm enhancing mass along the left ventrolateral pons in the area of the trochlear nerve, extra-axial but not dural-based.

The differential diagnosis included schwannoma, which was considered the likelier pathology, or meningioma, and the options of observation versus surgical removal were discussed. The progression of his diplopia suggested ongoing nerve damage caused either by compression or infiltration of the trochlear nerve. The options for observation or surgical intervention were discussed, and the patient elected to have surgery for diagnosis as well as surgical cure. The patient underwent a preauricular middle fossa craniotomy for resection of the mass. The trochlear nerve appeared to insert directly into the tumor, clinically more suggestive of a schwannoma. The proximal trochlear nerve was then sharply divided and the tumor was removed.

Pathological study demonstrated a meningiothelial meningioma (WHO Grade I) with no atypical histological features (~Fig. 1). The tumor was focally positive for EMA, but negative for progesterone receptors. The overall Ki-67 proliferation index was 3% to 4%. A neurofilament stain demonstrated invasion by the meningioma into a peripheral nerve segment (~Fig. 2).
Discussion

There are two prior cases in the literature of meningioma invading the trochlear nerve. The first case was described in a patient with von Recklinghausen’s disease, who had fusiform mass of the right trochlear nerve found incidentally during resection of a right cerebellopontine angle. The mass was resected and pathological study demonstrated a meningotheial meningioma with nerve fibers found in the periphery. The second case involved a cavernous sinus meningioma that had encased the right trochlear nerve. The nerve was resected, and pathological study demonstrated infiltration by a syncytial meningioma.

Conclusion

Nerve infiltration is not a criterion used in grading meningiomas, likely due to the rarity of cases and the lack of follow-up that would demonstrate that this feature is a risk factor for recurrence. Reports of nerve infiltration by meningioma include three cases of optic nerve invasion by optic nerve sheath tubular–diffuse type meningiomas; two cases of optic nerve invasion by intraorbital meningiomas, one transitional type found on autopsy and the other angiomatosus type that was disease-free 28 years after resection; trigeminal (V3) nerve invasion by a syncytial meningioma; and accessory nerve invasion by a meningotheial type meningioma. While one case series of jugular foramen meningiomas reports that these tumors commonly invade the lower cranial nerves, there is limited evidence of this behavior to be found in the literature.

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Fig. 1 Hematoxylin and eosin stain showing loose whorls and syncytia consistent with a meningotheial meningioma.

Fig. 2 Neurofilament immunostain showing invasion of tumor into peripheral nerve tissue.