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W C Shen, D Y Yang, W L Ho, Y J Ho and S K Lee

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Neurilemmoma of the Oculomotor Nerve Presenting as an Orbital Mass: MR Findings

Wu-Chung Shen,¹ Dar-Yu Yang,² William L. Ho,³ Yung-Jen Ho,¹ and San-Kan Lee¹

Summary: Neurilemmoma of the oculomotor nerve is rare and infrequently presents as an orbital mass. We report a neurilemmoma arising from the cavernous sinus with its major part extending into the orbit. The Antoni A part of the neurilemmoma showed intermediate signal intensity in both T1- and T2-weighted MR images and exhibited postcontrast enhancement. The Antoni B part revealed hypointensity on T1-weighted images, hyperintensity in T2-weighted images, and showed no contrast enhancement.

Index terms: Neuroma; Nerves, oculomotor (III); Orbits, neoplasms; Orbits, magnetic resonance

Although neurilemmoma (or schwannoma) arising from the oculomotor nerve is rare, oculomotor neurilemmoma arising from the cavernous sinus and extending into the orbit is even rarer (1–5). Herein we report the magnetic resonance (MR) findings of an oculomotor neurilemmoma involving the cavernous sinus, with the majority of the tumor extending into the orbital cavity via the dilated orbital fissure.

Case Report

A 43-year-old man complained of slowly progressive protrusion of the right globe with blurred vision for 7 years. Physical examination revealed right proptosis, ptosis of the right eyelid, poor visual acuity, and impairment of upward gaze. Computed tomographic (CT) scan disclosed a fusiform tumor mass, the majority of which was in the retrobulbar region of the right orbit, and the rest in the right cavernous sinus. There was widening of the right superior orbital fissure (Fig 1A). CT scan showed only faint contrast enhancement of the tumor compared with the enhancement of the normal cavernous sinus. MR imaging showed a tumor with the same shape as that found in the CT scan, and two distinct signal intensities. The central part revealed intermediate signal intensity on T1- and T2-weighted images, having signal characteristics similar to those of gray matter. The periphery of the tumor appeared as low signal intensity on the T1-weighted image (Fig 1B) and hyperin-

tensity on the T2-weighted image (Fig 1C). After intravenous administration of gadopentetate dimeglumine, the central part of the tumor showed clear, uniform contrast enhancement (Fig 1D), whereas the periphery of the tumor showed no enhancement.

Right-subfrontal craniotomy was performed, and after the cavernous sinus was opened, the surgeon could clearly see that the tumor was connected to the oculomotor nerve and extended into the orbital cavity via the widened right-superior orbital fissure. The orbital part of the tumor was easily removed as was the rest of the tumor in the cavernous sinus.

Grossly, the tumor was fusiform and well encapsulated. The cut surface was grayish, firm in the central portion, and soft in the periphery. There was no gross cystic change. Microscopy revealed neurilemmoma with a central area that was cellular, composed of spindle cells with palisading arrangements of nuclei consistent with Antoni type-A tissue. The peripheral area was less cellular, composed of loosely arranged cords of spindle cells separated by a myxoid matrix consistent with Antoni type-B tissue.

Discussion

Accounting for only 1% to 5.7% of all orbital tumors (3), neurilemmomas of the orbit are rare (1–3), as are neurilemmomas of the oculomotor nerve (3–5), most of which occur in the cavernous sinus. However, there are reports of the tumor arising from the cavernous sinus with orbital extension (1, 2).

In our case, the component of the tumor in the orbit was much larger than that in the cavernous sinus, so that before surgery we considered it as a retrobulbar orbital tumor with backward extension into the cavernous sinus via the superior orbital fissure. However, the surgeon clearly identified the tumor as being connected to the oculomotor nerve in the cavernous sinus with extension into the orbital cavity via the widened superior orbital fissure.

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¹Department of Radiology, ²Section of Neurosurgery, and ³Department of Pathology, Taichung Veterans General Hospital, Taichung, 407 Taiwan, Republic of China. Address reprint requests to Wu-Chung Shen.

