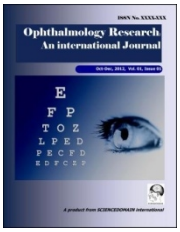

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Presumed, Adult-onset Eosinophilic Granuloma of the Orbital Roof Showing Spontaneous Resolution

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Authors' contributions

This work was carried out in collaboration between both authors. Authors FB and AP designed and conducted the study, collected, analyzed, and interpreted the data. Author AP managed the literature search and wrote the first draft of the manuscript. Both authors read and approved the final manuscript.

Case Study

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ABSTRACT

Aims: To describe an unusual case of presumed, adult-onset eosinophilic granuloma of the orbital roof, which showed spontaneous resolution.
Presentation of Case: A 42-year-old man presented with a three-month history of relapsing oedema in his right upper eyelid. Slit-lamp examination of both eyes was unremarkable, ocular motility was normal, and there was no proptosis. CT and MRI head scans disclosed a 15mm solid mass in the right anterior cranial fossa, which had eroded the orbital roof and penetrated into the orbit. An initial diagnosis of orbital intradiploic meningioma was made. Total resection of the lesion was planned and the patient was put on the waiting list for neurosurgery. One year later, preoperative MRI disclosed a significant regression of the lesion. Six months later, CT and MRI showed complete disappearance of the lesion and full re-ossification of the orbital roof. A presumptive, final diagnosis of eosinophilic granuloma was made and the patient was cancelled from the waiting list for neurosurgery. There have been no MRI changes in the last 24 months.
Discussion: In this report, initial radiological images were apparently consistent with an orbital intradiploic meningioma. However, this diagnosis turned out to be wrong, because spontaneous resolution of adult-onset orbital intradiploic meningioma has never been reported. Conversely, orbital eosinophilic granuloma may heal without treatment, even

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though its occurrence in adults is exceptional.
Conclusion: Upper eyelid oedema may be the presenting sign of an osteolytic intracranial mass eroding the orbital roof and extending into the orbit. Radiological differential diagnosis may be difficult. In adults, the detection of an osteolytic lesion and its subsequent spontaneous resolution may be suggestive of an eosinophilic granuloma.

Keywords: Upper eyelid oedema; presumed eosinophilic granuloma; orbital roof; CT; RMI; adult onset; spontaneous resolution.

1. INTRODUCTION

Relapsing upper eyelid oedema without proptosis may be the first sign of an orbital mass originating from the orbital roof, such as a meningioma or granuloma. We describe herein an unusual case of presumed, adult-onset eosinophilic granuloma of the orbital roof, which showed spontaneous resolution.

2. PRESENTATION OF CASE

A 42-year-old man presented at the Section of Ophthalmology, Department of Surgical, Microsurgical, and Medical Sciences, University of Sassari, Sassari, Italy, with a three-month history of relapsing oedema in his right upper eyelid. There was no history of previous head trauma. Visual acuity was 6/6 bilaterally. Slit-lamp examination of both eyes was unremarkable. Ocular motility was normal and there was no proptosis. Blood examination revealed no pathological findings, with no increase in leukocytes and eosinophils or in the erythrocyte sedimentation rate. CT and MRI head scans disclosed a 15mm solid mass in the right anterior cranial fossa, which had eroded the orbital roof, penetrated into the orbit, and caused mild displacement of the levator palpebrae and superior rectus muscles (Figs. 1 and 2). An initial diagnosis of presumed intradiploic meningioma of the orbital roof was made. Two different neuro-surgical units in Italy agreed with this diagnosis. As a result, total resection of the lesion was planned and the patient was put on the waiting list for neurosurgery. One year later, preoperative MRI head scans disclosed a significant regression of the lesion; consequently, neurosurgery was postponed. Six months later, CT and MRI scans showed complete disappearance of the solid mass and full re-ossification of the orbital roof (Fig. 3). A presumptive, final diagnosis of solitary eosinophilic granuloma of the orbital roof was made. Systemic investigation excluded involvement of other organs. He was cancelled from the waiting list for neurosurgery and MRI scans at 6-month intervals have been scheduled. There have been no MRI changes in the last 24 months.

