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Intraosseous Meningioma: A Rare Entity with Multiple Clinical and Radiological Faces

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Abstract | **Case Report**

Primary intraosseous meningiomas are rare ectopic meningiomas, accounting for 1-2% of all meningiomas. The aim of this work is to illustrate the different imaging aspects contributing to a diagnostic approach and to discuss their various differential diagnoses. We report a case of an ethmoido-orbital intraosseous meningioma, documented by CT and MRI.

Keywords: Intraosseous Meningiomas, CT, MRI.

INTRODUCTION

Encephalic meningiomas account for 20% of primary intracerebral tumors and usually considered benign lesions. Intraosseous localizations are rare, accounting for only 1-2% of all intracranial meningiomas. We report a case of ethmoido-orbital intraosseous meningioma documented by CT and MRI.

PATIENT AND OBSERVATION

Observation: A 61-year-old woman with hypertension presented with exophthalmos and right periorbital swelling for 1 year (**Figure 1)**.

Figure 1 : Right orbital exophthalmos with periorbital swelling

A CT scan revealed a bony lesion process on the upper and inner orbital walls of the right orbit, with endo-orbital and retroocular development, of condensing matrix with cortico-spongy dedifferentiation, some local osteolytic lesions associated with discrete thickening and meningeal enhancement opposite the condensed bone.

MRI confirms the intraosseous lesion process, with a dual component, a predominantly frank T2.T1 and T2 hyposignal portion on all sequences, and a T1 iso

signal and moderate T2 hyposignal portion. The latter enhances after gadolinium injection. There also discrete thickening and enhancement of the adjacent meninges after injection of Gadolinium.

