CASE REPORT

3T magnetic resonance imaging in management of orbital venolymphatic malformations: two case reports

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Abstract

Vascular malformations may occur during the development of the arterial, venous, and lymphatic systems in isolation or in combination. Orbital venolymphatic malformations (OVLM) are particularly challenging to treat because they infiltrate and remodel normal structures within the tight orbital space and lead to substantial ocular discomfort, visual disability, and facial disfigurement. Herein, we report two cases of OVLMs with different presenting clinical features and treatment options. We compare the image qualities among computed tomography and 1.5T and 3T magnetic resonance imaging and discuss how better anatomical resolution in the latter aid in the management of OVLM.

Key words: Congenital abnormalities; Magnetic resonance imaging

Case presentation

Patient 1

In March 2020, a 26-year-old Chinese woman presented to the accident and emergency department with a 5-day history of spontaneous right eye painful swelling and periorbital bruises. She had congenital right facial and parotid cystic hygroma. At age 3 to 5 years, she had undergone three debulking surgeries, which led to postoperative right facial nerve palsy. She also had four episodes of spontaneous retrobulbar hemorrhage that required hospital admissions at age 9, 11, 13, and 16 years. Each episode was preceded by upper respiratory tract infections and resolved after a course of oral antibiotics and steroids. Computed tomography (CT) of the orbit was performed during each episode.

On examination, her right eye had a 5-mm proptosis (baseline, 2 mm), near-complete ophthalmoplegia, and an intraocular pressure (IOP) of 48 mmHg. Best-corrected visual acuity was 20/30 without relative afferent papillary defect or optic disc swelling. Examination of the left eye was unremarkable. Urgent contrast CT of the orbits showed a homogenous soft tissue mass measuring 2.0×3.9×2.1 cm at the right retrobulbar intraconal region, which had increased in size compared with previous scans in 2014 (**Figure 1**). The mass appeared inseparable from the optic nerve and the medial and inferior recti and involved the right orbital apex and cavernous sinus. The orbital compartment syndrome resolved gradually after bed rest, anti-glaucomatous drops, and oral steroid. At 4 weeks after onset, 3T magnetic resonance imaging (Prisma Magnetom,

