Case 16230

Eurorad

Primary orbital syringoid eccrine carcinoma

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ISSN: 1563-4086
Section: Head & neck imaging
Area of Interest: Head and neck
Procedure: Imaging sequences
Procedure: Biopsy
Procedure: Contrast agent-intravenous
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: Image manipulation / Reconstruction
Imaging Technique: MR
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 79 years, female

Clinical History:

A 79-year-old woman referred to our Department to investigate a progressive right eye proptosis and diplopia over the last two months. Ophthalmologic examination did not reveal significant findings and a brain CT was scheduled. Her past medical history included bilateral mastectomy for breast cancer.

Imaging Findings:

Contrast-enhanced brain CT showed a homogeneously enhancing solid mass of 3 cm occupying predominantly the medial aspect of the right orbit in the intraconal and extraconal compartment (Fig.1). On subsequent MRI, the mass invaded the retrobulbar fat on the medial side, infiltrating the extrinsic muscles of the right eye (medial rectus muscle and inferior rectus muscle) and surrounding the optic nerve sheath. The mass was iso-hypointense on T1-weighted images (Fig. 2a), hypointense on T2 weighted images (Fig. 2b), hyperintense on fat saturated images (Fig. 2c), with restriction on diffusion-weighted images (Fig. 3) and with intense enhancement after the administration of gadolinium (Fig. 4). The orbital biopsy revealed microcystic adnexal carcinoma with eccrine differentiation containing syringoid cell profiles. Histomorphology and the immunohistochemical profile were compatible with syringoid eccrine carcinoma (Fig. 5-6).

Discussion:

Syringoid eccrine carcinoma is a tumor originating from eccrine sweat glands, included in the group of syringomatous carcinomas [1-3]. It occurs most frequently in the cutaneous and subcutaneous region of the head and neck, often as a slow-growth nodule or plaque on the eyelid (with eyelid entropion or eyelid thickening) or on the central face area. It often demonstrates invasive destructive local growth, with skin and muscle involvement and perineural invasion. It usually occurs in the middle age and in the elderly, with equal gender distribution [4]. In the rare case of orbital extension, exophthalmos, reduced ocular movements and diplopia may occur [3, 4-6]. We described an extremely rare case of syringoid eccrine carcinoma due to its pure orbital presentation with lack of any cutaneous or subcutaneous lesion at the external examination. Syringoid carcinoma is a rare entity that should be

considered after exclusion of other possible diseases in the broad spectrum of orbital masses. A multidisciplinary approach, including ophthalmologic examination and cross-sectional imaging (CT and MRI), is essential in the diagnostic work-up. However, some extremely rare malignant neoplasms, such as syringoid eccrine carcinoma, represent an imaging diagnostic challenge in which final diagnosis should be entitled to histologic examination. To our knowledge, only one similar case of syringomatous carcinoma (microcistic adnexal carcinoma) has been previously reported in literature [7]. The origin of this tumor is still unknown. We supposed it arose from a small orbital dermoid cyst or some microscopic sweat glands. As an alternative, an origin from a developmental malformation containing adnexal structures may be considered.
Our imaging diagnostic work-up included some differential diagnoses. First, we considered a lymphoproliferative disorder, the most common primary orbital tumor in the elderly [8]. However, it is mostly extraconal, it tends to mold orbital structures (globe, optic nerve, orbital wall) and it is usually hyperintense on T2 weighted images at MR imaging [9]. Secondly, we hypothesized an orbital inflammatory pseudotumor, the third most common primary tumor of the orbit and a common cause of unilateral proptosis in adults [10]; however, it is commonly painful, while in our patient the lesion was indolent. Finally, we considered an orbital metastasis from previous breast cancer, although very unlikely considering the extreme rarity of orbital involvement by breast cancer metastasis and the fact that the patient's breast cancer occurred many years before.

