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Case Report

Intraocular Ciliary Nerve Schwannoma—Emphasis on Preoperative Diagnosis

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Abstract

Intraocular ciliary schwannoma is extremely rare and accurate diagnosis prior to surgery is difficult. In this article, we demonstrate a precise preoperative diagnosis of ciliary nerve schwannoma by meticulous neurological evaluation, combined with high-resolution magnetic resonance imaging with three-dimensional reconstruction of the lesion and adjacent structures. The tumor was identified to be arising from the ciliary nerve during operation and was fully excised. After 9 months of observation, the function of the third nerve recovered except for the ciliary branch. In conclusion, through high-resolution imaging and scrupulous cranial nerve examination, an accurate preoperative diagnosis of intraocular ciliary nerve schwannoma was achieved. This enabled the design of an appropriate surgical procedure and prediction of the surgical outcome. (*Tzu Chi Med J* 2009;21(3):265–268)

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1. Introduction

Oculomotor nerve schwannoma has been anatomically divided into three groups: cisternal, cistern cavernous, and cavernous (1). These aside, restricted intraorbital oculomotor schwannoma has been seldom reported (2). These infrequently located tumors could arise from any branch of the oculomotor nerve. However, prior to surgery, determining the tumor's origin is difficult, as is differentiation from more common intraorbital tumors arising from the optic nerve (3–5). In this article, we outline a case of schwannoma derived from the ciliary branch of the oculomotor nerve, which was accurately diagnosed preoperatively using careful neurological and ophthalmological evaluation results associated with comprehensive preoperative three-dimension image planning.

2. Case report

A 66-year-old woman became aware of blurred vision of her right eye for 6 months prior to her admission. Neurological examination showed no weakness, and no hypoesthesia or hyperalgesia of her limbs or trunk. Cranial nerve examination results were normal except for the nerves associated with vision. The ophthalmologic evaluation of her right eye revealed obvious exophthalmos, a slightly enlarged

