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Tectal Plate Glioma with Optic Nerve Glioma in Neurofibromatosis Type 1: A Case Report

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ABSTRACT

Tectal plate gliomas are uncommon low-grade brain tumors that typically present with obstructive hydrocephalus, while optic pathway gliomas are among the most frequent central nervous system tumors associated with neurofibromatosis type 1 (NF1). We describe an 11-year-old boy who presented with progressive macrocephaly. Clinical examination revealed multiple café-au-lait macules, decreased visual acuity in the right eye, and a family history of NF1. Brain magnetic resonance imaging (MRI) demonstrated a well-circumscribed 5 × 6 × 6 mm tectal plate lesion that was hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR sequences, without diffusion restriction or post-contrast enhancement, causing aqueductal obstruction and triventricular hydrocephalus without transependymal cerebrospinal fluid permeation. MRI also showed fusiform enlargement of the intraorbital segment of the right optic nerve with T2/FLAIR hyperintensity and no contrast enhancement. These imaging findings were highly suggestive of a tectal plate glioma associated with a right optic nerve glioma in the setting of NF1. This case highlights the essential role of MRI in identifying the cause of obstructive hydrocephalus, detecting associated NF1-related intracranial tumors, and guiding appropriate clinical management and imaging surveillance.

KEYWORDS

Neurofibromatosis type 1; Tectal glioma; Optic nerve glioma; Obstructive hydrocephalus; Magnetic resonance imaging

MAIN ARTICLE

INTRODUCTION

Neurofibromatosis type 1 (NF1) is a common autosomal dominant tumor-predisposition syndrome characterized by pigmentary abnormalities, neurocutaneous manifestations, and an increased risk of central nervous system tumors [1].

Optic pathway glioma is the most frequent central nervous system tumor associated with NF1 and occurs in approximately 15%–20% of affected children [2].

Tectal plate gliomas are uncommon pediatric low-grade gliomas that typically present with obstructive hydrocephalus due to aqueductal compression [3].

The coexistence of a tectal plate glioma and an optic pathway glioma in a child with clinical features of NF1 is rare and highlights the importance of MRI in identifying the underlying cause of hydrocephalus and associated NF1-related intracranial lesions [3,4].

We report the case of an 11-year-old boy with suspected NF1 presenting with progressive macrocephaly, in whom MRI demonstrated a small tectal plate glioma responsible for triventricular hydrocephalus and an associated right optic nerve glioma [1–4].

CASE PRESENTATION

An 11-year-old boy was referred to our institution for evaluation of progressive macrocephaly that had gradually increased in size over several years. His family history was notable for a sibling diagnosed with neurofibromatosis type 1 (NF1).

Physical examination revealed marked macrocephaly and multiple café-au-lait macules, raising the suspicion of NF1.

An earlier non-contrast brain computed tomography (CT) scan had demonstrated hydrocephalus without an identifiable underlying cause. Brain magnetic resonance imaging (MRI) was subsequently performed for further evaluation. MRI revealed a well-circumscribed 5 × 6 × 6 mm lesion centered in the tectal plate, adjacent to the cerebral aqueduct. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR sequences, without diffusion restriction or contrast enhancement following gadolinium administration. It caused obstruction of the cerebral aqueduct, resulting in triventricular hydrocephalus without imaging evidence of transependymal cerebrospinal fluid permeation.

MRI also demonstrated fusiform enlargement of the intraorbital segment of the right optic nerve, which appeared hyperintense on T2-weighted and FLAIR images without post-

contrast enhancement. These imaging findings were highly suggestive of a tectal plate glioma associated with a right optic nerve glioma in the setting of neurofibromatosis type 1.

DISCUSSION

NF1 is diagnosed on the basis of clinical and/or genetic criteria, including café-au-lait macules, axillary or inguinal freckling, neurofibromas, optic pathway glioma, characteristic osseous lesions, or an affected first-degree relative [1].

In our patient, the presence of multiple café-au-lait macules, decreased visual acuity, and a family history of NF1 supported a clinical suspicion of NF1 [1].

Children with NF1 are predisposed to low-grade gliomas, particularly optic pathway gliomas, but non-optic gliomas may also occur in the brainstem and other intracranial locations [2,5].

Optic pathway gliomas in NF1 usually arise during childhood and may involve the optic nerves, optic chiasm, optic tracts, hypothalamus, or optic radiations [2].