Written informed patient consent for publication has been obtained.
Differential Diagnosis List: Syringoid eccrine carcinoma, Lymphoproliferative disorder, Orbital inflammatory pseudotumor, Orbital metastasis

Final Diagnosis: Syringoid eccrine carcinoma

References:

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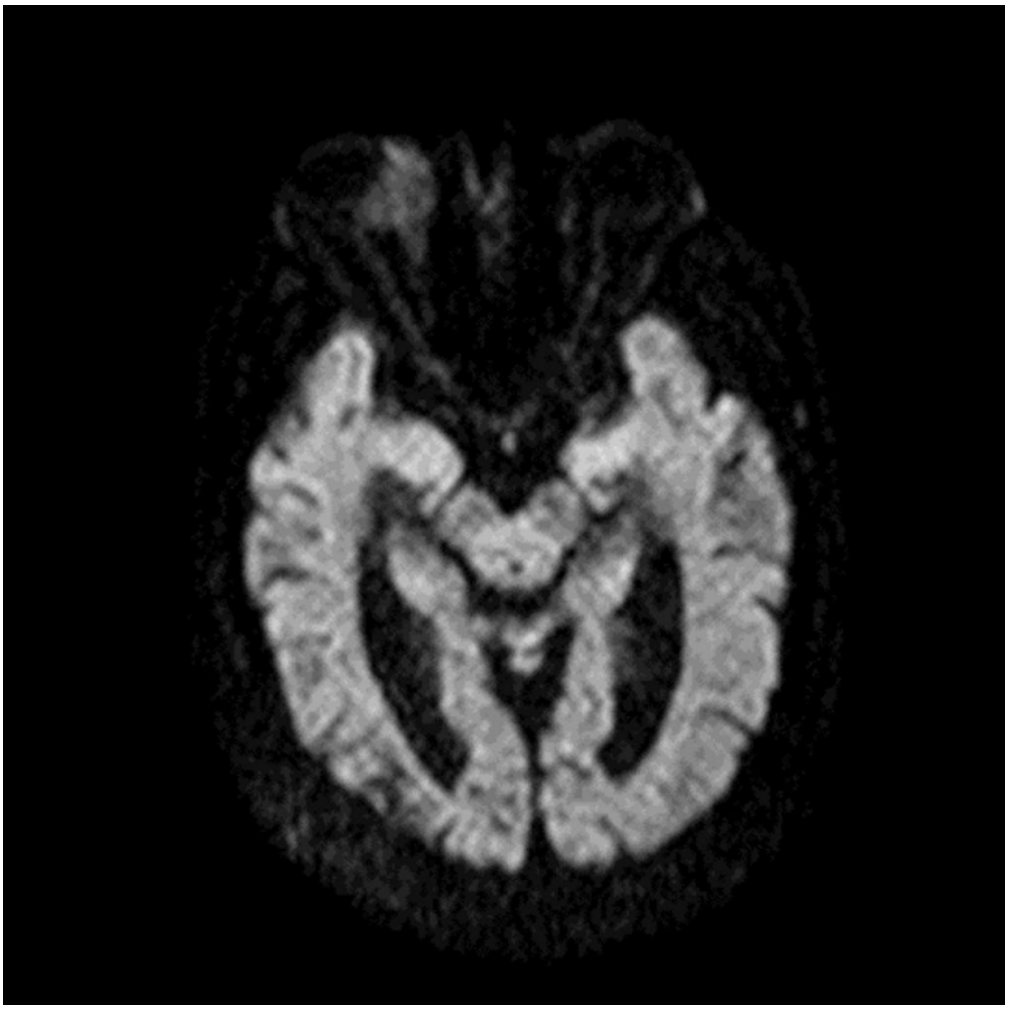
Khalil M, Brownstein S, Codère F, Nicolle D (1980) Eccrine sweat gland carcinoma of the eyelid with orbital involvement. Arch Ophthalmol 98:2210–4 (PMID: 7447777)
Mayer MH, Winton GB, Smith AC, Lupton GP, Parry EL, Shagets FW (1989) Microcystic adnexal carcinoma (sclerosing sweat duct carcinoma). Plast Reconstr Surg 84:970–5 (PMID: 2587662)
Wu-Chen WY, Weng CY, Rajan KD, Eberhart C, Miller NR (2011) Unusual presentation of primary orbital microcystic adnexal carcinoma. J Neuroophthalmol 31:147–50 (PMID: 21368669)
Demirci H, Shields CL, Shields JA, Honavar SG, Mercado GJ, Tovilla JC (2002) Orbital tumors in the older adult population. Ophthalmology 109:243–8 (PMID: 11825802)
Tailor TD, Gupta D, Dalley RW, Keene CD, Anzai Y (2013) Orbital Neoplasms in Adults: Clinical, Radiologic, and Pathologic Review. RadioGraphics 33:1739–1758 (PMID: 24108560)
Harr DL, Quencer RM, Abrams GW (1982) Computed tomography and ultrasound in the evaluation of orbital infection and pseudotumor. Radiology 142:395–401 (PMID: 7054828)