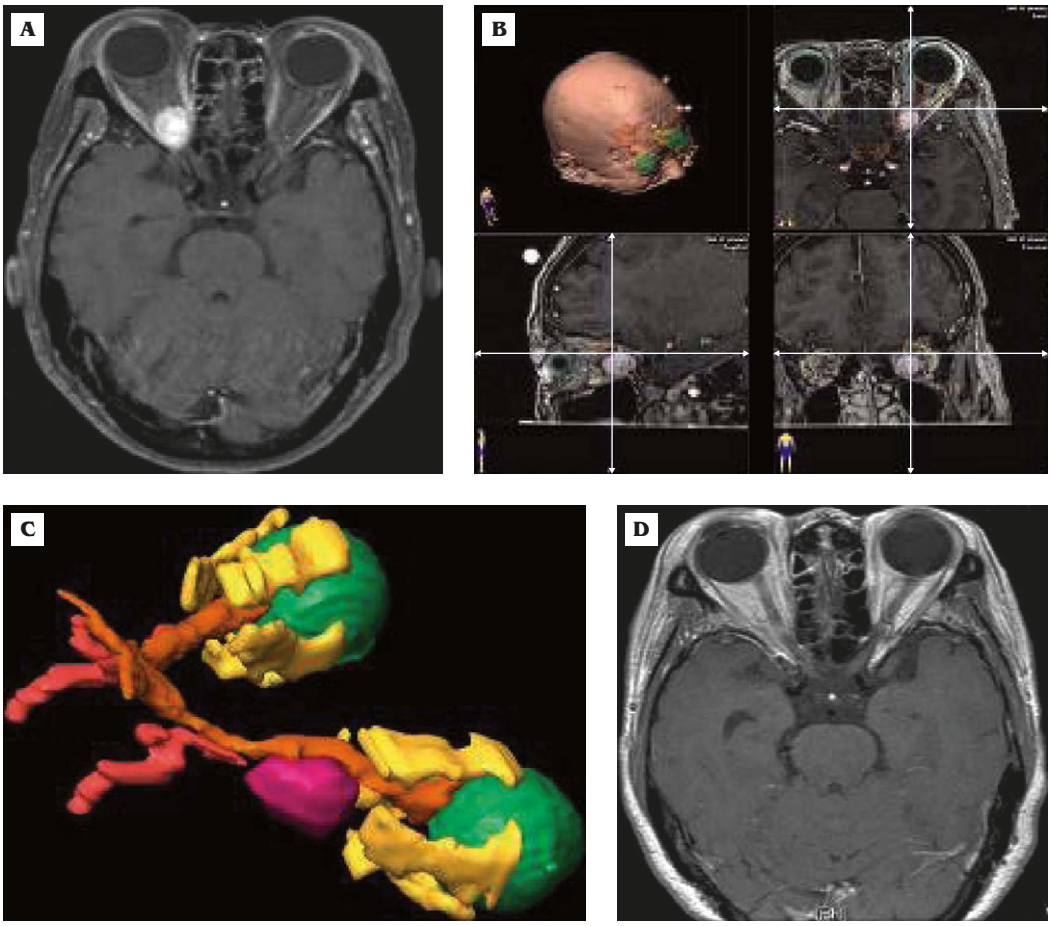

Fig. 1 — Preoperative magnetic resonance imaging (MRI) was transferred to the Brain LAB system for delineation of the tumor and adjacent structures. (A) T1-weighted MRI with contrast showed a well-enhanced round lesion in the right orbital apex. (B) Surgical planning of the intraorbital anatomic structures and tumor shown by reconstructive three-dimensional images (green: eyeball; yellow: intraocular muscles; orange: optic nerve and radiation; red: internal carotid and ophthalmic arteries; purple: tumor). (C) Three-dimensional imaging excluding non-delineated tissue showed the correlation of intraorbital structures and tumor. (D) T1 flair MRI with contrast showed total removal of the tumor, demarcation of the right optic nerve, and recovered anteroposterior diameter of the orbit 3 months after surgery.

pupil size, sluggish light reflex, positive indirect light reflex, only light perception of the right visual acuity, and no limitation of extraocular movement. Magnetic resonance imaging (MRI) revealed an intraorbital mass with low intensity on T1-weighted imaging, high intensity on T2-weighted imaging, and contrast-enhanced features (Fig. 1A).

Surgery was performed via the subfrontal approach, under computer-assisted stereotactic navigation. After unroofing the orbital cavity, an elastic and well-encapsulated mass was found deviating the optic nerve (superior-medially)—this was easily separated from the optic nerve and surrounding adipose tissue. Through the opening of the capsule, the tumor was removed piece by piece and total excision was achieved after resection of the two ends of the tumor (Figs. 1B–D).

Immediately after the operation, the patient developed third nerve palsy with proptosis, dilated pupil without light reflex, and lateral position of the right pupil. The pathological examination elicited schwannoma (Figs. 2A–C). Nine months after the operation, the neurological examination results exhibited resolution of the proptosis and malposition of the right pupil, complete recovery of extraocular movement, and normal right pupil size (which measured 3 mm). The direct light reflex of the right pupil still showed no response, but the indirect light reflex was detected (Fig. 3).

3. Discussion

Intraorbital ciliary schwannoma is a very rare orbital tumor entity—fewer than 20 cases have been reported