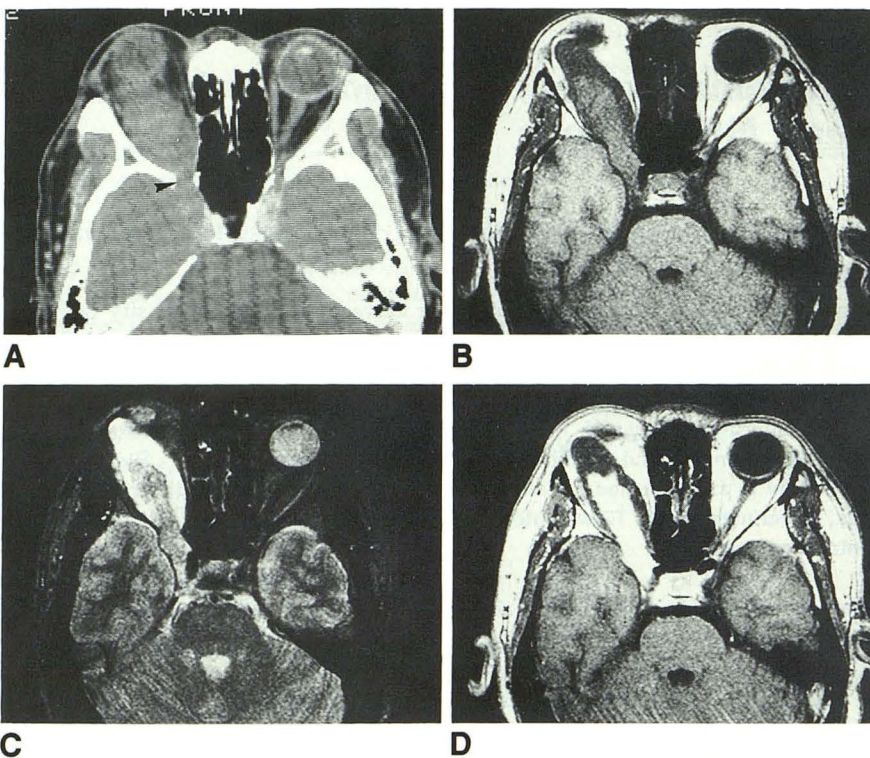
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Fig. 1. A, Axial CT of the right orbit with intravenous contrast enhancement shows a fusiform tumor mass—the majority of the tumor in the retrobulbar region of the orbit and the rest in the right cavernous sinus with widening of the right superior orbital fissure (arrowhead). Compared with the normal cavernous sinus, the tumor is poorly enhanced.

B, Axial MR, T1-weighted image (600/20/2) (repetition time/echo time/excitations). There are two kinds of signal intensities in the tumor mass—the central part shows intermediate signal intensity, reflecting soft-tissue entity, whereas the periphery of the tumor reveals hypointensity, suggesting sparse cellularity.

C, Axial T2-weighted image (2000/80). The central part of the lesion shows intermediate signal intensity. The periphery changes to a very high signal intensity.

D, Axial T1-weighted image (600/20) with intravenous injection of gadopentetate dimeglumine. The central part of the tumor shows homogeneous enhancement, whereas the periphery of the tumor is not enhanced.



It is generally recognized that the histologic pattern of neurilemmoma can be divided into two types. In the Antoni A type, the tumor cells form compact interlacing bundles of elongated bipolar spindle elements, and arrangement of the tumor cells in regular bundles often suggest a palisading effect. The Antoni B type of tissue is found in contiguity with the Antoni A type, and consists of a much looser pattern of more pleomorphic cells that lack the arrangement in bundles and palisades (6).

Neurilemmoma usually shows heterogeneous density on CT scan. The low-density part has been attributed to cystic change, Antoni type-B cells or Antoni type-A cells mixed with lipid-rich schwann cells (7, 8). The soft-tissue part presumably represents Antoni type-A cells. Earlier reports of oculomotor neurinoma (4, 9) show hyperintensity on T2-weighted images and intermediate intensity (9) or hyperintensity (4) on T1-weighted images. The periphery of the tumor, which showed slightly decreased density on CT scan, hypointensity on T1-weighted images, and hyperintensity on T2-weighted images, histologically showed an Antoni type-B area with sparse cellularity. The central part of the tumor, which appeared as a soft-tissue mass on both CT and

MR and showed contrast enhancement by gadopentetate dimeglumine, was histologically an Antoni type-A area with denser cellularity.

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