Color Doppler Ultrasound Imaging of Primary Optic Nerve Sheath Meningioma

Tzong-Shyue Liu and Mon-Te Kahn

A 72-year-old woman presented with a history of progressive blurred vision of the left eye for more than 10 years and left proptosis for about 3 years. Her best-corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. Examination of the left fundus revealed temporal pallor of the optic disc and abnormal retinal vascular architecture. A retrobulbar intraconal lesion was seen on magnetic resonance imaging (MRI), which displaced the optic nerve upward and laterally to the left. Color Doppler flow imaging showed a globular, retrobulbar solid mass with the high-blood-flow hemodynamics of a tumor, compared to the low-blood-flow hemodynamics of normal orbital tissue. The tumor was partially excised and the immunohistopathologic diagnosis was meningothelial meningioma. The proptosis improved after surgery, but the corrected visual acuity remained the same in the first year of postoperative follow-up.

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KEY WORDS: • optic nerve sheath meningioma • doppler imaging • orbital tumor • blood flow

INTRODUCTION

Meningiomas are the second most common brain neoplasm after gliomas, representing 15% to 20% of all intracranial tumors in adults [1]. Some of them, usually from the sphenoid ridge or tuberculum sellae, invade the optic canal and orbit by extending between the dura and arachnoid membranes of the optic nerve in these regions. These intracranial tumors involving the orbit are considered secondary optic nerve sheath meningiomas. Primary optic nerve sheath meningiomas are those that arise from the cap cells of the arachnoid membrane surrounding the intraorbital, or less commonly, the intracanalicular portions of the optic nerve [2]. Optic nerve sheath meningiomas account for only 1.7% of all orbital tumors. Primary optic nerve sheath meningiomas are even rarer: only 10% of optic nerve sheath meningiomas are primary [1].

Color Doppler imaging (CDI) is an advanced ultrasound technique that combines B-mode ultrasonography, color representation of blood flow and pulsed-Doppler measurement of blood flow velocities. CDI is widely used as a powerful imaging technique in many medical specialties, but its application in ophthalmology is limited. There are only a few reports of the use of CDI in the study of orbital tumors in ophthalmic literature. We report a case of primary optic nerve sheath meningioma and the hemodynamic analysis from CDI.
CASE REPORT

A 72-year-old woman presented with a history of slowly progressive blurred vision in the left eye for more than 10 years and left proptosis for about 3 years. Her best-corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. The intraocular pressure and anterior segment were normal in both eyes. Exophthalmometry showed left proptosis of 16 mm in the left eye (right eye, 13 mm). Examination of the left fundus revealed temporal pallor of the optic disc, fine macular chorioretinal folds, and distorted retinal vascular architecture.

There was mild macular fluorescent staining on fluorescence angiography, but neither disc dye leakage nor optociliary shunt vessels were noted. B-mode ultrasonography showed a solid retrobulbar tumor measuring approximately 17 mm x 12 mm. Magnetic resonance imaging (MRI) confirmed a retrobulbar intraconal lesion, which displaced the optic nerve upward and laterally to the left (Fig. 1). The tumor was confined within the intraconal space and no evidence of intracranial spread was noted. No intracranial tumor was found, ruling out the possibility of secondary intraorbital tumor from an intracranial lesion.

CDI showed a globular retrobulbar solid mass with high-blood-flow hemodynamics. The tumor had two feeding arteries. These had different orientations:

- Feeding artery 1 (FA1) was in the oblique posterior orientation;
- Feeding artery 2 (FA2) was in the oblique anterior orientation. Both arteries showed high blood flow, compared with the low blood flow of normal ocular arteries. The peak systolic velocities (PSV) were 71.4 cm/sec in FA1 and 29.0 cm/sec in FA2 (Figs. 2 and 3). The respective PSVs of the ophthalmic artery and central retinal artery were 15.2 cm/sec and 11.7 cm/sec – far slower than the feeding arteries of the tumor. The CDI results in our patient are compared with those of normal Taiwanese eyes in the Table [3].

**Fig. 1.** A retrobulbar tumor, shown on MRI, displacing the optic nerve upward and laterally. The tumor was confined within the intraconal space and no evidence of intracranial spread was noted.