They are often indolent, but visual decline may occur, especially in young children and in tumors involving the chiasm or posterior optic pathways [2,6].

MRI is the imaging modality of choice because it allows accurate assessment of optic nerve enlargement, signal abnormalities, contrast enhancement, and extension along the visual pathway [2,6].

In this case, fusiform enlargement and T2/FLAIR hyperintensity of the intraorbital segment of the right optic nerve, without enhancement, were highly suggestive of an NF1-associated optic nerve glioma [2,6].

Tectal plate gliomas are usually low-grade astrocytic tumors centered in the tectum and are classically associated with aqueductal obstruction and triventricular hydrocephalus [3,4].

On MRI, they typically appear as well-defined tectal lesions with T2 hyperintensity, variable T1 signal, and absent or mild enhancement [3].

Our patient's tectal lesion showed T1 hypointensity, T2/FLAIR hyperintensity, no diffusion restriction, and no post-gadolinium enhancement, which is consistent with the usual imaging profile of a low-grade tectal glioma [3,4].

The associated triventricular hydrocephalus without transependymal cerebrospinal fluid permeation suggested chronic aqueductal obstruction without acute decompensation [3,4].

The main role of imaging in such cases is to identify the obstructive lesion, assess the severity of hydrocephalus, search for additional NF1-related lesions, and provide a baseline for surveillance [3,4].

Management of tectal plate gliomas is generally conservative, with treatment directed

primarily toward cerebrospinal fluid diversion when hydrocephalus is symptomatic or progressive [3,4].

Endoscopic third ventriculostomy is commonly used for obstructive hydrocephalus related to tectal plate gliomas and may avoid the need for tumor resection in many patients [4,7].

Biopsy or resection is usually reserved for atypical imaging features, radiological progression, contrast enhancement, or clinical deterioration [3,4].

The coexistence of optic pathway glioma and tectal plate glioma in a child with NF1 emphasizes the need for careful ophthalmologic evaluation and long-term MRI follow-up [2,6].

Treatment of NF1-associated optic pathway glioma is generally considered when there is progressive visual impairment, radiological progression, or clinically significant mass effect [2,6].

Chemotherapy has traditionally been used as first-line therapy for progressive pediatric low-grade optic pathway gliomas, while radiotherapy is generally avoided in young NF1 patients because of the risk of long-term complications and secondary tumors [2,6].

Therefore, in this case, MRI was essential not only for explaining the hydrocephalus but also for detecting a second clinically relevant NF1-associated tumor involving the right optic nerve [2–4].

CONCLUSION

This case highlights the central role of MRI in identifying the cause of obstructive hydrocephalus and detecting associated intracranial manifestations of neurofibromatosis type 1. Although uncommon, the coexistence of a tectal plate glioma and an optic nerve glioma should be considered in children with clinical features of NF1. Early recognition of this association is essential for appropriate management, ophthalmologic assessment, and long-term imaging surveillance.

FIGURES :