Approval from the Ethics Committee/Institutional Review Board of the Department of Surgical, Microsurgical, and Medical Sciences, University of Sassari, Sassari, Italy, was obtained. The patient received detailed information and provided written informed consent.

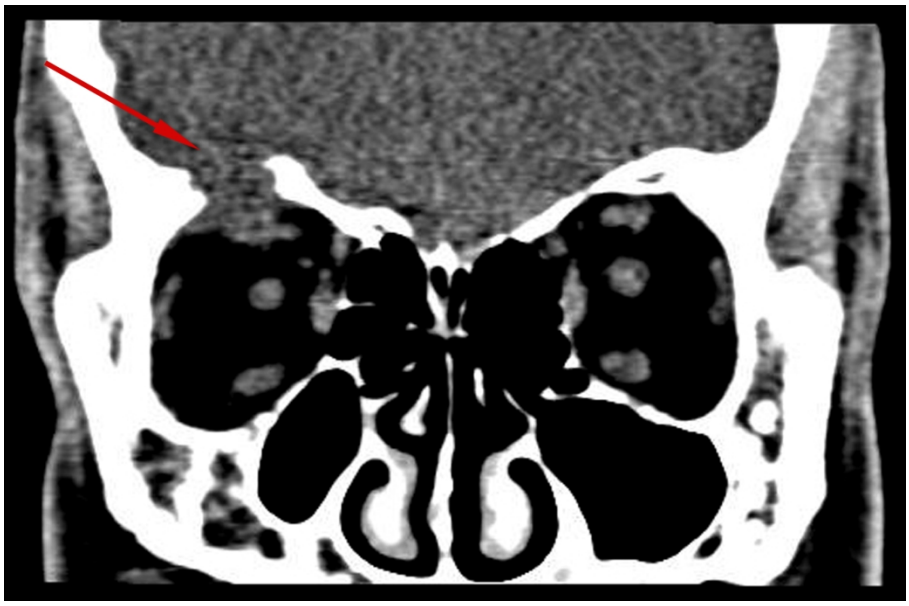

Fig. 1. Computed tomography (initial examination): coronal scan of the head showing a solid mass eroding the right orbital roof (arrow)

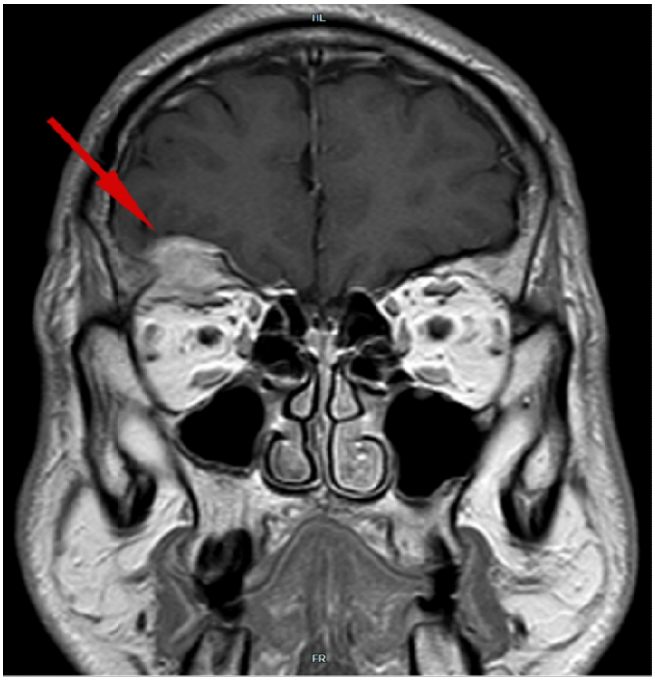

Fig. 2. Magnetic resonance imaging (initial examination): coronal head scan with gadolinium showing a solid mass in the right orbital roof extending into the orbit (arrow)