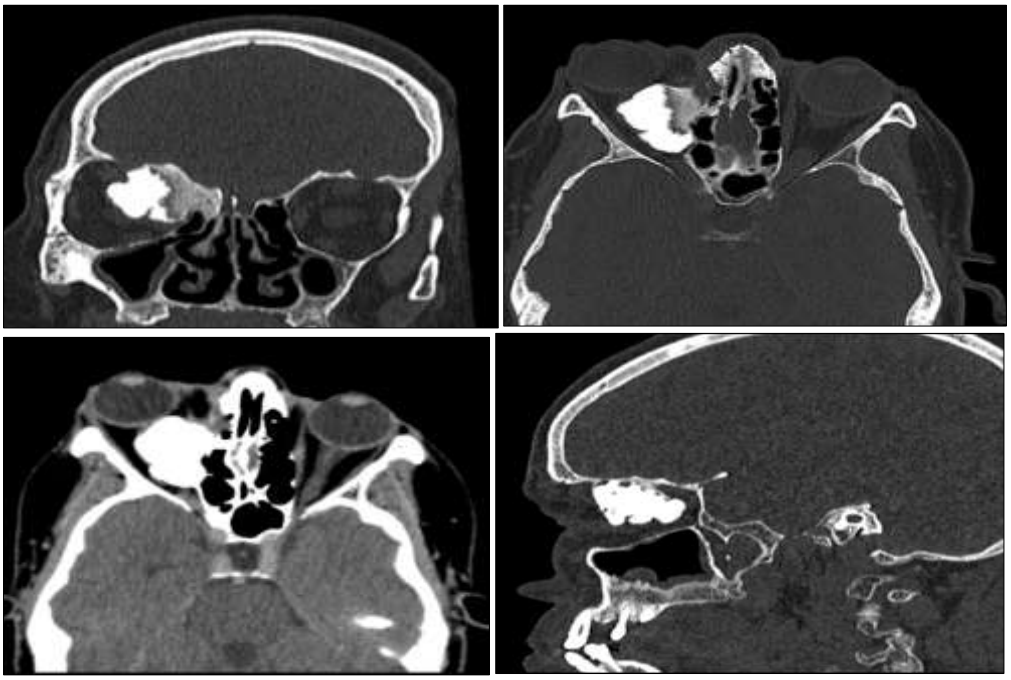

Figure 2: CT scan of the orbit in axial, sagittal and coronal sections

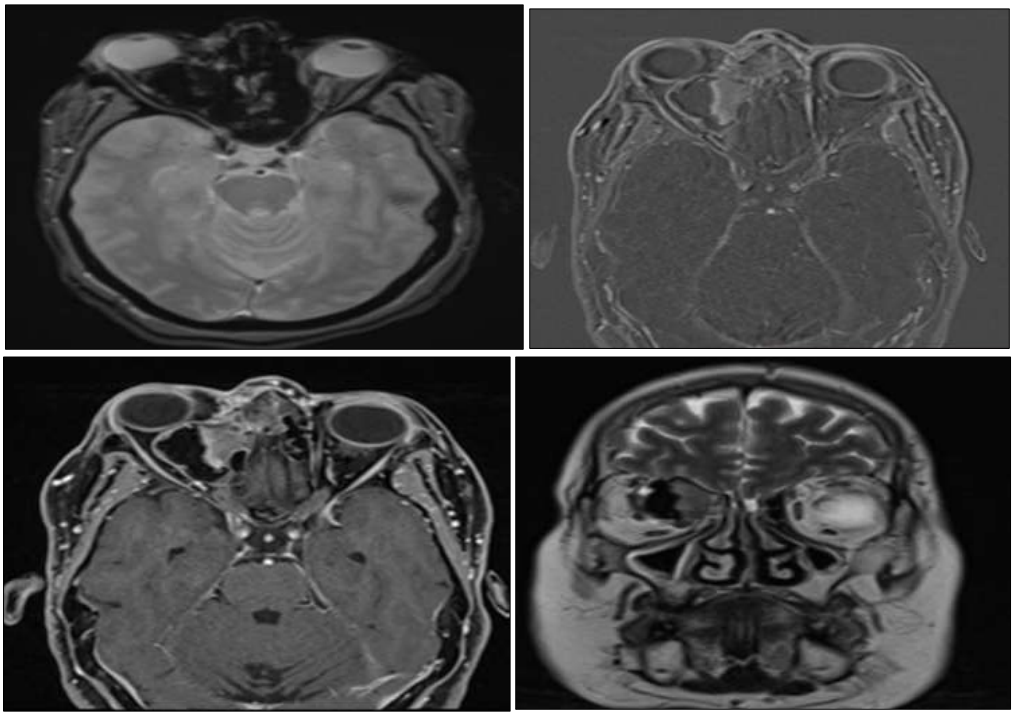

Figure 3: axial T2 and coronal T2 sequences, axial T1 with gadolinium injection/subtraction

This process pushes the optic nerve downwards and outwards, causing it to appear laminated, and the right eyeball forward, resulting in Grade II exophthalmos. It also comes into contact with the rectus obliquus, superior rectus and medial rectus muscles.

DISCUSSION

Meningiomas are the most common primary intracranial non-glial tumors, most often located in the subdural space [1]. Ectopic meningiomas can be found in the facial sinuses, nasal cavity, orbits, salivary glands, nasopharynx and cranial vault. Intraosseous primary meningiomas are rare and constitute a distinct entity.

Their first case was described by Winkler in 1904, with the convexity and base of the skull being the most common locations [2].

Pathophysiology

Their pathophysiology is poorly understood, and several hypotheses have been put forward: Sequestration of arachnoid cells in fracture lines after head trauma [2]. Proliferation of ectopic arachnoid cells. Proliferation of extra-dural arachnoid cell remnants located at cranial sutures during childbirth and cranial remodeling. Development from pluripotent mesenchymal cells or from metaplasia of mesenchymal cells such as fibroblasts [3].

Clinical:

Clinical symptoms are not specific. Manifestations vary according to the site of the lesion, its size and its relationship with neighbouring organs [2]. Most often, these lesions manifest as a fixed, hard, painless swelling. Neurological examination is normal, apart from intermittent headaches. In the case of tumors extending to the inner table and causing a mass effect on the cerebral parenchyma, neurological signs may be observed. In temporo-spheno-orbital forms, the clinical picture is dominated by slowly progressive, axial, painless exophthalmos associated with swelling of the homolateral temporal fossa. Decline in visual acuity is variable [1, 2].

Means of Exploration

Standard Radiography:

Intraosseous meningiomas can take two forms: the hyperostotic form is the most frequent, with thickening of the vault, sometimes bristling with tangential bone spicules, and the osteolytic or erosive form, with bone lysis with or without reconstruction. Intratumoral calcifications may occur in 20% of cases.

CT Examination:

Reveals the meningioma, whose tissue density is usually greater than that of the cortex. In the hyperostotic form, a CT scan reveals the squat, regularly arranged appearance of the spicules, which differs from the fine, anarchic appearance of a malignant tumor.

Magnetic Resonance Imaging:

Provides better anatomical definition by showing its various components. The tumor shows a marked hyposignal on T1- and T2-weighted sequences. Intravenous injection of gadolinium leads to contrast enhancement within the intra-osseous component and in the meningeal envelopes opposite. This enables a meningeal extension assessment that is often underestimated by CT scans, due to the low differential contrast between bone and enhanced meninges, thus avoiding recurrence by partial excision. MRI can also detect intracranial extension [4].

Technetium scintigraphy: shows hyperfixation in the majority of cases [5].

Differential Diagnosis:

In hyperostotic forms, they are discussed with sclerotic hemangiomas (well-limited, trabecular honeycomb lesion, periosteal reaction, heterogeneous T1 hypersignal on MRI), fibrous dysplasia (ground-glass appearance, sharp contours), osteoma (regular, sharp contours), osteoblastic metastases and Paget's disease. In lytic forms, the differential diagnosis may involve osteolytic metastases, osteosarcoma (fine, anarchic spicules, whereas in MIO they are squat and regular) and myelomatous lacunae. In young subjects, eosinophilic granulomas and epidermoid cysts should also be discussed [1, 2].

Treatment:

Treatment is surgical, involving excision to establish the histological diagnosis and avoid neurological complications. Complementary radiotherapy may be recommended in cases of incomplete excision [6].

CONCLUSION

Intraosseous meningiomas are rare, benign ectopic tumors with a progressive course. Their imaging appearance is characteristic, with a very good topographical study. The diagnosis of certainty is histological.

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