Complete, Pupil-Sparing Third Nerve Palsy in a Patient With a Malignant Peripheral Nerve Sheath Tumor

Pupil-sparing but otherwise complete oculomotor nerve pareses (OMNPs) usually are caused by ischemia, whereas pupil-involved OMNPs or incomplete, pupil-sparing OMNPs usually require an evaluation for a compressive or infiltrative process. There are, however, exceptions. For example, Lustbader and Miller reported a case of a complete, pupil-sparing OMNP caused by a giant basilar tip aneurysm. Herein, we report a case of a complete pupil-sparing OMNP caused by a malignant peripheral nerve sheath tumor (MPNST) that arose in the sphenoid sinus.

Report of a Case. A 72-year-old man with controlled hypertension had a 5-week history of left ptosis and diplopia. The patient had normal visual acuity, color vision, and visual fields in each eye. The pupils were isocoric and equally and normally reactive to light and near stimulation. The right upper eyelid was in normal position and the right eye moved fully in all directions. The left upper eyelid was completely ptotic. The left eye had no elevation, depression, or adduction, but there was normal abduction and intorsion on attempted downgaze in abduction. The rest of the examination revealed no abnormalities. Magnetic resonance imaging revealed a hyperintense, heterogeneous, multilobulated mass centered in the sphenoid sinus on T2-weighted and fluid-attenuated inversion recovery images (Figure 1A), with heterogeneous enhancement and central nonenhancing parts with evidence of bilateral cavernous sinus invasion on contrast-enhanced T1-weighted images (Figure 1B). The patient underwent a gross total resection of the lesion, which was diagnosed as an intermediate-grade MPNST with the neoplastic cells staining strongly and diffusely for S-100 protein (Figure 2). No mutations were detected in the 4 KIT exons analyzed; ultrastructural analysis revealed no melanosomes.

Within a month after surgery, the patient's eyelid had returned to a normal position and his double vision had resolved. Examination at that time revealed normal visual sensory function, full extraocular movements, and no ocular misalignment in any field of gaze.

Figure 1. Magnetic resonance images. A, Coronal T2-weighted image showing a sphenoid sinus mass. B, Coronal contrast-enhanced T1-weighted image. R indicates right; L, left; H, head; and F, feet.

Figure 2. Immunohistochemical analysis of a malignant peripheral nerve sheath tumor. A, The lesional cells show both paucicellular and hypercellular zones and display a proclivity to proliferate in the subendothelial zones as seen in the vessel in the center of the field (hematoxylin-eosin, original magnification ×10). B, In this case, there is strong diffuse immunolabeling, an unusual feature for malignant peripheral nerve sheath tumor, which typically displays weaker, more focal S-100 protein expression. This finding prompted the consideration of both spindle cell melanoma and cellular schwannoma, both of which were excluded (S-100 protein immunohistochemistry, original magnification ×20).
Comment. Malignant peripheral nerve sheath tumors are rare tumors that originate from peripheral or cranial nerves or result from malignant transformation of a benign schwannoma or neurofibroma. They may sometimes be confused with amelanotic melanomas. In our case, the lack of KIT mutations and the absence of melanosomes in any of the cells were consistent with a diagnosis of MPNST rather than amelanotic melanoma.

Although MPNSTs can invade the paranasal sinuses, origin within the sinuses is extremely rare. These tumors almost never affect the cranial nerves, but when they do, the nerve most commonly affected is the facial nerve. We are unaware of any cases in which an OMNP was the manifesting sign. This case is even more unusual in that neither the size nor the reactivity of the pupil was affected.

What was the cause of third nerve palsy in our case? One possibility is that it was related not to the tumor but to the patient’s underlying vascular disease. This would be consistent with the painless onset of the palsy and its complete resolution after about 7 months, although this is somewhat longer than one would expect in such a setting. Although lesions of the sphenoid sinus do not commonly produce third nerve palsy, a few such examples have been reported. Thus, the mass could have affected the oculomotor nerve in the anterior cavernous sinus by compression, by producing an inflammatory reaction, or by causing local ischemia. Any of these mechanisms could account for the rapid resolution of the palsy following resection of the lesion. In any event, this case provides support for the recommendation that a patient with an isolated, complete, pupil-sparing OMNP be evaluated with neuroimaging when there is no evidence of improvement after several months.

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