**Fig. 2.** Feeding artery 1 of the tumor shows high blood flow. Peak systolic velocity is 71.4 cm/sec, mean velocity is 40.7 cm/sec, and end diastolic velocity is 17.1 cm/sec.

**Fig. 3.** Although slower than feeding artery 1, feeding artery 2 of the tumor still shows high blood flow; peak systolic velocity is 29.0 cm/sec. Feeding artery 2 may be independent of feeding artery 1 or it may be the distal end of feeding artery 1.
Due to the indeterminate diagnosis, an excisional biopsy through a medial orbitotomy was performed. During surgery, the tumor could not be separated from the optic nerve without damaging the optic nerve, so the tumor was only partially excised. The immunohistopathologic diagnosis was meningothelial meningioma. The patient’s proptosis improved after surgery, but her corrected visual acuity remained the same in the first year of postoperative follow-up.

**DISCUSSION**

Visual impairment, proptosis, disc swelling, and optic atrophy are the common findings of optic nerve sheath meningiomas. Although visual loss, optic atrophy, and optociliary shunt vessels are considered the pathognomonic triad of optic nerve sheath meningiomas [4], the simultaneous occurrence of all three findings is uncommon. Gradual visual loss is the most frequent presenting symptom. Disc atrophy and disc swelling are also typical findings of optic nerve sheath meningiomas. Overall, more than 90% of patients will show one or the other of these two findings [1]. Optociliary shunt vessels represent dilated collateral channels between the retinal veins and the choroid, resulting from compression of the central retinal vein. Only about 30% of optic nerve sheath meningiomas demonstrate this finding. Our patient presented with decreased vision, proptosis and a pale disc; all are typical findings of optic nerve sheath meningiomas. Optociliary shunt vessels were not seen in our patient.

There are three types of optic nerve sheath meningiomas that can be seen on computed tomography (CT) or MRI: a diffuse tubular enlarged type (64%), a fusiform type (23%), and a globular type (12%) [5]. "Railroad tracks" (or "tram-tracking") is a CT sign found in the diffuse tubular enlarged type, in which the denser and thickened optic nerve sheath outlines a central lucency representing the residual optic nerve. This sign is seen in about 25% of all optic nerve sheath meningiomas [6] and is a characteristic finding. The fusiform type is sometimes confused with an optic nerve glioma. About 10% to 20% of optic nerve sheath meningiomas cannot be differentiated from gliomas without a tissue diagnosis [7]. The relatively uncommon globular type is due to tumor growth outside the dura sheath. On MRI, optic nerve sheath meningiomas present iso-intense or slightly hyper-intense signal intensity compared to the normal optic nerve on both T1- and T2-weighted sequences. In our patient, MRI showed an intracanal mass lesion, without evidence of intracranial invasion. The picture was compatible with a primary optic nerve sheath meningioma.

Although CDI is widely used in many medical specialties, it is not as popular in ophthalmology. However, there are some published reports of CDI data from normal eyes [3, 8–12]. The applications of CDI in ophthalmic diseases include orbital vascular occlusion, orbital tumor, glaucoma, diabetic retinopathy and carotidocavernous fistula [13]. There are only a few reports of the use of CDI in the study of orbital tumors. In a series of 44 patients with

<table>
<thead>
<tr>
<th>Indices</th>
<th>PSV</th>
<th>MV</th>
<th>EDV</th>
<th>PI</th>
<th>RI</th>
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<tr>
<td>Meningioma</td>
<td>OA</td>
<td>15.2</td>
<td>8.78</td>
<td>6.53</td>
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<tr>
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<td>CRA</td>
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<td>8.15</td>
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<td>40.7</td>
<td>17.1</td>
<td>1.30</td>
</tr>
<tr>
<td></td>
<td>FA2</td>
<td>29.0</td>
<td>14.0</td>
<td>7.25</td>
<td>1.55</td>
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| Normal eyes | OA   | 32.86 ± 13.38 | 13.69 ± 4.38 | 7.15 ± 2.36 | 1.81 ± 0.45 | 0.7 ± 0.08 |
|             | CRA  | 12.48 ± 4.03  | 7.61 ± 2.48  | 5.55 ± 2.09 | 0.93 ± 0.30 | 0.56 ± 0.11 |