Figure 1

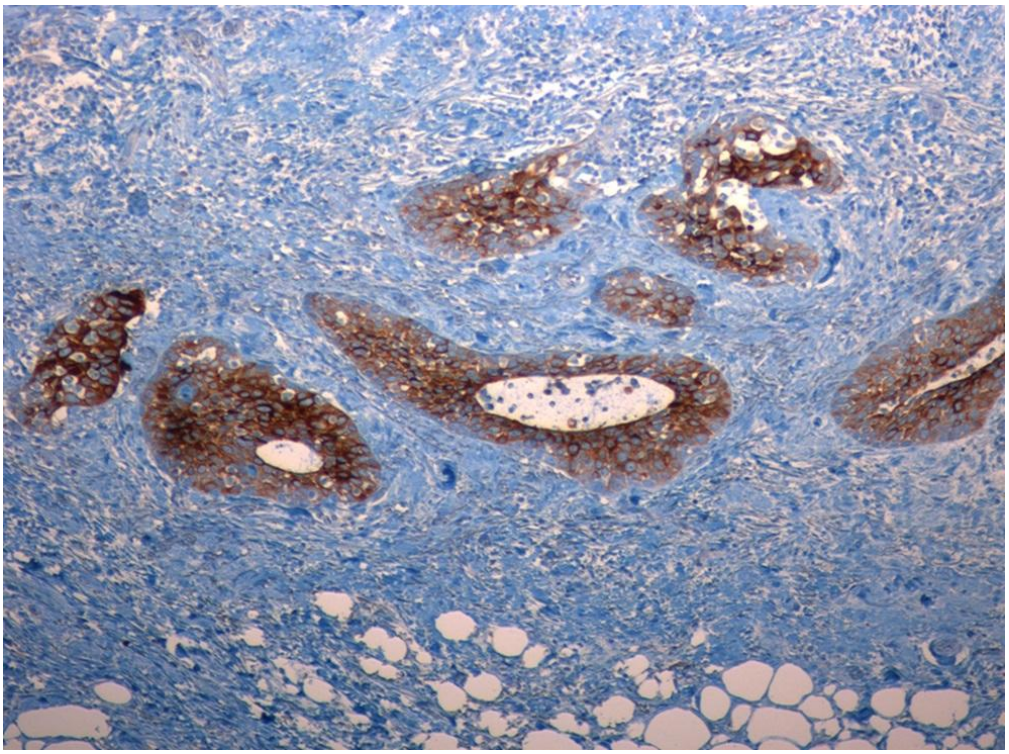
a



Description: Diffusion-weighted imaging shows mild diffusion restriction of the lesion **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy

