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Isolated Trochlear Palsy Due to Pilocytic Astrocytoma Involving the Pineal Gland

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Dear Editor,

Pilocytic astrocytoma mostly arises from the cerebellum,[1] and rarely involves the pineal region.[2] Isolated diplopia has rarely been reported in association with pilocytic astrocytoma, and isolated trochlear palsy has only been described once, in a child with pilocytic astrocytoma involving the posterior fossa.[3] The purpose of this report is to describe a rare manifestation of isolated unilateral trochlear palsy due to pilocytic astrocytoma originating from the pineal gland and symmetrically involving the tectal area.

A 22-year-old man presented progressive diplopia with a 4-year history. He denied any history of significant vomiting, headache, weight loss, or head trauma. A neurologic examination revealed that his head was tilted to the right, while his visual acuity, visual fields, and pupils were normal. His eyelid function was also normal, with no ptosis or retraction. At the primary position, he had a left hypertropia of 8 prism diopters (PD) that increased during rightward (20 PD) and downward (20 PD) gaze and during leftward head tilt (15 PD) (Fig. 1A). The smooth pursuit and saccades were normal in both the horizontal and vertical directions, and convergence was intact. The findings of a funduscopic examination were normal, with no abnormal ocular torsion. The subjective visual vertical was within the normal range in both eyes. A review of photographs of the patient during childhood did not reveal any compensated head tilt. He was diagnosed as isolated left trochlear nerve palsy of acquired origin.

All findings of routine blood tests were normal or negative. Magnetic resonance imaging (MRI) revealed a lobulated mass of 1.8×4 cm with gadolinium enhancement in the area of the pineal gland. The lesion was symmetric around the midline and extended into the tectal area (Fig. 1B and C). Craniotomy and removal of the tumor disclosed a pilocytic astrocytoma that probably originated from the pineal gland. The diplopia remained unchanged after the operation.

Pineal tumors comprise 0.4−1.0% of all intracranial tumors and can be classified into three major types: germ cell tumors, pineal cell tumors, and glial tumors.[4] Germ cell and pineal cell tumors comprise 78−86% of all pineal tumors, while gliomas account for only 14–22%.[4] A study of gliomas involving the pineal region found pilocytic astrocytoma in only 49 (14%) out of 349 patients with a histologic confirmation.[4] The present patient was also initially diagnosed with a germ cell tumor based on the MRI findings, because most pineal tumors are germ cell tumors at his age. However, a histopathologic study disclosed a pilocytic astrocytoma.

The symptoms of pineal tumors are mostly due to compression of the surrounding structures and resultant hydrocephalus.[5] About 90% of pineal tumors therefore present with symptoms of increased intracranial pressure, and approximately 50% show pretectal syndrome.[5] In contrast, only 11% of gliomas involving the pineal region present with diplopia.[4] The present patient is unique in that his pineal glioma manifested with an isolated unilateral

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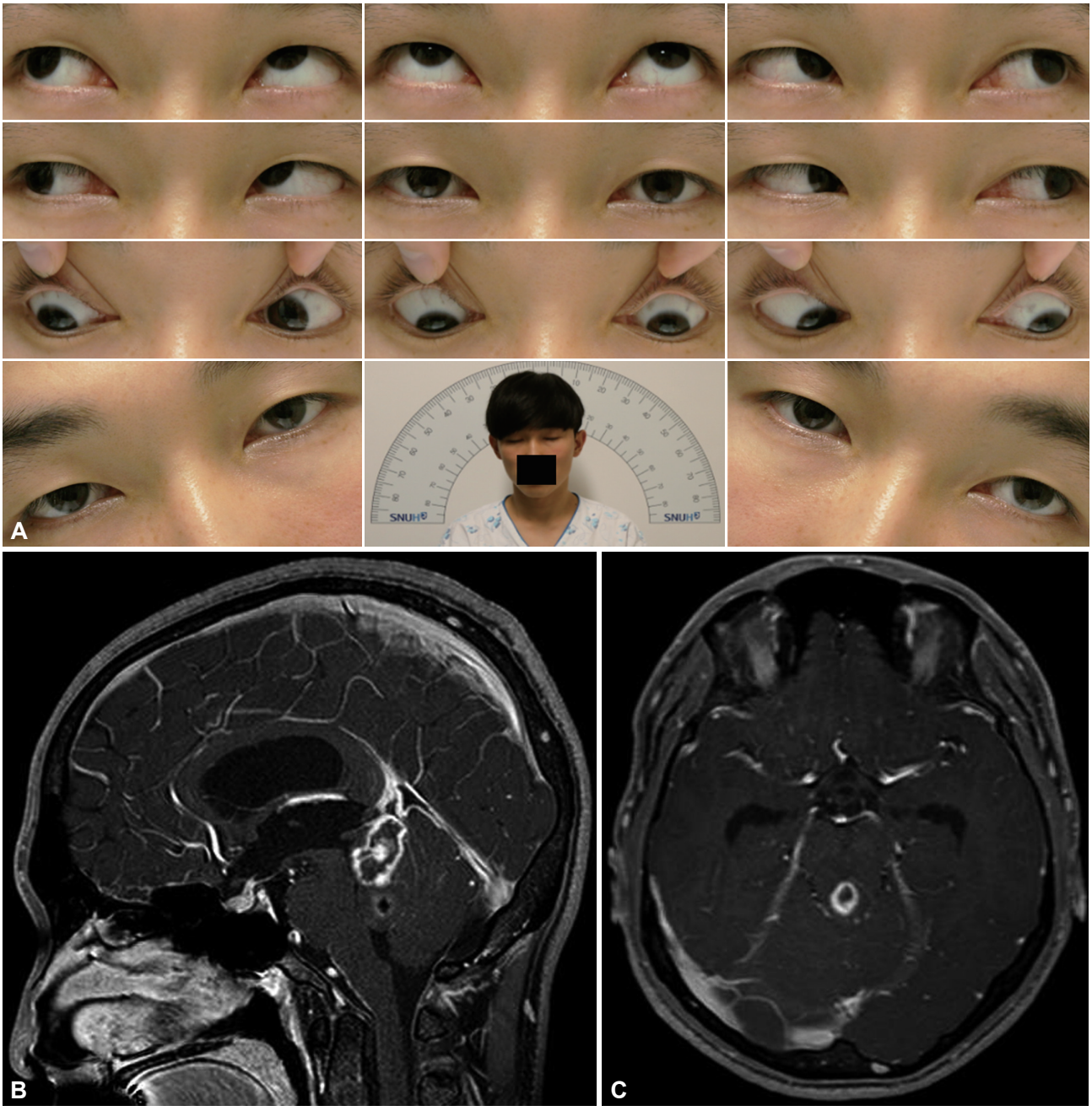

Fig. 1. Eye movements, head posture, and MRIs of the patient. A: Gaze photographs showing left hypertropia that increases during rightward and downward gazes, and during leftward head tilt. B: Gadolinium-enhanced sagittal MRI showing a well-circumscribed pineal tumor extending from the pineal region. C: Gadolinium-enhanced axial MRI shows compression of the tectum in the midbrain.

trochlear nerve palsy. The mechanism appears to be compression of the trochlear fascicle in the brainstem or the trochlear nerve in the subarachnoid space. The normal ocular torsion and subjective visual vertical also indicate the chronicity of the underlying lesion. Pineal glioma should be considered a rare cause of isolated trochlear nerve palsy, especially in younger patients.

Conflicts of Interest

The authors have no financial conflicts of interest.

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