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Concomitant Meningioma and Glioma Within the Same Optic Nerve in Neurofibromatosis Type 1

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Abstract

A patient with neurofibromatosis type 1 and the rare finding of concomitant meningioma and optic pathway glioma within the same optic nerve is presented. A 4-year-old boy was admitted to our hospital with right-sided proptosis. He also had numerous café-au-lait macules and axillary freckling on physical exam. According to National Institutes of Health (NIH) criteria, he met the diagnostic criteria for neurofibromatosis type 1. On magnetic resonance imaging (MRI), a mass originating from the right optic nerve sheath with normal appearance of the optic nerve was observed, which was consistent with optic nerve sheath meningioma. Another mass lesion was observed in the prechiasmatic region of the same optic nerve, which was consistent with optic nerve glioma. Two different types of optic pathway tumors in the same optic nerve is an extraordinary case. It is important to recognize imaging findings of these tumors and make correct diagnosis.

Keywords

neurofibromatosis type 1, optic nerve sheath meningioma, glioma, magnetic resonance imaging (MRI)

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Optic nerve tumors are rare and they can be difficult to diagnose. More than 90% of primary optic nerve tumors are either benign gliomas of childhood or optic nerve sheath meningiomas.[1] The percentage of optic glioma in patients with neurofibromatosis type 1 is unclear (ranges from 10% to 70%).[2,3] Fifteen percent of neurofibromatosis type 1 patients are reported to have optic pathway gliomas.[4] Meningiomas constitute 0.4% to 4.6% of the pediatric central nervous system tumors.[5,6] The incidence of meningiomas seems to be same in the general population and in neurofibromatosis type 1 patients.[7]

We present an extraordinary neurofibromatosis type 1 case with these 2 different tumors in the same optic nerve. Only a few cases of concomitant meningioma and glioma have been reported, but there is no reported case of neurofibromatosis type 1 with concomitant optic nerve glioma and optic nerve sheath meningioma within the same optic nerve in the current literature. Distinction of these 2 tumors is important because of the different treatment modalities.

Methods

An orbit magnetic resonance imaging (MRI) was performed before and after contrast administration, using a Philips Achieva 3-T MR scanner. For enhanced imaging, we used 0.1 mmol/kg gadolinium diethyltriaminepentaacetic acid (Gd-DTPA).

We also performed physical examination and ophthalmic examinations including visual acuity determination, slit-lamp biomicroscopy, and fundus examination.

Case Summary

A 4-year-old boy was admitted to our hospital with a history of gradual right-sided proptosis. MRI showed a mass in the right retrobulbar optic nerve that was consistent with meningioma and concomitant another mass in the prechiasmatic region of the same optic nerve that was consistent with optic glioma. We did not perform biopsy because of risk of vision loss. The patient was diagnosed with neurofibromatosis type 1 according to National Institutes of Health (NIH) criteria. Our patient fulfilled 3 criteria, including 6 or more café-au-lait spots

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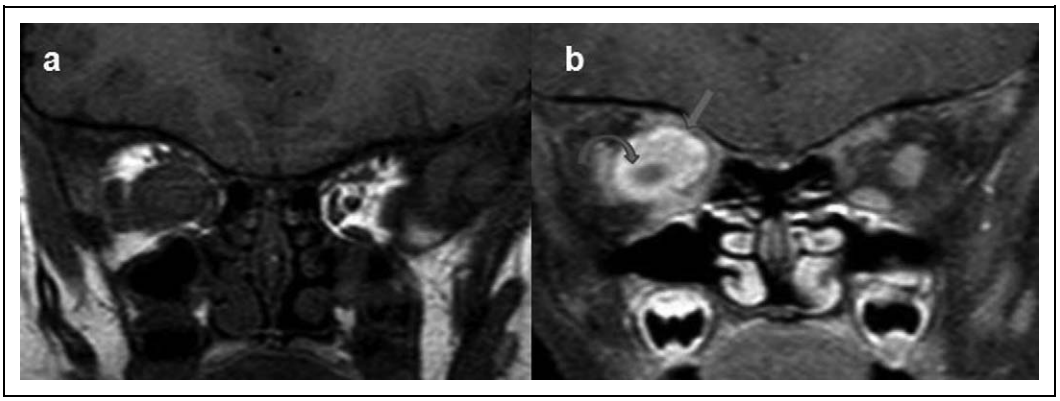

Figure 1. Magnetic resonance imaging (MRI) of a mass lesion originating from the right optic nerve sheath is seen. (A) The lesion is hypointense on T1-weighted images. (B) Following contrast administration, unenhanced optic nerve (curved arrow) can be clearly distinguished from the strongly enhanced mass lesion (straight arrow).