Figure 2

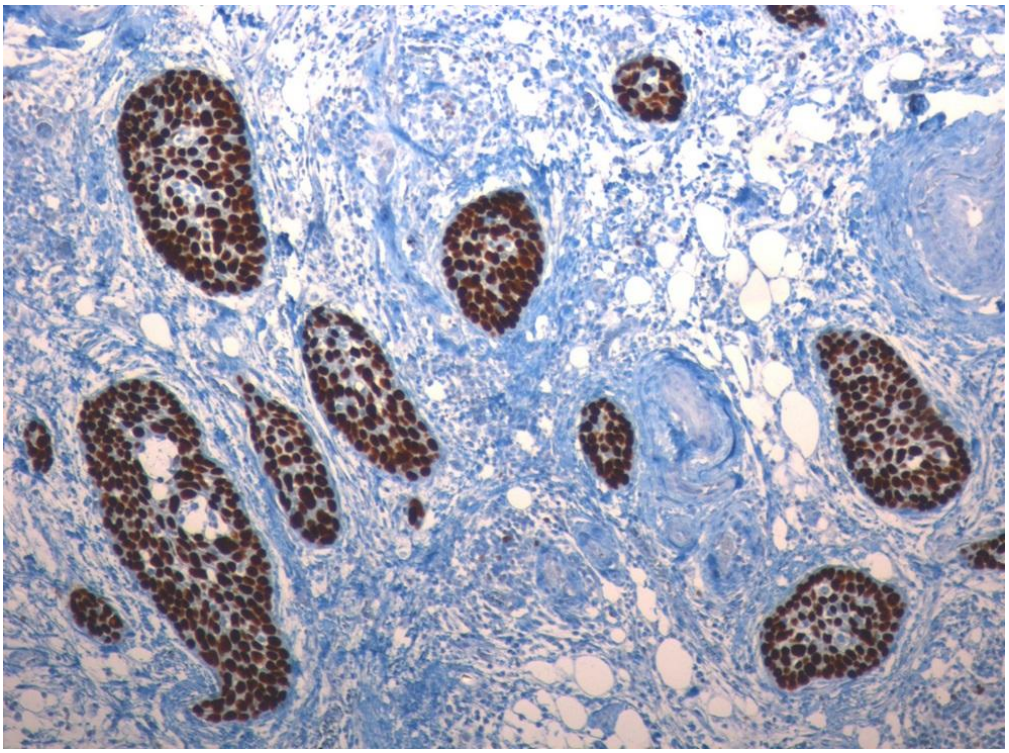
a



Description: EMA was diffusely positive in the neoplastic cells (magnification x10) **Origin:** Department of Pathology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy