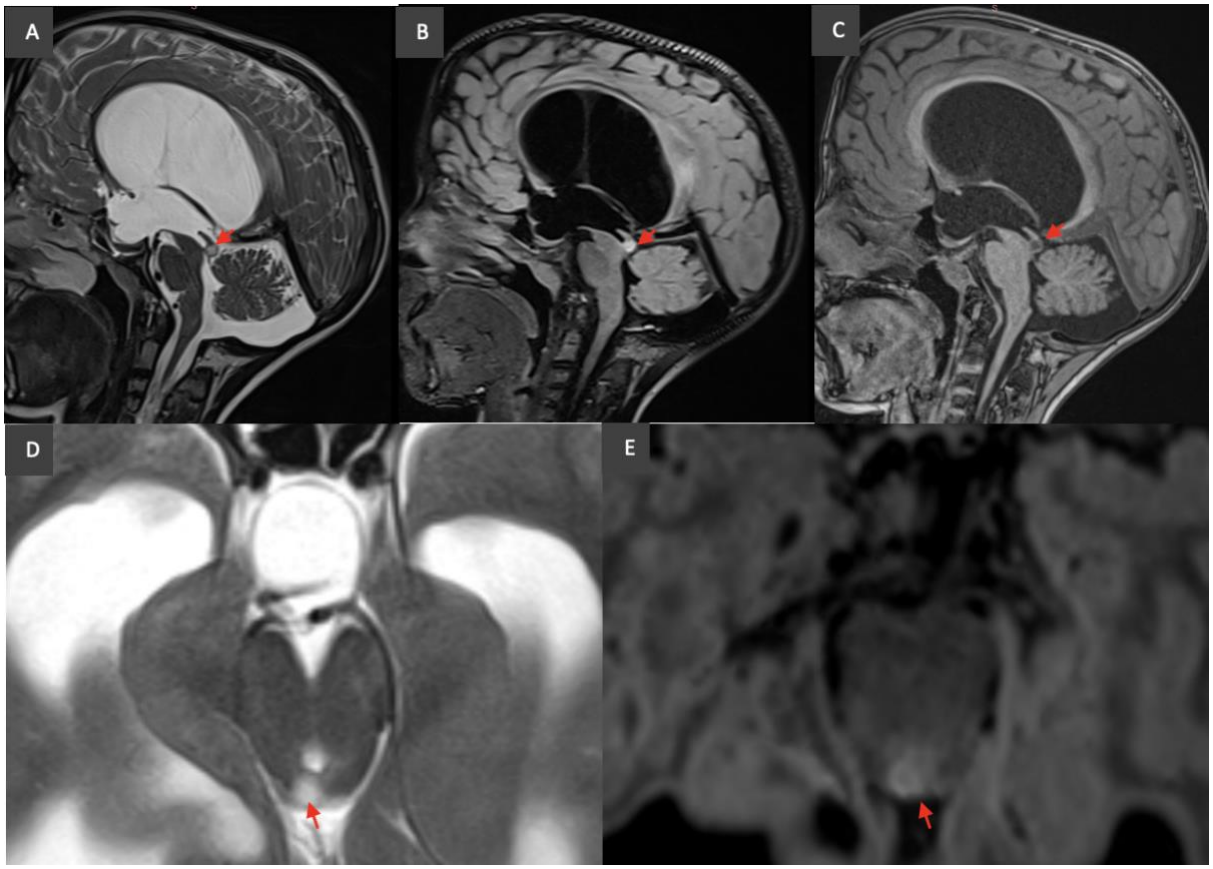


Figure 1: Brain MRI demonstrating a well-circumscribed tectal plate lesion (white arrow). (A) Sagittal T2-weighted image, (B) sagittal FLAIR image, and (C) sagittal T1-weighted image show a small tectal plate mass, hypointense on T1-weighted imaging and hyperintense on T2-weighted and FLAIR sequences, causing compression of the cerebral aqueduct with associated triventricular hydrocephalus. (D) Axial T2-weighted image and (E) axial FLAIR image confirm the hyperintense tectal lesion (white arrow)

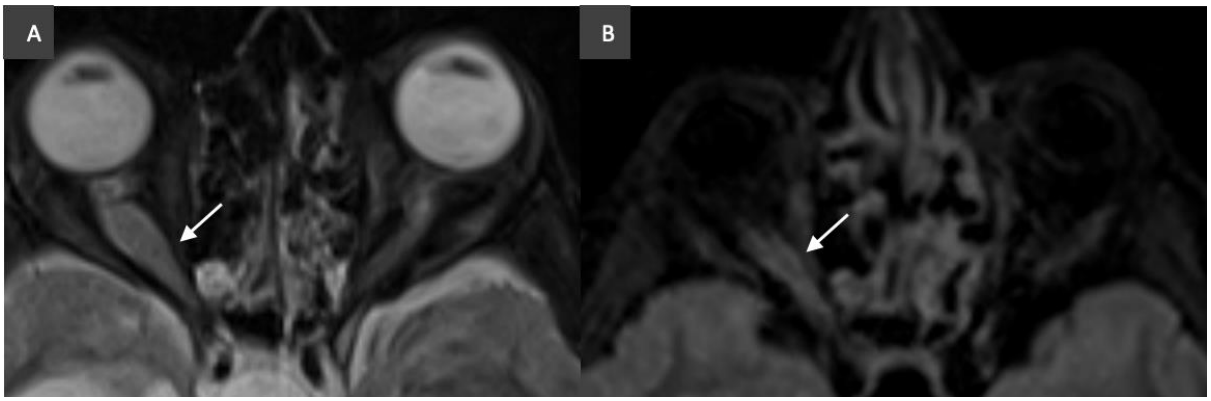


Figure 2: (A) Axial fat-suppressed T2-weighted image and (B) axial FLAIR image demonstrate fusiform enlargement of the intraorbital segment of the right optic nerve (white arrow), which appears hyperintense relative to the contralateral side.

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