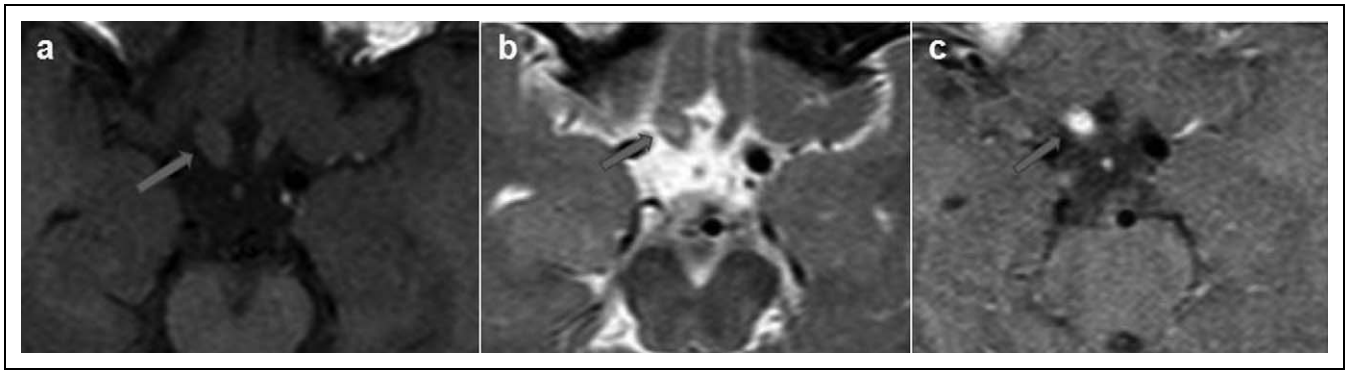

Figure 2. (A) T1-weighted magnetic resonance imaging (MRI) shows a hypointense mass lesion in the region of the right prechiasmatic optic nerve. (B) The lesion is slightly hyperintense on T2-weighted images. (C) Note that different from the first, this lesion demonstrates global enhancement following contrast administration.

>0.5 cm in diameter in prepubertal children, axillary freckling, and optic pathway glioma. We decided to observe for meningioma and treat optic glioma because of the patient's age. Vincristine in combination with carboplatin was used in the treatment of optic glioma. We performed MRI and ophthalmic examination at the 4-month follow-up.

Results

His physical examination revealed café-au-lait macules over the whole body, axillary freckling, mild cognitive impairment, and right-sided proptosis. His father also has had cutaneous neurofibromatosis. Ophthalmic examination revealed no abnormality. No lisch nodule was observed and the optic disc was normal. MRI revealed a mass originating from the right optic nerve sheath in the retrobulbar optic nerve with a low signal intensity on T1-weighted images and a high signal intensity on T2-weighted images (Figure 1a). MRI revealed another mass in the prechiasmatic region of the right optic nerve with a low signal intensity on T1-weighted images and a slightly high signal intensity on T2-weighted images (Figure 2A and B). Contrast-enhanced MRI showed significant enhancement of the retrobulbar mass with normal appearance of the optic nerve centrally (Figure 1B). In addition, contrast-enhanced MRI revealed total enhancement of the mass in the region of the prechiasmatic optic nerve without normal appearance of the optic nerve (Figure 2c). At the 4-month follow-up, the tumors had shown no radiologic progression. There was no abnormality on ophthalmic examinations, and the patient was stable.

Discussion

Neurofibromatosis type 1 is an inherited tumor predisposition syndrome. Diagnosis can be made using the National Institutes of Health[8] diagnostic criteria. Patients are diagnosed with neurofibromatosis type 1 who met 2 or more criteria. These criteria are as follows: (1) 6 or more café-au-lait spots (>0.5 cm diameter in prepubertal children, >1.5 cm after puberty); (2) axillary or inguinal freckling; (3) cutaneous neurofibromas or 1 plexiform neurofibroma; (4) 2 or more iris Lisch nodules; (5) optic pathway gliomas; (6) specific bony lesions (sphenoid wing dysplasia, pseudoarthrosis of the tibia); and (7) a first-degree relative with neurofibromatosis type 1.