Figure 3

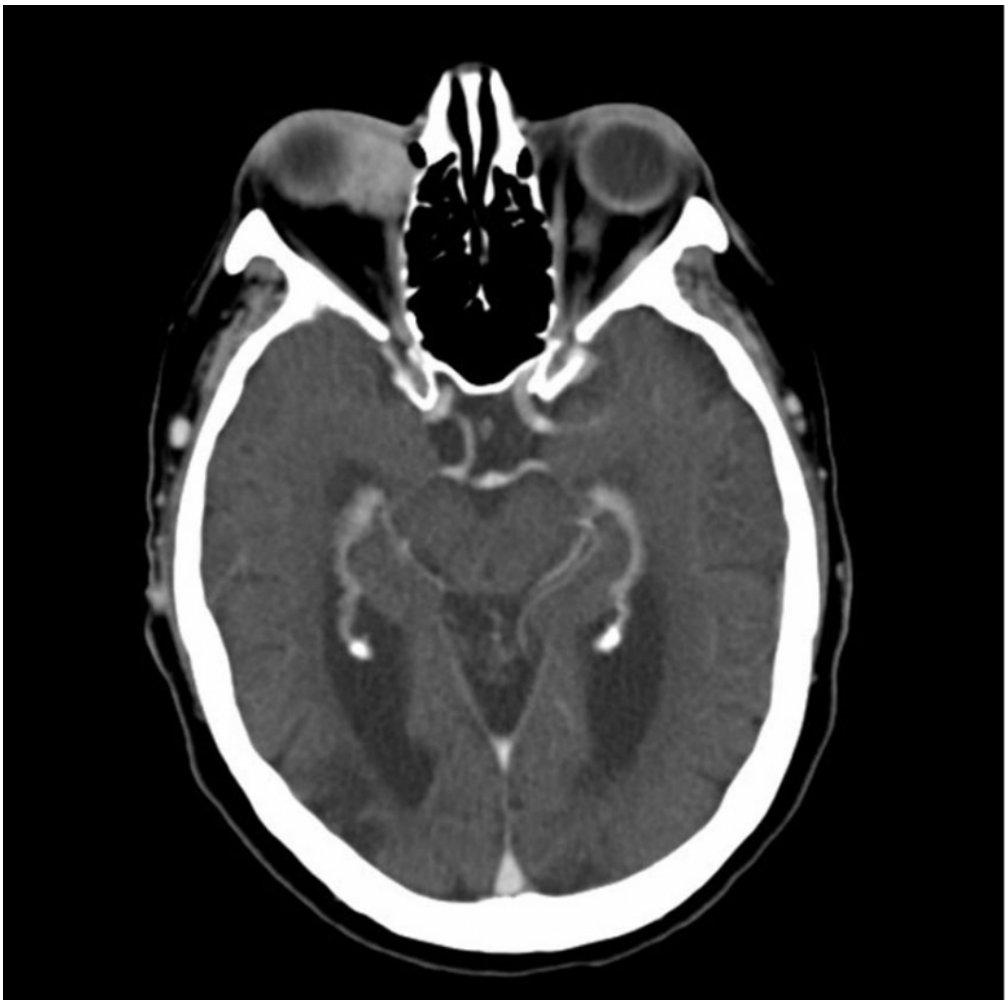
a



Description: The immunohistochemistry marker p63 stained positively in the lesion (magnification x10)
Origin: Department of Pathology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy