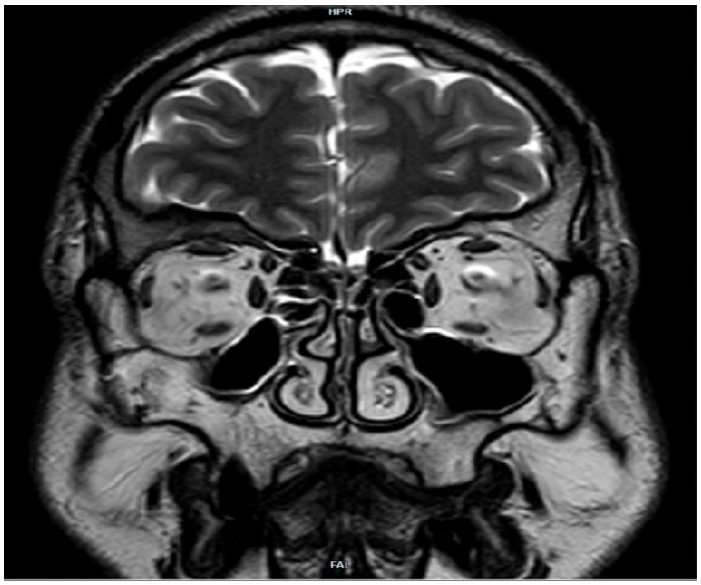

Fig. 3. Magnetic resonance imaging (18 months later): coronal T2-weighed head scan showing complete disappearance of the solid mass

3. DISCUSSION

Meningiomas arising from sites different from the meninges are termed as ectopic or extradural meningiomas. Intraosseus meningiomas usually originate from the diploe of the calvarial bone at the suture lines and often have osteoblastic activity. They have been postulated to originate from arachnoidal cell nests captured in extradural tissues during embryonal development or after head trauma with fracture and dural tear [1]. Intradiploic meningiomas of the orbital roof are exceedingly rare [1-6]. They usually occur during the second decade of life and present with proptosis on the side of the tumor, due the tumor's extension into the orbit. Coronal and lateral CT and MRI scans are necessary to demonstrate clearly the intradiploic involvement of the orbital roof. Treatment consists of total resection of the lesion with reconstruction of the orbital roof by autogenous split calvarial bone graft [2,3].

Eosinophilic granuloma of the orbit is a subtype of Langerhans cell histiocytosis, an idiopathic reticuloendothelial proliferative disorder with clonal proliferative Langerhans cells. Isolated eosinophilic granuloma of the orbit is uncommon, with onset in the first or second decade, but predominantly among 2- to 5-years-olds [7]. Symptoms include rapidly progressive upper eyelid oedema and erythema, bone pain, and tenderness. The blood tests show an increase in leucocytes and eosinophils in approximately 7% of cases [8]. The bones of the lateral orbital roof are usually affected, with osteolytic lesions with regular or irregular borders. CT and MRI show the exact size and borders of the tumour, which usually appears as an isointense mass on T1-weighted images and a high-intensity mass on T2-weighted images. The definitive diagnosis is made by histological examination. All patients need systemic investigation to exclude involvement of other organs. This tumour, as well as orbital Langerhans cell histiocytosis [8], may heal without treatment, but recurrence may occur during the first year after diagnosis. Thus, even after surgical curettage a careful follow-up

must be conducted. If a secondary lesion appears, then chemotherapy or further excision has to be done [9].

In the case reported here, CT and MRI images were initially believed to be consistent with an intradiploic meningioma of the orbital roof. However, luckily for the patient, delays in neurosurgery showed that this initial diagnosis was most likely wrong. Indeed, to our knowledge, spontaneous resolution of adult-onset intradiploic meningioma of the orbital roof has never been reported. Conversely, eosinophilic granuloma of the orbit may heal without treatment, even though its occurrence in adult patients is exceptional. On the whole, the clinical course of the orbital lesion observed in our patient seems to be much more in favour of an eosinophilic granuloma rather than an intradiploic meningioma. Of course, histological evaluation would be essential to establish the right diagnosis; however, whatever the lesion's aetiology, our report clearly demonstrates that, in this case, a relatively large mass arising from the orbital roof showed gradual, spontaneous resolution in an 18-month period, thus sparing the patient a major operation.

4. CONCLUSION

Upper eyelid oedema may be the presenting sign of an osteolytic intracranial mass eroding the orbital roof and extending into the orbit. Radiological differential diagnosis may be difficult. In adults, the detection of an osteolytic lesion and its subsequent spontaneous resolution may be suggestive of an eosinophilic granuloma.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that this study was approved by the appropriate ethics committee and was therefore performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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