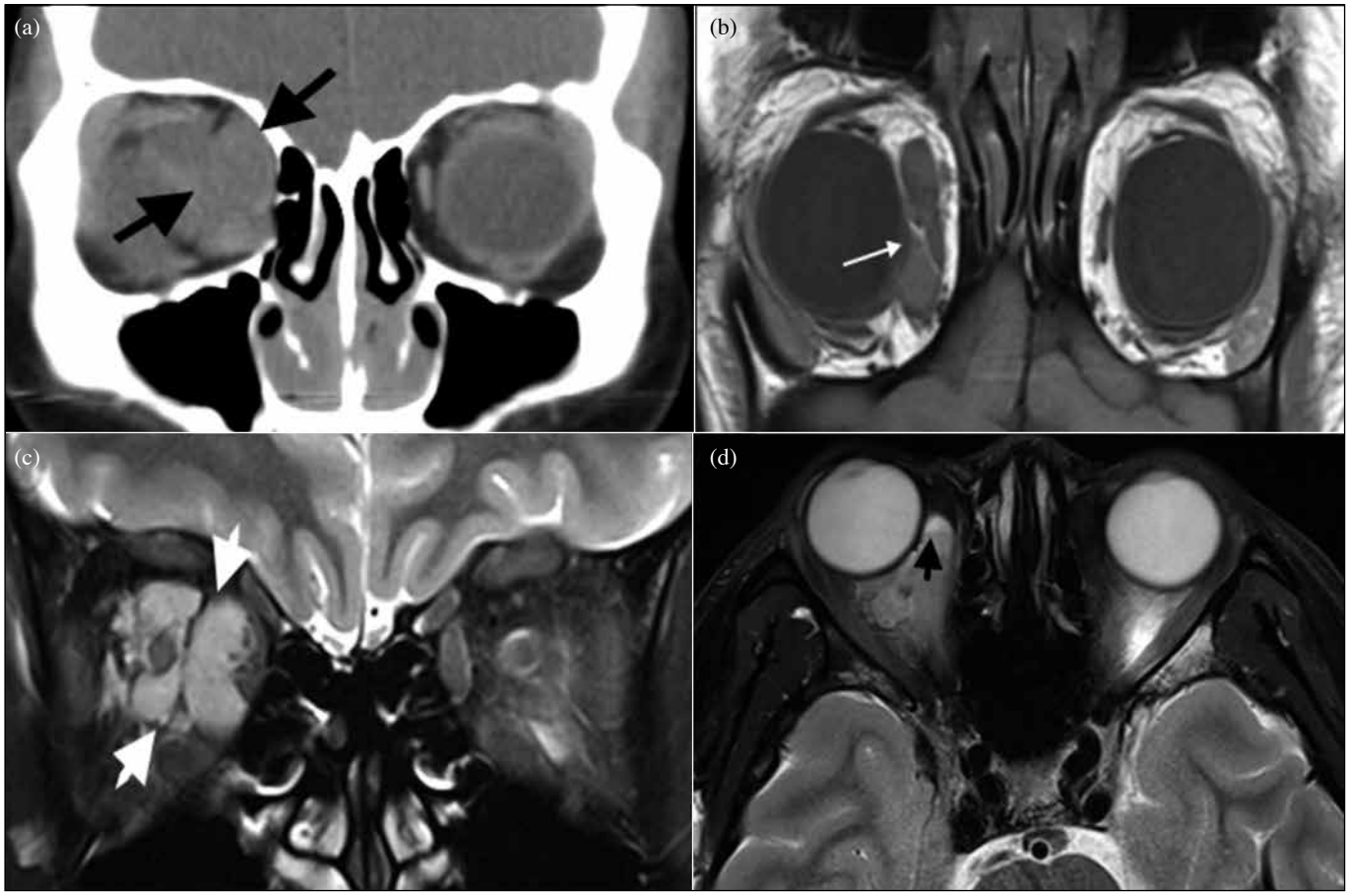

Figure 1. Patient 1: (a) Contrast computed tomography showing a soft tissue mass measuring 2.0×3.9×2.1 cm at the right retrobulbar intraconal region. (b) 3T coronal T1-weighted magnetic resonance imaging (MRI) at 4 weeks after onset of symptoms showing a multilobulated septated trans-spatial cystic lesion, slightly hyperintense to the extraocular muscle, occupying the right retrobulbar region. (c) Coronal T2-weighted MRI showing encasement of the optic nerve by the lesion and medial displacement of the medial rectus muscle. (d) Axial fat-suppressed T2-weighted MRI showing distinct fluid-fluid levels within the lesion suggestive of resolving intralesional hemorrhages.

Siemens Healthineers, Germany) showed a multi-lobulated septated cystic and venolymphatic malformation occupying almost the entire right retrobulbar region (**Figure 1**). The optic nerve was encased by the lesion and multiple intralesional fluid-fluid levels were present, suggestive of recent hemorrhages.

The patient was given treatment options of glue-assisted excision, sclerotherapy, combination of both, and oral Sirolimus. She opted for oral Sirolimus 1.5 mg daily.

Patient 2

In April 2018, a 30-year-old Chinese man presented with an enlarging purplish lesion in the medial canthus of the right eye, which increased in size with Valsalva maneuver and dependent posture. Physical examination showed a Valsalva-positive medial palpebral and caruncular conjunctival vascular lesion. His visual acuity was 20/18 in either eye with no diplopia or proptosis. Contrast CT of the orbits showed an enhancing soft tissue at the medial aspect of the right orbit, with the presence of phlebolith and bony remodeling, suggestive of long-standing communicating venous-dominant type of venolymphatic malformation (**Figure 2**).

Contrast 1.5T MRI performed elsewhere showed a poorly circumscribed, predominantly extraconal, transcompartmental mass measuring 13×21×17 mm suggestive of orbital communicating venous malformation (**Figure 2**). The lesion was T1-isointense to muscle and showed homogeneous contrast enhancement. 3T MRI showed multiple cystic components and fluid-fluid levels within the lesion, with better anatomically defined margins (**Figure 2**). The patient was given treatment options of glue-assisted excision or oral Sirolimus. He opted for glue-assisted excision.

