Abstract

Combined venous lymphatic malformation is a benign, nonhereditary vascular anomaly. These unencapsulated hamartomas can be associated with spontaneous, recurrent hemorrhage leading to proptosis and pain. We describe a patient with this lesion to discuss the various management strategies and surgical challenges encountered with this condition.

Key Words: venous lymphatic malformation, lymphangioma, vascular anomaly

Introduction

The appropriate classification of combined venous lymphatic anomalies has been controversial. These congenital lesions display features of both venous and lymphatic tissue and are characterized by tortuous, frail vessels with a propensity to hemorrhage, posing a medical challenge for proper management.

Case Report

A six-year-old boy was referred to the orbital and oculoplastics service for fullness of the right upper eyelid, which had been present since birth. Examination showed 5mm increased right side proptosis with 2mm right hypogobulus and ptosis of the right upper eyelid. A mass was palpated along the right superomedialed orbit (Figure 1a). Best-corrected visual acuity was 20/20 OU; extraocular movements were full, with normal pupillary reaction and color vision. MRI confirmed the diagnosis, demonstrating a multicystic soft tissue mass extending from the medial canthus of the right orbit into the intraconal space, enveloping the optic nerve, without evidence of direct compression (Figure 1b).

Due to the significant cosmetic effect of the anterior component of the malformation and our patient’s negative social interactions at school, an anterior surgical approach through the right upper eyelid was undertaken to debulk the hamartoma and correct the ptosis. Care was taken to avoid extensive intraconal manipulation, as this can result in hemorrhage and optic nerve compromise. Microscopic study of the excised tissue demonstrated irregularly dilated vessels with scant stroma and smooth muscle fibers.

One week after surgery the patient was very happy showing significant decrease in the mass effect with 20/20 vision with normal optic nerve function. Three days after, the patient presented to the emergency department with no light perception vision and a relative afferent pupillary defect in the right eye (Figure 2a). MRI demonstrated increased right side proptosis with acute and extensive intraconal and extracranal hemorrhage. An urgent medial orbitotomy with drainage of intraconal blood cysts was performed, and our patient recovered normal optic nerve function and vision to 20/20 (Figure 2b).

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Discussion

Combined venous lymphatic malformations of the orbit are benign lesions, typically unilateral, with no gender predominance. These lesions are present at birth; however, they often remain inconspicuous due to small size.\(^1\)

The exact pathogenesis is unknown; many speculate that these hamartomas embryologically derive from the vascular system with differentiation into both lymphatic and venous tissue. Microscopic study and immunohistochemical staining confirms the presence of two distinct tissues.\(^1,2,4\)

Clinically, patients may present with pain, proptosis, diplopia or a visible mass along the eyelid. Many cases are diagnosed during childhood due to sudden enlargement of the vascular hamartoma.\(^1,2,3\)

This acute expansion can be secondary to lymphatic tissue hyperplasia due to an upper respiratory infection, or due to intrinsic hemorrhage from small intrallesional fragile vessels.\(^1,2\)

Once significant intracranial hemorrhage has formed, compressive optic neuropathy may develop.

If a combined venous lymphatic malformation is suspected, appropriate orbital imaging is recommended and magnetic resonance imaging (MRI) has emerged as the modality of choice. On MRI, an unencapsulated soft tissue mass with multiple cystic spaces in the intraconal or extraconal space can be identified and the intensity of the lesion may indicate the age of the hemorrhage.\(^5\)

Other more invasive imaging studies such as venography and Doppler do not adequately highlight these hamartomas because these lesions are isolated from the normal orbital vasculature.\(^1,2\)

This unique property distinguishes them from orbital varices. When the diagnosis is confirmed, additional imaging of the brain is recommended as an association with intracranial vascular anomalies has been documented.\(^6\)

Generally, conservative management is encouraged as the risk of surgical removal can threaten both vision and ocular motility.\(^7\)

With this approach, frequent clinical exams and radiologic monitoring is required as the malformation can suddenly increase in size. Interestingly, spontaneous regression of these lesions and associated hemorrhage has been reported.\(^3,7\)

The indications for surgery are controversial, as intraoperative manipulation of the lesion can induce future hemorrhage due to the thin-walled, fragile vessels and their tendency to rupture.\(^8\)

Surgical intervention is indicated if there is significant acute expansion leading to compression of the optic nerve. In addition, if amblyopia is of major concern or a well-isolated lesion is extensively affecting cosmesis, surgery can be considered.\(^1,2\)

Most lesions do extend deep into the orbit and are not well circumscribed, often surrounding the optic nerve and other key vessels, preventing complete surgical excision. Therefore, many surgeons advocate drainage of the blood cysts and conservative debulking of the lesion.\(^1,3,8,9\)

Materials such as cyanoacrylate glue and ethanol can be used to embolize these malformations, as an adjunct to subsequent surgical removal. A newer sclerosing agent, n-butyl-2-cyanoacrylate (Trufill\(^8\)), DePuy Synthes – Johnson&Johnson) has allowed for greater successful excision with relatively less blood loss and complications.\(^8\)

Combined venous lymphatic malformations are complex vascular lesions that pose clinical and surgical challenges for the ophthalmologist and patient. Conservative treatment with meticulous follow-up is advocated, with surgery reserved for critical cases with evidence of optic nerve compromise, amblyopia or significant cosmetic detriment.

\[\text{REFERENCES}\]