CASE REPORT

RETROBULBAR LYMPHANGIOMA– A CASE REPORT

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ABSTRACT: We are presenting a rare case of 8months old female baby from Gulbarga who was brought to our hospital with chief complaints of swelling in right eye since 6 days. There was no history of trauma or fever. Patient was subjected to ultrasound and a hypoechoic lesion was noted in the retrobulbar area. Later patient underwent CECT and MRI for further evaluation and it was finally diagnosed as lymphangioma, also called lymphatic-venous malformation. The patient underwent surgery and the final histopathological diagnosis came to be lymphangioma.

KEYWORDS: Lymphangioma, retrobulbar.

INTRODUCTION: Lymphangiomas are benign hamartomatous tumors which are usually diagnosed in early childhood. About 20% of these tumors involve the orbit and ocular adnexa¹. These usually present at an early age, from infancy to the first decade of life.

Venous-lymphatic malformation in the orbit is an anomaly of venous and lymphatic development that is characterized by non-enhancing, cystic lymphatic and enhancing solid venous components. Intralesional hemorrhage is common and frequently produces distinctive fluid-fluid levels within the cystic portion.

CASE SUMMARY: An 8 month old female baby from Gulbarga was brought to our hospital with swelling of right eye. Patient was referred to radiology department for evaluation.

Ultrasound: Hypoechoic solid mass lesion was noted in the retrobulbar area in right eye. Optic nerve appeared normal. No evidence of calcification was noted within the lesion or periphery of the lesion. Bulbar area of right eye appeared normal. Left eye appeared normal.

On color Doppler: No vascularity was noted within the centre or periphery of the lesion.

NECT: Evidence of hyperdense lesion with CT value of +50 to + 60 HU was noted in retrobulbar area in right eye. Lesion involved intraconal compartment. The lesion was predominantly located in the medial aspect of right eye. Optic nerve appeared normal but was pushed superiorly and laterally by the lesion. Proptosis of right eye was noted. Scalloping of right orbit was noted. No evidence of calcification was noted within the lesion or periphery of the lesion. Bulbar region of right eye appeared normal. Left eye appeared normal.

CECT: Lesion showed moderate enhancement.

MRI: Evidence of solid mass in right orbit was noted. It appeared to be heterogenous with large component which was hypointense on T1WI and hyperintense on T2WI. Lesion was intraconal in location. It measured around 32 x 22 mm in longitudinal and transverse dimensions and was located predominantly in the postero-medial aspect of the globe, with components also located anterior-inferior to the globe. Component located antero-inferior to the globe showed evidence of fluid levels. The globe itself appeared normal. Left eye appeared normal. Visualized intracranial structures were normal.

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On the basis of imaging patient was diagnosed to have lymphangioma of right eye, after which patient underwent surgery and the specimen was send for histopathology.

Histopathology: At histologic analysis, the malformation was composed of a network of irregular, anastomosing vascular channels of varying sizes and lined by a flat layer of endothelium within a loose, hypocellular stroma. Poorly developed smooth muscle fibers, well-developed lymphoid aggregates, and loose connective tissue septa were typically found randomly distributed within the surrounding stroma. The dysplastic channels were filled with serous fluid. Features were suggestive of lymphangioma.

DISCUSSION: Venous-lymphatic malformations are prevalent vascular lesions in the orbit. They represent 4% of all expanding pediatric orbital masses. The malformations are present at birth, but they may not be discovered until they undergo expansion. Patients present in childhood with slowly progressive exophthalmos or with sudden proptosis due to intratumoral hemorrhage and expansion.² The mass is irregular, lobulated, and infiltrating with ill-defined margins, features that reflect the lack of a capsule. The mass often involves both pre- and postseptal and intra- and extraconal portions of the orbit. The lesion can have components with cysts more than 1–2 cm in size (macrocytic) and portions that appear more solid (microcytic).³ Intralesional hemorrhage is common and frequently produces distinctive fluid-fluid levels within the cystic portions. Venous-lymphatic malformations of the orbit are associated with noncontiguous, ipsilateral, intracranial vascular anomalies.

CONCLUSION: This study highlights the significance of ultrasound, CT and MRI in diagnosis of lymphangioma. CT provides quicker scans, is able to image bone directly, and shows the presence of calcification better as well as involvement of bone. MRI has advantages over CT in its superior soft tissue contrast, its ability to image the orbit and intracranial structures free of beam hardening artifacts from the skull base.

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Fig. 1: Ultrasound - Hypoechoic solid mass lesion noted in right retrobulbar area.

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Fig. 2: Non-enhanced CT - Hypodense lesion in right retrobulbar region predominantly in postero-medial aspect.

Fig. 3: Contrast enhanced CT - Lesion shows moderate enhancement.

Fig. 4: MRI T1W Axial Images - Hypointense lesion involving extraconal as well as intraconal compartment of right eye

Fig. 5: MRI T1W Saggital Image - Fluid level in antero-inferior component of the lesion.

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Fig. 6: MRI T2W Axial image - Heterogenously hyperintense lesion with multiple septations and few hypointensities in intraconal

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