An unusual case of extranodal Rosai-Dorfmann disease presenting as simultaneous orbital and intracranial masses

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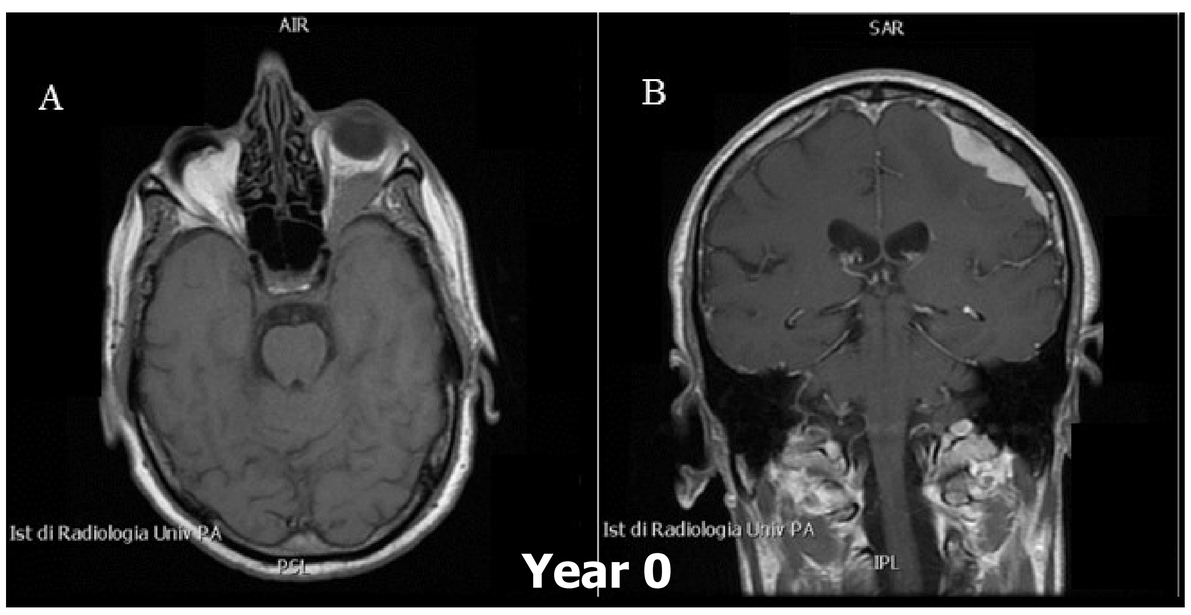
OBJECTIVES: To describe an unusual case of an old patient with extranodal Rosai-Dorfmann disease with orbital and intracranial involvement, poor response to steroid therapy, but good response to mercaptopurine.

INTRODUCTION: Rosai-Dorfmann-Disease (RDD) is a rare benign lymphoproliferative disorder, of unknown aetiology commonly presenting as painless, massive cervical lymphadenopathy with fever, weight loss and polyclonal hypergammaglobulinemia. The disease mainly affects children and young adults. Over 90% of patients present with cervical lymphadenopathy, and extranodal involvement (including paranasal sinuses, respiratory tract, skin, nose and bone) occurs in 40% of cases. Ophthalmic involvement is seen in 10% of cases. These include eyelid and orbital mass, and rarely uveitis. Rosai-Dorman disease could mimic lymphoma, histiocytic and lacrimal gland tumors. Localisation in the central nervous system (CNS) is rare (4% of cases). Various treatment have been proposed, including steroid therapy, chemotherapeutic regimens, radiotherapy, surgery, and combinations of the above but optimal treatment has yet to be established.

CASE REPORT: A 70-years-old man, with blindness in the right eye from infancy, was referred to our hospital for a history of right upper limb focal seizures and paraesthesia followed by a recent onset of episodes with secondary generalization, loss of consciousness and amnesia about the event. The clinical examination revealed a slight right hemiparesis.
An MRI revealed (fig 1 A-B):

- **in the orbital region:** an isointense enhancing mass with dislocation of the optic nerve
- **in the left fronto-parietal-temporal region**: an extra-axial thickening of the dura mater with enhancement and perilesional oedema, infiltrating the spheno-orbital fissure.



