Localized Neurofibromas in the Bilateral Orbits

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Abstract Localized neurofibromas are rare in the orbit and, unlike the more common plexiform neurofibromas, are not typically associated with von Recklinghausen neurofibromatosis. We present a rare case of localized neurofibromas in the bilateral orbits.

Keywords Orbital tumor · Localized neurofibroma · Bilateral

A 34-year-old woman presented with increasing exophthalmos and retro-orbital pain of both eyes over a 2-year period. Ophthalmologic examinations revealed decreased visual acuity in both eyes. She had no symptoms to indicate von Recklinghausen neurofibromatosis. Brain magnetic resonance imaging (MRI) showed retrobulbar extraconal mass lesions with peripheral enhancement in both eyes (Fig. 1).

Fig. 1 Brain magnetic resonance images showing retrobulbar extraconal mass lesions with peripheral enhancement in both eyes (arrows): (a) Coronal T1-weighted image, (b) coronal T2-weighted image, and (c and d) T1-weighted images with gadolinium enhancement images (c, coronal; d, axial; e, sagittal)

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Bilateral tumor resection was performed. A histological examination revealed bilateral localized neurofibromas. The patient's symptoms improved after surgery.

Neurofibromas are categorized into plexiform, localized, and diffuse subtypes. Localized neurofibromas are rare in the orbit and, unlike the more common plexiform neurofibromas, are not typically associated with von Recklinghausen neurofibromatosis [1–3]. Furthermore, bilaterality of localized neurofibromas in the orbits is extremely rare [1, 2]. A complete systemic evaluation in such cases is essential to rule out any associated disorders, including von Recklinghausen neurofibromatosis; however, such an association may not be universal. Several imaging features, including multiplicity,

multilobulation, ring-configured contrast enhancement, and MRI signal intensity heterogeneity, may be diagnostic of localized neurofibromas [3].

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