Discussion

Vascular malformations may occur during the development of the arterial, venous, and lymphatic systems in isolation or in combination.[1] OVLMs are non-neoplastic congenital

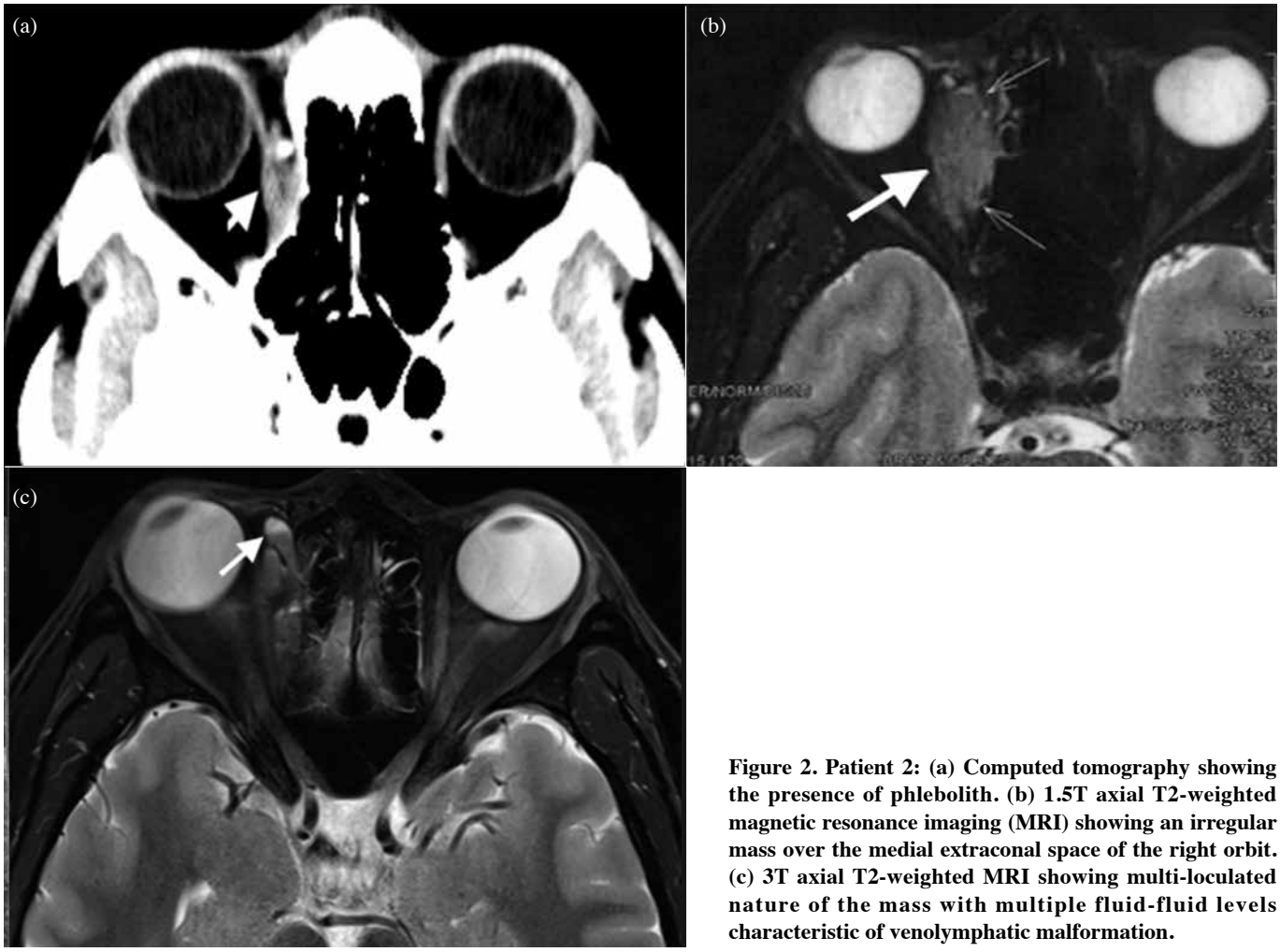

Figure 2. Patient 2: (a) Computed tomography showing the presence of phlebolith. (b) 1.5T axial T2-weighted magnetic resonance imaging (MRI) showing an irregular mass over the medial extraconal space of the right orbit. (c) 3T axial T2-weighted MRI showing multi-loculated nature of the mass with multiple fluid-fluid levels characteristic of venolymphatic malformation.

