Successful regression of orbital lymphatic-venous malformation with sclerotherapy

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ABSTRACT

Orbital lymphatic-venous malformation is a rare multicystic malformation of the lymphatic and vascular system commonly affecting the paediatric population. A 16-year-old female with no known medical illness and no known drug allergy presented to us with gradual onset of left eye painless proptosis for 3 months, associated with blurring of vision for 2 weeks. There was no prior history of fever or trauma. Visual acuity was 3/60 (pinhole 6/36) in left eye and 6/9 in the right eye. She had grade 1 relative afferent pupillary defect. She was found to have restricted left extra-ocular muscle movements in all directions, associated with a 7 mm non-axial proptosis. There was also evidence of optic disc swelling and macula striation in the fundus examination. Magnetic resonance imaging of brain and orbit showed heterogenous enhancing mass at medial left orbit and presence of multiple phleboliths and cystic components with fluid filled level occupying the medial extraocular and retroorbital space involving the orbital apex suggestive of left orbital veno-lymphatic malformation with venous component predominance. There was no intracranial involvement. Due to the close proximity of the tumour to the optic nerve and high risk of bleeding intraoperatively as it was a highly vascularized tumour, interventional radiologist was consulted for sclerotherapy instead of debulking surgery. The patient underwent bleomycin sclerotherapy and showed an excellent response with the resolution of proptosis and improvement in extraocular muscle movement. Sclerotherapy is a promising and less invasive alternative treatment for orbital-venous malformation.