MRI is an excellent method of choice in the follow-up of patients with neurofibromatosis, as well as detecting lesions in asymptomatic patients.[9] These patients are prone to

developing both peripheral and central nervous system tumors. Gliomas are the most common primary central nervous system tumors that especially seen in the optic pathways in children.[10] Although the neurofibromatosis type 1–associated optic pathway gliomas exhibit indolent behavior, approximately from one-third to half of these tumors cause clinical symptoms.[4] The diagnosis of an optic nerve glioma and optic nerve sheath meningioma can be confirmed by computed tomography (CT) or MRI.[11] Optic nerve biopsy has no influence on therapeutic decisions. In addition, biopsy may cause severe visual loss.[1,11] The appearance of optic nerve glioma depends on whether or not the patient has neurofibromatosis type 1. There is almost always a fusiform enlargement of the optic nerve with a clearcut margin produced by the intact dural sheath in patients without neurofibromatosis type 1, whereas the nerve is more irregular and tends to show both kinking and buckling in neurofibromatosis type 1 patients.[12] On MRI, gliomas usually appear as hypo- to isointense on T1-weighted images and mildly to strongly hyperintense on T2-weighted images. After contrast administration, gliomas show total or partial enhancement. MRI can also show extension of tumor and tumor-associated changes.[11]

The treatment options for optic nerve glioma include surgery, chemotherapy, and radiotherapy.[1] The effective treatment modality is chemotherapy for unresectable gliomas, which delays growth of tumor. It also delays the use of radiotherapy and thus prevents the undesirable side effects of irradiation,[13] including secondary malignancy, hearing loss, visual loss, radiation-induced optic neuropathy-retinopathy, and moyamoya syndrome.[1]

Optic nerve sheath meningiomas can be categorized into 2 groups as (1) primary meningiomas that arise from the cap cells of the arachnoid of the optic nerve and (2) secondary meningiomas that arise intracranially and invade the optic canal and orbit by extending between the dura and arachnoid of the optic nerve.[11] Diagnosis can be made by a combination of clinical history and imaging findings.[1] Patients with optic nerve sheath meningiomas may complain of decreased or blurred vision or transient visual loss lasting for a few seconds or visual loss occurring only when moving the eye into a particular field of gaze. However ophthalmic examination may reveal normal vision[11] as in our case. Proptosis may rarely be an initial symptom and usually occurs after visual loss.[11] Our patient had proptosis but visual acuity was normal. The characteristic computed tomographic (CT) features of optic nerve sheath meningiomas include the presence of calcification surrounding the nerve (which occurs in 20%-50%), tubular enlargement of the optic nerve, and a bulbous enlargement of the optic nerve at the apex with distal tubular enlargement. The appearance of the optic nerve on coronal section is a hypodense area surrounded by a dense peripheral ring. Marked, homogenous enhancement with gadolinium is also typical. MRI findings are similar to CT findings.[11] Optic nerve sheath meningiomas are not life threatening, are unlikely to spread intracranially and be compatible with good vision; thus the treatment has altered from surgery to conservative approach. In some instances, the appropriate treatment is radiotherapy.[11,14] In addition, optic nerve sheath meningiomas cannot be separated from the optic nerve completely because, in most cases, the tumor completely surrounds and occasionally invades the optic nerve.[11,15] More recently, it has become clear that surgery is usually unsuccessful to completely remove an optic nerve sheath meningioma, and radiation is the optimum therapy.[14] But it must be kept in mind that radiotherapy increases the risk of secondary malignancy, especially in patients with neurofibromatosis type 1.[1] Roser et al[15] recommend surgery only in case of intracranial extension and rapid deterioration.

Miller[11] recommends follow-up without intervention in patients with optic nerve sheath meningioma who have good visual function, as in our case. Miller[11] also reported that the management of such patients should be regular clinical examinations every 3 to 6 months and repetition of MRI examination every 6 to 12 months.

As a result, although the appearance rarely varies, optic nerve gliomas may be mistaken for an optic nerve sheath meningioma.[16] In most of the cases, distinction can be made by combining clinical history and imaging findings. This distinction is important because the treatment differs between these 2 tumors.

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This child was managed by the pediatric oncology of the department of pediatrics at the Kocaeli University school of medicine, Kocaeli, Turkey.

Author Contributions

SB and AA drafted the manuscript. MK and FÇ contributed to the study design, acquisition and analysis of data, and clinical evaluation. AA and YA performed radiologic evaluation and approved the final draft.

Declaration of Conflicting Interests

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Ethical Approval

Ethical approval was not necessary because this study did not involve research on human subjects and there were no patient identifiers.

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