Figure 4

a



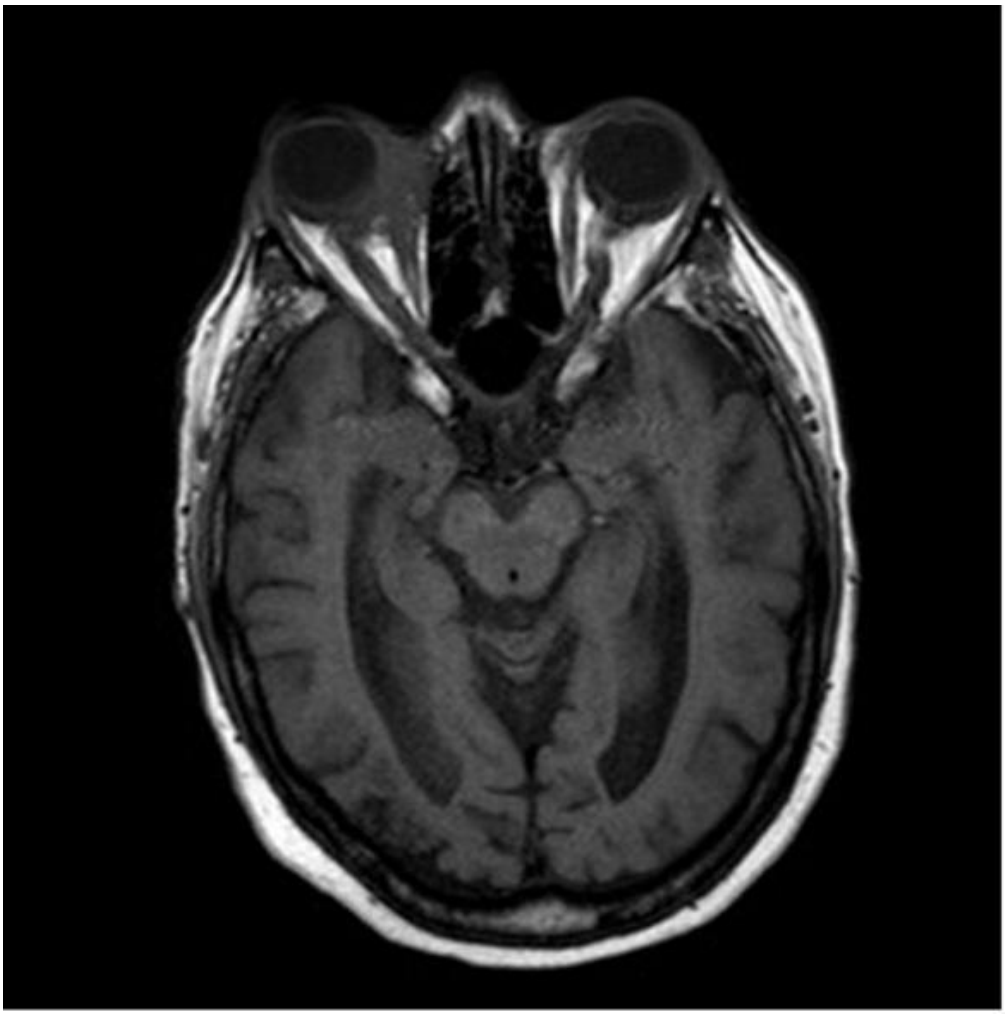
Description: CT (axial plane) shows an intra-extraconal enhancing solid mass located in the medial aspect of the right orbit **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