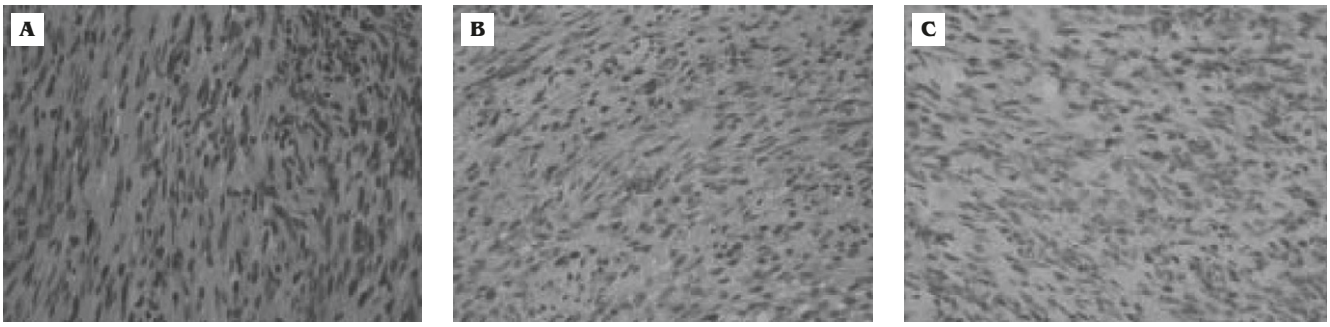

Fig. 2 — Pathological findings: (A) hematoxylin and eosin staining demonstrated wavy spindle cells; (B) S100 immunohistochemistry showed positive results; (C) glial fibrillary acidic protein staining showed negative results.

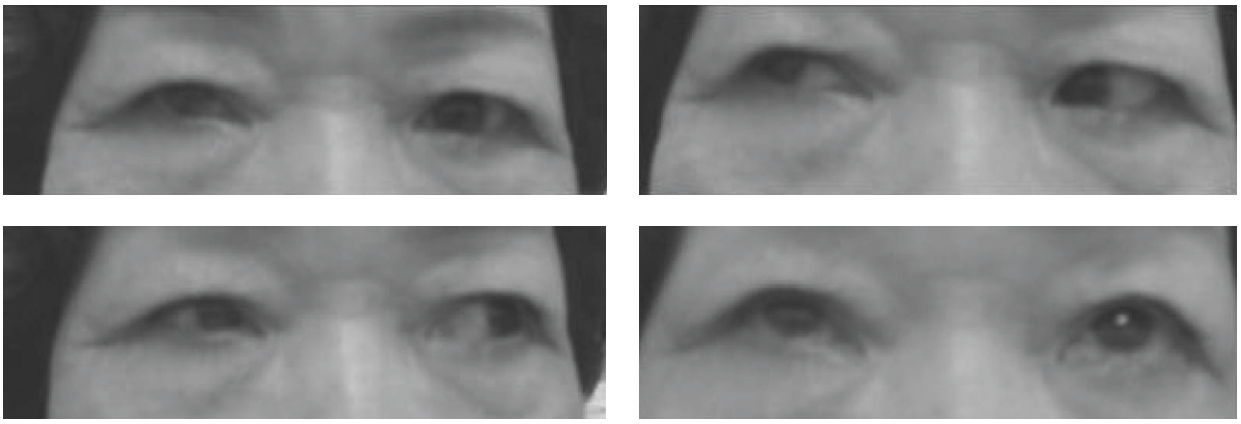

Fig. 3 — Photographs taken 9 months after surgery show no limitations in eye movement.

so far (6,7). In this case, the normal indirect right papillary light reflex disclosed intact function of the right oculomotor nerve, and normal indirect left papillary light reflex indicated a functional right optic nerve. However, sluggish direct right papillary light reflex and slight mydriasis of the right pupil suggested malfunction of the right optic nerve or intraorbital parasympathetic nerve. From the clinical presentation, a lesion involving the parasympathetic short ciliary and/or the optic nerve with partial compromise of function were interpreted. For further differentiation of the lesion origin, thin slice thickness MRI was used to delineate and reconstruct the optic nerve, lesion, and adjacent structures via colored and stereo images. In differentiating the origin of the tumor, the characteristic picture of an optic glioma or meningioma on MRI is expansion and enlargement of the optic nerve; in contrast, a tumor arising from branches of the oculomotor nerve displays a compressed and flattened optic nerve on MRI (8). In this case, the navigational imaging demonstrated that a tumor was located at the apex and compressed the optic nerve but did not enter or flatten any of the intraocular muscles (Figs. 1B and 1C). These findings—associated with neurological deficits—convinced us that the tumor originated from the short ciliary nerve with compression of the optic nerve.

Nine months after tumor resection, the intraocular movement recovered completely, but the direct light reflex was still absent and indirect light reflex was preserved to preoperative status. This indicated impairment of the ciliary nerve, but sparing of the third branches. These postoperative findings reconfirmed the preoperative diagnosis (Fig. 3).

In conclusion, by using high quality MRI imaging and meticulous neurological examination, more accurate preoperative diagnosis and more precise tumor excision can be achieved.

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