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Case Report

Large rhabdoid meningioma presenting prominent hyperintensity in the optic nerve: An indicator of visual disturbance on constructive interference steady-state sequence?

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ABSTRACT

Background: Rhabdoid meningiomas (RMs) are a rare type of malignant meningioma. Here, we report a case of intracranial RM presenting with visual disturbance and prominent hyperintensity in the optic nerve (ON).

Case Description: A 20-year-old female presented with a 1-year history of headache. At presentation, her visual acuity (VA) was 20/50 on the right side and 20/40 on the left, with an intraocular pressure of 17 mmHg on both sides. Cerebral magnetic resonance imaging revealed a broad-based tumor in the right frontal convexity. It measured 82 mm × 65 mm × 70 mm in diameter, accompanied by cystic components, and was inhomogeneously enhanced. The intraorbital ONs demonstrated prominent intramedullary hyperintensity on the constructive interference steady-state sequence. Gross total tumor resection was performed and the pathology was consistent with RM. Immediately after surgery, her VA and IOP were 20/17 and 10 mmHg, respectively, with a remarkable resolution of the intramedullary hyperintensity.

Conclusion: Prominent hyperintensity in the ON identified in patients with chronic intracranial hypertension may be an indicator of visual disturbance. It can rapidly resolve after resolution of intracranial hypertension with functional recovery.

Keywords: Chronic intracranial hypertension, Optic nerve, Intramedullary hyperintensity, Rhabdoid meningioma, Visual disturbance

INTRODUCTION

Meningiomas are the most common primary brain tumors in adulthood. Most of them are benign tumors and classified as the World Health Organization (WHO) Grade I. The epidemiology and treatment outcomes are reported to differ among children/adolescents (0–21 years), young adults (22–45 years), and older adults (>45 years).[3] Rhabdoid meningiomas (RMs) are a rare type of malignant, WHO Grade III, meningioma with an increased tendency for recurrence and possible metastasis and leptomeningeal dissemination.[4,5,9,12,13] The overall prognosis for

these patients is commonly poor.[16] Prominent peritumoral edema, cystic components, and bone involvement have been documented as characteristic neuroimaging findings.[7] Gross total resection followed by adjuvant radiotherapy has been recommended for the treatment of primary intracranial RMs.[2,14,15]

Studies have suggested that hyperintense areas in the optic nerves (ONs) and tracts that can be detected using T2-weighted imaging, constructive interference steady-state (CISS) sequences, and contrast-enhanced fluid-attenuated inversion recovery imaging may be important for maintaining normal visual function.[6,8,11]

Here, we report a 20-year-old woman with intracranial RM who presented with visual disturbances and prominent CISS hyperintensity in the intraorbital ONs that rapidly resolved following tumor resection.

CASE PRESENTATION

A 20-year-old woman without neurofibromatosis presented with a 1-year history of headaches. At presentation, the patient was well oriented and did not show any focal neurological deficits. Ophthalmological examination revealed papilledema in both the optic fundi [Figure 1]. Visual acuity (VA) was 20/50 on the right side and 20/40 on the left, with an intraocular pressure of 17 mmHg on both sides [Table 1]. Cranial computed tomography revealed a large tumor in the frontal lobe accompanied by focal erosion in the adjacent skull [Figure 2a]. Magnetic resonance imaging (MRI) showed a tumor with broad-based attachment to the convexity dura mater. It appeared heterogeneous intensity on both T1- and T2-weighted sequences, accompanied by cyst components, measuring 82 × 65 × 70 mm in diameter, and inhomogeneously enhanced. No apparent dural tail was identified, and peritumoral brain edema was scant [Figures 2b-d]. On the CISS sequence, the intraorbital ONs appeared prominent hyperintensity [Figure 3]. The widths and heights of the optic sheaths (OSs) and nerves were measured at the orbital apex as shown in Figure 4 and Table 1. The patient underwent microsurgical tumor resection. The tumor tissue was grayish, elastic soft and supplied by the peripheral branches of the middle meningeal artery at the site of the skull erosion. A gross total resection was achieved. Resected specimens revealed rhabdoid-shaped anaplastic cells with pleomorphisms. Focal microvascular proliferation and geographic necrosis were observed, in addition to four mitotic figures identified in

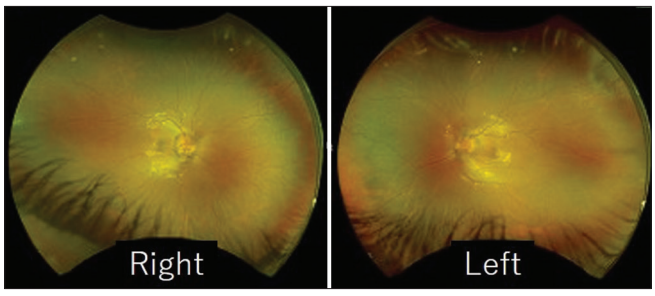

Figure 1: Fundus photos showing a papilledema on both sides.

Table 1: Summary of measurements.