b

Description: CT (coronal plane). The enhancing mass is located in the medial aspect of the right orbit
Origin: Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

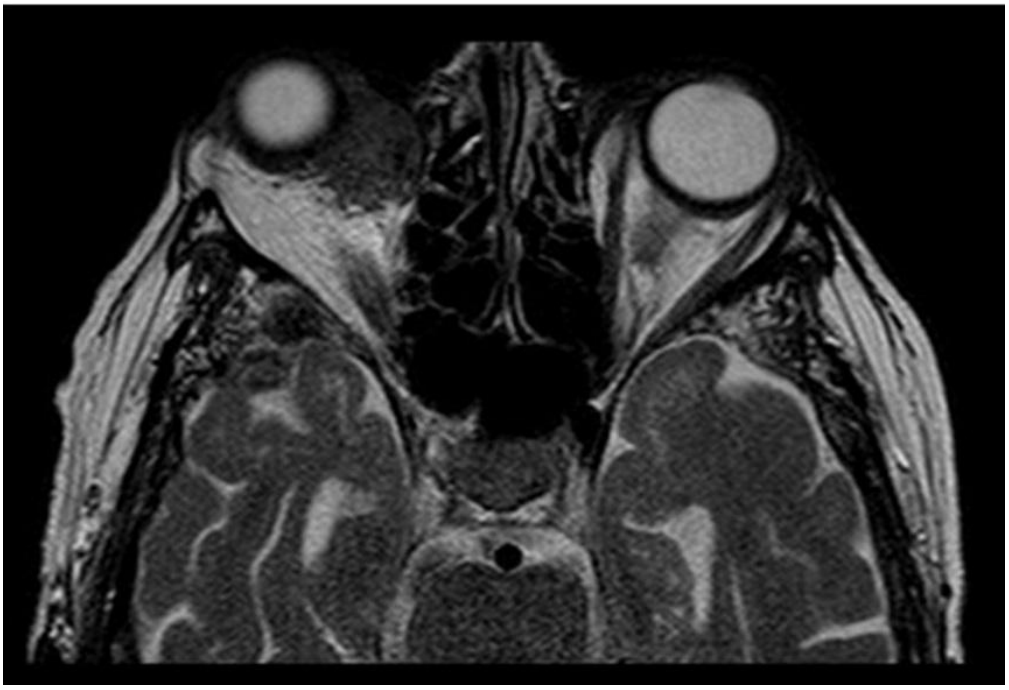
Figure 5

a



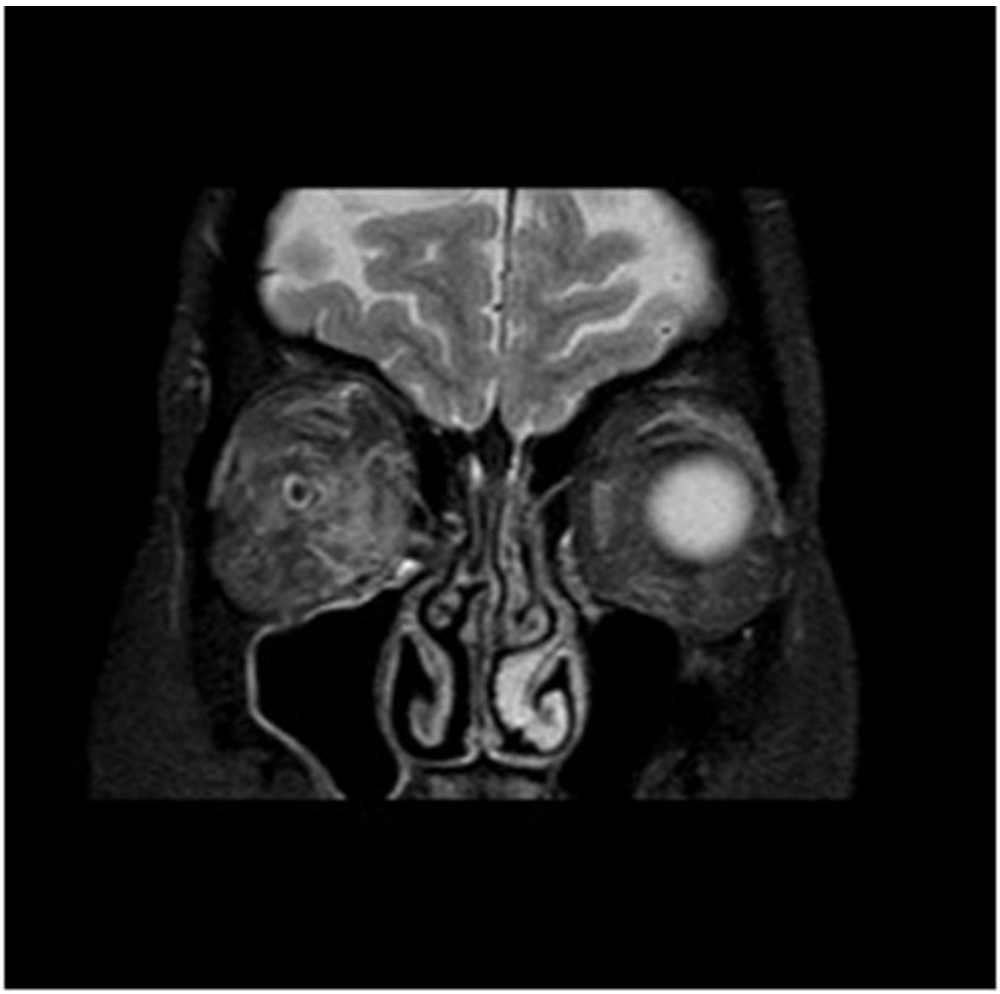
Description: The lesion is iso-hypointense on T1-weighted images. **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

b



Description: The lesion is hypointense on T2 weighted images **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

