Two-in-one: concomitant diffuse large B-cell lymphoma and cavernous haemangioma within the same orbit

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Orbital tumours encompass a wide range of benign and malignant space occupying lesions that may arise primarily from the orbit or have spread from other sites in the body. They are rare with an incidence of 1 in every 100 000 and may lead to devastating complications of which mechanical compression causing optic neuropathy is the most important.[1] Multiple orbital tumours of the same orbit are even rarer with most reported cases being benign homologous tumours such as cavernous haemangiomas or myxofibrosarcomas.[2,3] A 58-year-old ethnic Han Chinese male presented in December 2021 with a 1-month history of right eye proptosis. He had no clinical sign of optic neuropathy. Computed tomography of the orbit revealed right axial proptosis and two separate lesions in the right orbit. One lesion appeared infiltrative and measured 2.2 × 1.5 × 1.7 cm^3 (anteroposterior × transverse × longitudinal) at the extraconal space with retrobulbar extension between the lamina papyracea and the medial rectus. The other was an encapsulated mass with regular border located in the superolateral intraconal region measuring 1.9 × 1.7 × 2.0 cm^3 and abutting the optic nerve. Both lesions enhanced mildly with intravenous contrast (Fig 1). Blood results revealed normal thyroid function and immunoglobulin G4 and white blood cell levels. Based on the distinguishing radiological features of each lesion, we performed a two-stage surgery for theranostic reasons. An incisional biopsy of the medial infiltrative lesion was performed first through an anterior orbitotomy via an upper lid skin crease approach. Frozen section of the medial yellow jelly-like mass revealed atypical lymphoid cells with enlarged vesicular nuclei and amphophilic cytoplasm, highly suspicious of lymphoproliferative malignancy. We then performed a complete excision of the vascular encapsulated intraconal lesion using cryotherapy via a lateral orbitotomy. Formal histopathology reports revealed the first medial infiltrative lesion to be consistent with diffuse large B-cell lymphoma with positive immunostaining for CD20, BCL2, BCL6, MUM1, and CMYC1 (Fig 2). The second intraconal lesion was consistent with cavernous haemangioma (Fig 3).

At 4 months post-surgery there was no clinical sign of optic nerve damage and best-corrected visual acuity was 0.8 in the right eye. Positron emission tomography–computed tomography showed a residual hypermetabolic lesion over the medial

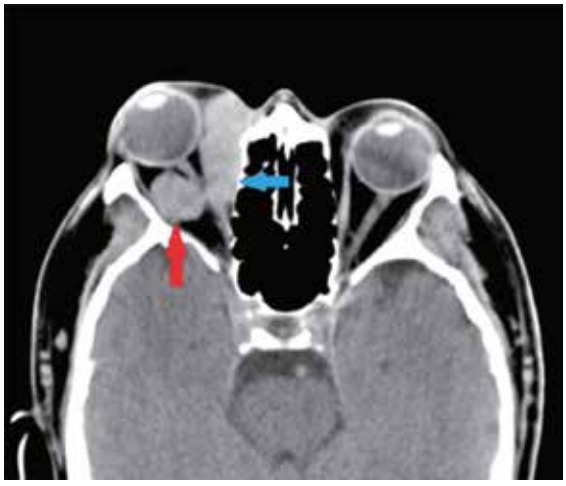

FIG 1. Computed tomography scan with intravenous contrast of the patient showing a cavernous haemangioma (red arrow) and diffuse large B-cell lymphoma (blue arrow)

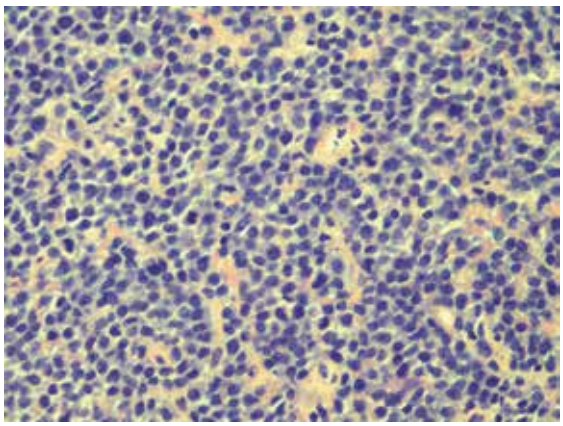

FIG 2. High-power view (×400) of a fresh frozen section with immunostaining of the patient's medial orbital mass showing diffuse sheets of lymphoma cells with focal apoptotic bodies and mitotic figures

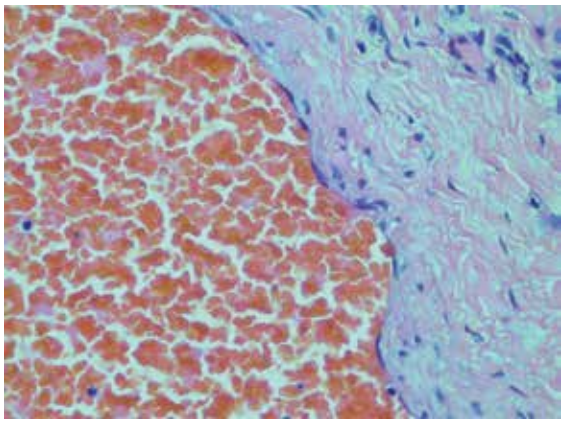

FIG 3. High-power view (×400) of a section from the patient's lateral orbit mass, stained with haemotoxylin and eosin showing dilated congested venous type–looking vessels lined by bland-looking endothelial cells, consistent with cavernous haemangioma

aspect of the right orbit with no extra orbital lesion. The patient is receiving chemotherapy under the care of our haematology team.

Benign multiple homogenous lesions in the same orbit have been reported. Although multiple solitary fibrous tumours of the same orbit without malignant degeneration have been reported,[4] multiple heterogeneous tumours in the same orbit are extremely rare. Ma et al[5] reported a case of concurrent schwannoma and cavernous haemangioma in the same orbit of a 54-year-old female. To the best of our knowledge, concurrent benign and malignant lesions of the same orbit have not been reported in the English literature.

Author contributions

All authors contributed to the concept and design of this study, acquisition of data, analysis of data, drafting the manuscript, and critical revision 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.

Conflicts of interest

The authors have no conflicts of interest to disclose.

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Ethics approval

The patient was treated in accordance with the Declaration of Helsinki and provided informed consent for the treatment and consent for publication.

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