malformations and are classified into superficial, deep, combined (superficial and deep), and complex (orbital and extra-orbital) types according to their locations.[2] Clinical manifestations and management are based on hemodynamics and classified into no flow (type 1), low flow (type 2), and high flow (type 3). According to International Society for the Study of Vascular Anomalies, vascular malformation can be divided into high-flow (type 3) and low-flow (types 1 and 2) lesions. High-flow lesions include arteriovenous malformation and congenital arteriovenous fistula. Low-flow lesions include venous malformations, lymphatic malformations, and veno-lymphatic malformations.[3]

OVLMs are the most common low-flow orbital lesions and have mixed vascular elements histologically with different degrees of lymphatic versus venous components. Patients may present with proptosis, pain, limited extraocular movement, globe dystopia, and rarely loss of vision. Acute hemorrhage occurs after minor trauma or viral infections and may result in orbital compartment syndrome.

Imaging is crucial in the management of OVLMs in terms of location, nature, progression, and treatment. CT provides excellent details related to the bony anatomy, bony changes, and phleboliths, which may be seen in slow-flow lesions.[4] MRI is superior to CT in differentiating acute from chronic blood- or fluid-filled cysts and in providing anatomical details including loculation within the lesion and their relationship with the optic nerve, extraocular muscles, and cavernous sinus. Imaging quality of MRI depends on the magnetic field strength measured in Tesla (T), usually between 0.5 T and 3.0 T. With increasing tesla strength, the image quality increases and the time taken to perform the imaging decreases.[5] In 2002, the first 3T MRI scanner was approved for clinical use by the US Food and Drug Administration.[4] 3T MRI offers a higher signal-to-noise ratio than does the lower-field strength MRI, which leads to a higher spatial resolution constant.[5] At present, 1.5T short-bore MRI remains the standard technology in most hospitals. For head and neck, neural, musculoskeletal, and vascular imaging, 3T MRI has a stronger signal strength and higher spatial image resolution and is better at delineating anatomical details and tissue planes, compared with 1.5T MRI or CT. 3T MRI can also generate a higher contrast-to-noise ratio. This is particularly useful as orbital structures can be difficult to differentiate owing to their limited inherent contrast.[6] Valsalva maneuver on supine position may be performed during MRI when orbital varices are

suspected in order to determine the presence and the relative contribution of the distensible (communicating) component.

The management of OVLMs is challenging. The first-line treatment includes observation or simple aspiration of the anterior cystic components. Surgical debulking is difficult because of the poorly defined margin of vascular malformations, the risk of uncontrolled bleeding, the proximity to orbital structures, and the high recurrence risk related to incomplete excision. Recent development of therapeutic agents and advancements in MRI enable image-guided, individualized treatment of symptomatic lesions. From oral (sirolimus) to intralesional agents using sclerosants (bleomycin, doxycycline, sodium tetradecyl sulfate, alcohol) targeted for micro- and macrocystic lesions, liquid polymers (N-butyl cyanoacrylate glue) for venous components followed by controlled surgical debulking has evolved as a staged, multimodal approach for complex OVLMs. Although percutaneous sclerotherapy under sonographic and/or fluoroscopic guidance has been performed, complex lesions may benefit from additional orbitotomy exposure and intraoperative navigation guidance. Intermittent 3T MRI was recently used with 'in-and-out' technique to monitor MRI-compatible needle placement and sclerosant delivery during percutaneous treatment of venous malformation.[6]

Conclusion

3T MRI is superior to 1.5T MRI or CT in management of OVLMs.

Contributors

PTYW and KKLC designed the study. PTYW, KKLC, and WCWC acquired the data. PTYW, KKWC, and KKLC analyzed the data. PTYW drafted the manuscript. All authors critically revised the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflict of interest

All authors have disclosed no conflicts of interest.

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Data availability

All data generated or analyzed during the present study are available from the corresponding author on reasonable request.

Ethics approval

The patients were treated in accordance with the tenets of the Declaration of Helsinki. The patients provided written informed consent for all treatment and procedures and for publication.

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