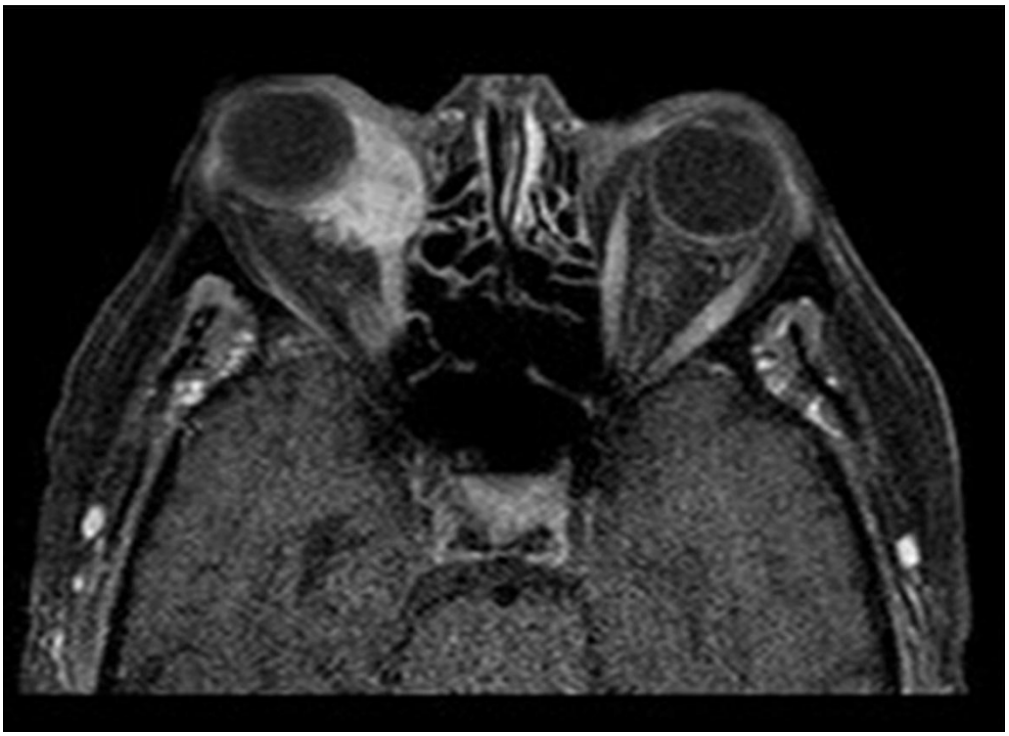
c



Description: The lesion is hyperintense on fat saturated images **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

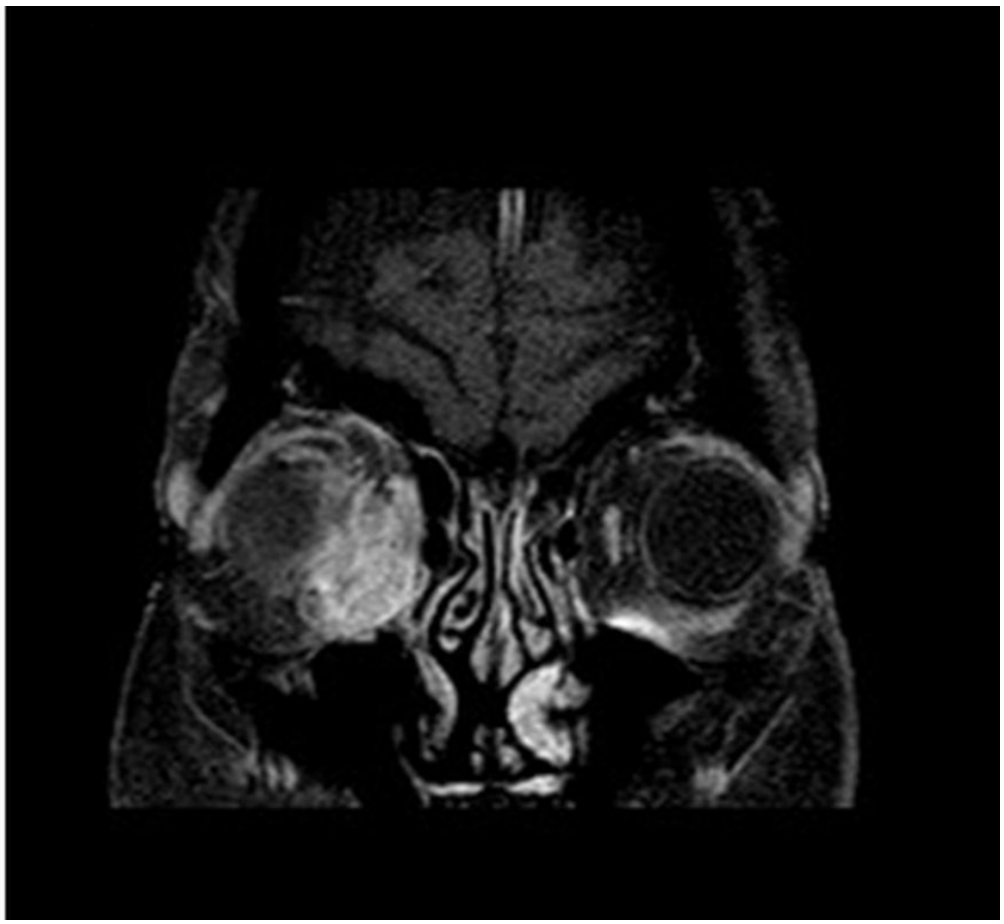
Figure 6

a



Description: Fat-suppressed axial T1-weighted image after the administration of Gadolinium shows intense enhancement of the mass **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.

b



Description: Fat-suppressed coronal T1-weighted image after the administration of Gadolinium shows intense enhancement of the mass **Origin:** Department of Radiology, Azienda Sanitaria Universitaria Integrata di Trieste, Trieste, Italy.