OA = ophthalmic artery; CRA = central retinal artery; FA1 = feeding artery 1; FA2 = feeding artery 2; PSV = peak systolic velocity (cm/sec); MV = mean velocity (cm/sec); EDV = end diastolic velocity (cm/sec); PI = pulsatility index, RI = resistivity index; SD = standard deviation.
intraocular mass lesions [14], Doppler signals of feeding arteries were detected in 38 (90.5%). Abnormal Doppler signals were detected in 26 (92.8%) out of 28 suspected choroidal melanomas, and in all four choroidal hemangiomas and choroidal metastases. In contrast, in three patients with tumor-simulating lesions (two cases of age-related macular degeneration with extensive subretinal hemorrhage and one case of dense vitreous hemorrhage), no abnormal flow could be detected. Lesions that can simulate choroidal tumors, such as large subretinal hemorrhages, usually do not have a distinctive blood supply. Therefore, they can be differentiated from neoplasms on the basis of the absence of feeding arteries.

In choroidal hemangiomas, high PSV in a feeding artery can be seen together with high end diastolic velocity (EDV). These findings are compatible with the pathologic features of this type of highly vascularized tumor. The flow pattern in choroidal melanomas is medium to high PSV with high EDV, which is characteristic of most neoplasms. Guthoff et al presented a CDI analysis of choroidal melanomas [15]. The average PSV of the tumor vessels of 41 melanomas was 18.8 ± 7.6 cm/sec. Moreover, they noted that the PSV often rose in the first few days after radiotherapy and then decreased significantly from pretreatment levels, frequently before tumor regression was detectable by conventional ultrasonography.

There was no report of CDI analysis on optic nerve sheath meningioma. Our data showed two feeding arteries (FA1 and FA2) with high PSVs (71.4 cm/sec and 29.0 cm/sec) and high EDVs (17.1 cm/sec and 7.25 cm/sec). FA1 was in the oblique posterior orientation, while FA2 was in the oblique anterior orientation. Due to the different orientations, the directions shown on Doppler imaging were the reverse of one another (FA1 had negative velocity and FA2 had positive velocity). These two feeding arteries may be independent or one may be the distal end of the other. Nevertheless, the high blood flow is compatible with previous reports of orbital tumors [14, 15] and represents the tumor vasculature. The PSV of the ophthalmic artery was slower than normal (15.2 cm/sec vs. 32.86 ± 13.38 cm/sec), and may be explained by the "steal phenomenon" in which the normal blood flow is shared by the feeding arteries of the tumor. The central retinal artery blood flow was within the normal range. Compared with previous reports [14, 15], we found that optic nerve sheath meningiomas, similar to choroidal melanomas, have high-blood-flow hemodynamics.

There is much literature regarding the treatment of primary optic nerve sheath meningiomas. Due to its critical location and unpredictable biologic activity, the management of primary optic nerve sheath meningiomas remains controversial. Surgical removal is the mainstay of therapy. In most cases, excision of orbital meningiomas results in blindness. The majority of such tumors share their blood supply with the pia mater and attempted tumor excision more often than not results in interruption of the optic nerve. Of 120 patients with primary optic nerve sheath meningiomas treated with surgery alone, 94 (78%) showed postoperative loss of vision to the extent of no light perception [1]. Due to the high risk of postoperative blindness, some authors recommend observation, not surgery, until all useful vision is lost or when there is evidence of intracranial tumor spread.

As our patient had some useful vision with no evidence of intracranial tumor spread, partial tumor resection, with vision preservation, was reasonable. Regular clinical examination and repeat CT are necessary to monitor the tumor.

**CONCLUSION**

We report a case of primary optic nerve sheath meningioma, with analysis of orbital hemodynamics. Although the CDI database of orbital tumors is limited, we still obtained useful information. CDI helps to differentiate neoplasms from neoplasm-like lesions. Furthermore, CDI can be used to monitor the therapeutic effects of radiotherapy [14, 15]. As CDI allows for non-invasive assessment of the retrobulbar vasculature, the applications of this technology in ophthalmology will further develop as the database becomes more complete.

**REFERENCES**