	PreOp		POD5	
	Right	Left	Right	Left
a (mm)	6.35	6.51	6.25	5.84
b (mm)	5.86	6.19	5.84	6.05
c (mm)	3.58	3.75	2.42	3.22
d (mm)	3.58	3.26	3.22	3.23
	POD6			
VA	20/50	20/40	20/17	20/17
IOP (mmHg)	17	17	10	10

IOP: Intraocular pressure, POD: Postoperative day, PreOp: Preoperative, VA: Visual acuity, a: Width of the optic sheath, b: Height of the optic sheath, c: Width of the optic nerve, d: Height of the optic nerve.

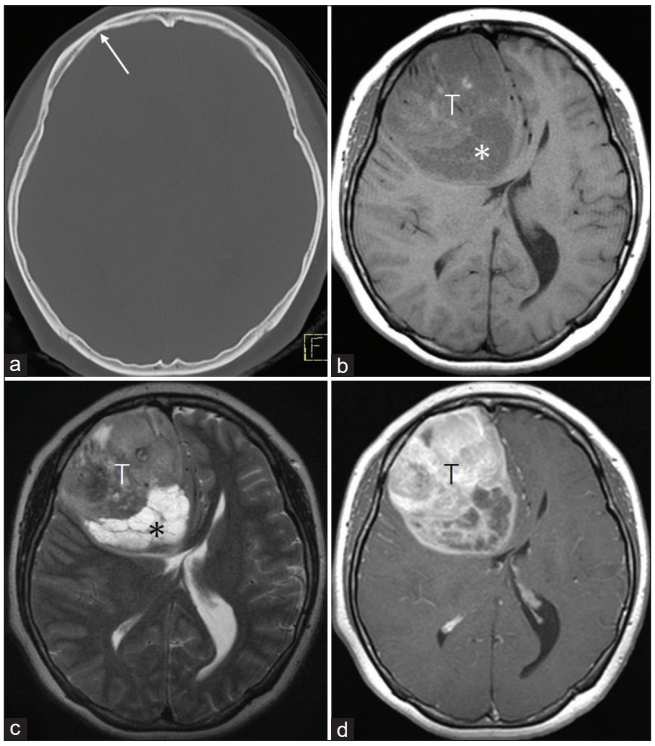

Figure 2: Axial computed tomography, bone-target image (a) and T1- (b), T2- (c), and postcontrast axial (d) T1-weighted magnetic resonance images at the same level showing a large tumor (T) in the right frontal lobe. The tumor has a broad-based attachment to the convexity dura mater. It appears heterogeneous intensity both on T1- and T2-weighted sequences, accompanied by focal erosion in the adjacent skull (a, arrow) and cyst components (b and c, asterisk), and inhomogeneously enhanced (d). No apparent dural tail is identified.

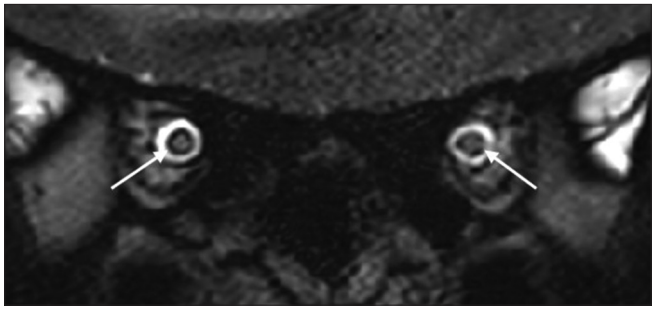

Figure 3: Coronal constructive interference steady-state image at the level of orbital apex, performed at the presentation, showing prominent hyperintensities in the intraorbital optic nerves (arrow).

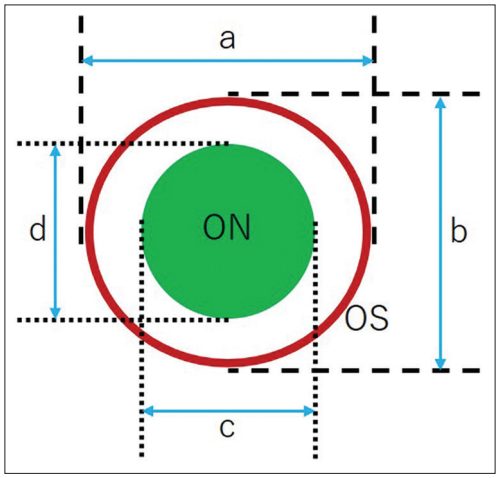

Figure 4: Schematic drawing showing measurements of the width and height of the optic sheath (a and b) and nerve (c and d), respectively. ON: Optic nerve, OS: Optic sheath.

10 high-power fields [Figure 5a]. Immunohistochemical examination showed positive staining for vimentin, S100 protein, progesterone receptor, and glial fibrillary acidic protein but negative for epithelial membrane antigen, CD34, STAT6, and Melan A. The MIB-1 index accounted for 50% [Figures 5b-f]. These findings highly suggested RM. The patient's postoperative course was uneventful. Her VA and IOP on postoperative day (POD) 6 were 20/17 and 10 mmHg, respectively [Table 1]. On the CISS sequence performed on POD 5, prominent hyperintensity in the ONs showed remarkable resolution [Figure 6]. Furthermore, measurements of the OSs and nerves revealed a clear decrease in the width of the right ON. No alterations were observed in the OS measurements [Table 1]. Although she did not receive immediate prophylactic radiotherapy, she remained under close surveillance, with MRI scans repeated every 6 months.

DISCUSSION

In this case, the prominent hyperintensity identified in the ONs presented simultaneously with long-term headache, bilateral papilledema, and visual disturbance, suggesting chronic intracranial hypertension. The offending tumor was large and occupied considerable space in the intracranial cavity. Furthermore, the intramedullary hyperintensity rapidly resolved after tumor resection, with remarkable improvement in visual function and decrease in IOP. Therefore, we assumed that the hyperintensity reflected the visual function both in before and after surgery. Recently, inactivation of the breast cancer-associated protein-1 tumor suppressor gene, BAP1, has been proposed to be the most

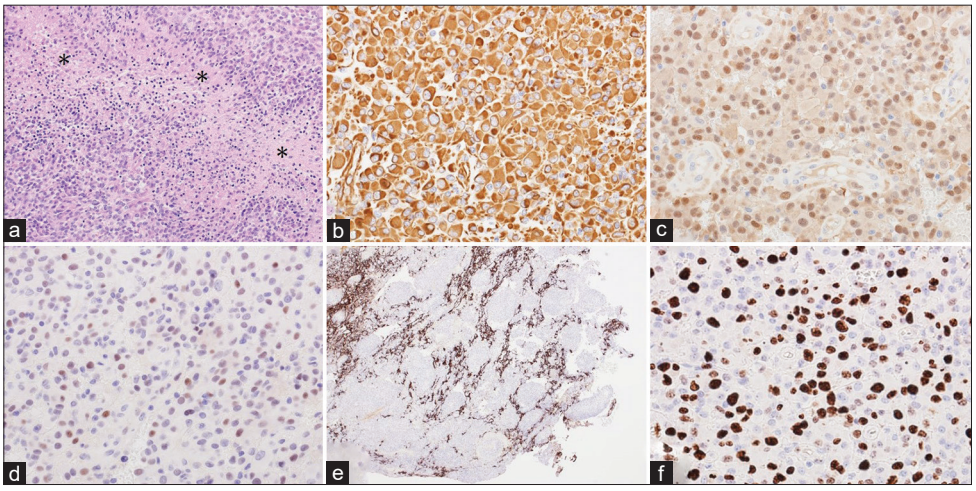

Figure 5: Photomicrographs of the resected specimens showing tumor tissue comprised of rhabdoid-shaped, anaplastic cells with pleomorphism and intervening necrotic foci (a, asterisks). Immunohistochemical analysis show positive staining for vimentin (b), S100 protein (c), progesterone receptor (d), and glial fibrillary acidic protein (e). The MIB-1 index accounts for 50% (f). (a) Hematoxylin and eosin stain, ×200; (b-d, f) ×400; (e) ×40.

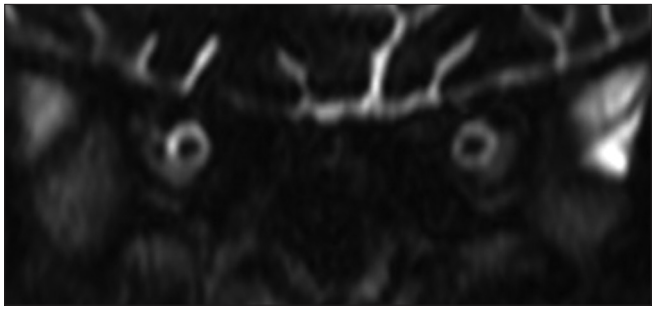

Figure 6: Coronal constructive interference steady-state image at the same level of Figure 3, performed on postoperative day 5, showing resolution of the hyperintensities in the intraorbital optic nerves.

common mutation associated with an aggressive nature of RMs.[10] In the present case, BAP1 was not explored.

In our patient, the width of the right ON decreased postoperatively. This probably indicates long-term, indirect tumor compression of the intracranial right ON, and consequent optic atrophy. Despite this, the visual disturbance improved immediately after surgery. Such intramedullary hyperintensity may serve as a radiological biomarker for indicating a prompt surgical intervention. The diameter of the OS did not show immediate shrinkage, consistent with the results of previous report.[1] This may be due to the hard composition of the OS with collagen fibers.

Prominent peritumoral edema, cystic components, and bone involvement are documented as characteristic neuroimaging findings of RMs.[7] This RM was accompanied by cysts, but lacked a dural tail with scant peritumoral edema. Furthermore, the patient presented with focal erosion of the skull, instead of hyperostosis. RM has been reported to exhibit heterogeneous histological findings and a wide variety of chromosomal alterations, which may contribute to atypical radiological findings like present meningioma.[5] It is important to recognize that RM may have a variable appearance on neuroimaging and attempt total resection when suspected.

CONCLUSION

Prominent hyperintensity in the ON identified in patients with chronic intracranial hypertension may be an indicator of visual disturbance. It can rapidly resolve after resolution of intracranial hypertension with functional recovery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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