Year 0

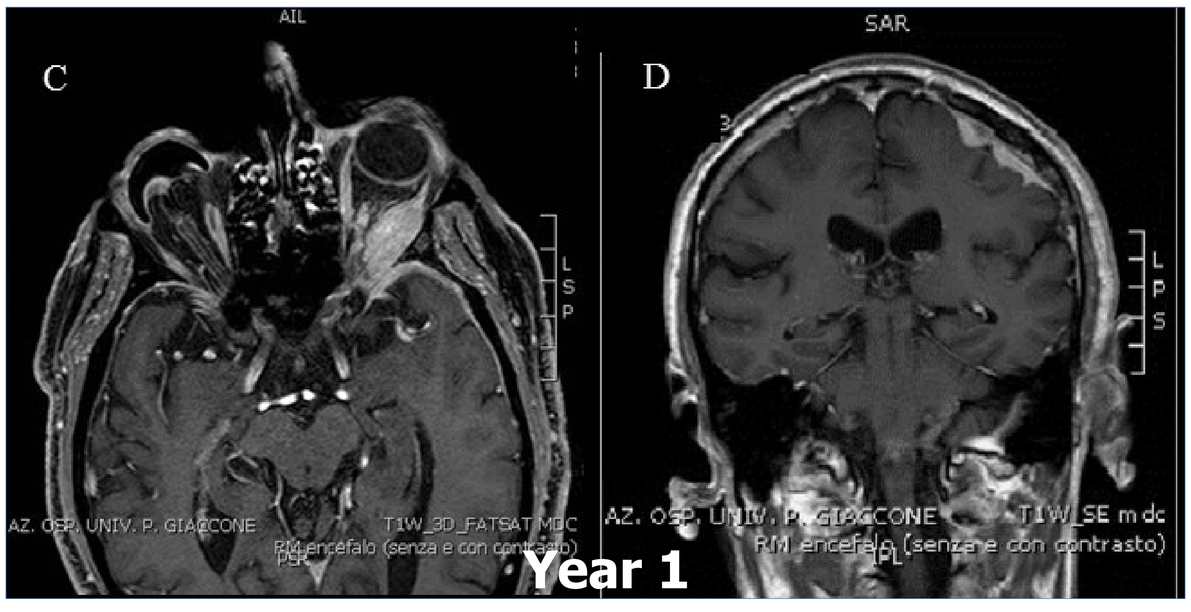
Histopathological evaluation: a fibrotic tissue with a histiocytic reaction and a large number of macrophage foam cells with intracytoplasmic lymphocytes (emperipolesis).

Immunohistochemistry: positive for S-100 protein and CD68 leucocyte antigens, and negative for CD1a.

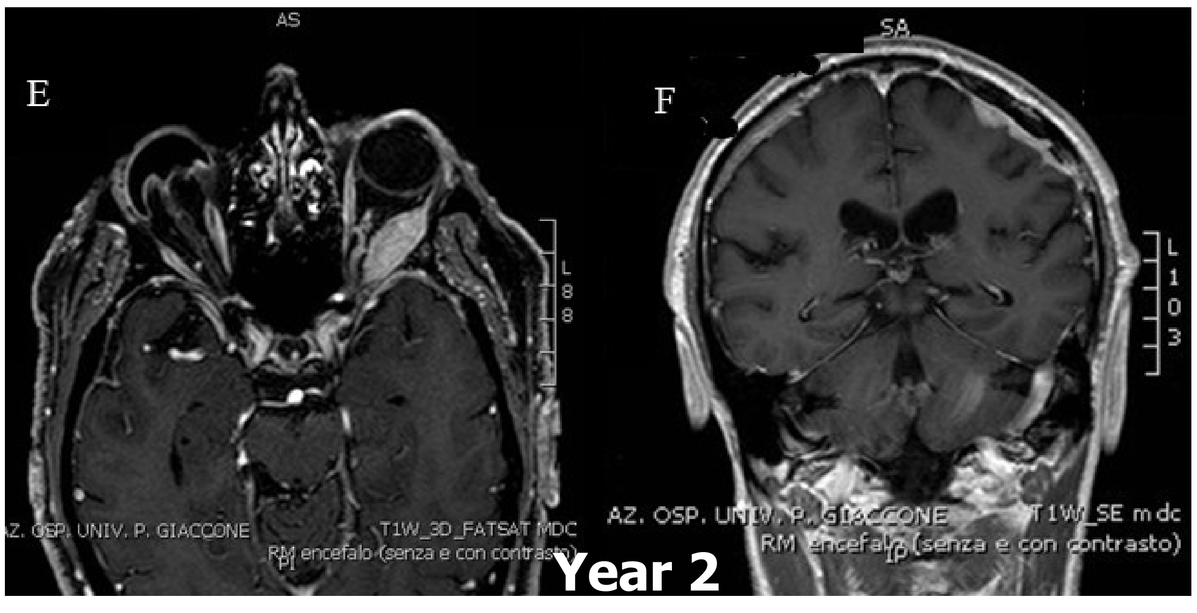
According to published criteria, these findings were consistent with a **diagnosis of Rosai-Dorfman disease (RDD)**.

The patient refused the surgical debulking to salvage his vision.

Treatment with levetiracetam (500 mg twice a day) and steroid (Desametasone 4 mg ev once a day) was started with remission of critical symptoms. Three months later, a therapy with mercaptopurine was started with a daily dosage of 2,5 mg/kg.



Year 1



Year 2

No seizure activity without further loss of vision.

MRI showed slight reduction of the parietal-temporal lesion (FIG. 2), and levetiracetam therapy was stopped

No seizure activity and further loss of vision.
MRI showed a further reduction of the parietal-temporal lesion and of the left intra-orbital mass that enhanced uniformly after gadolinium administration (FIG. 3).

DISCUSSION AND CONCLUSIONS

- ❖Very few cases of extranodal RDD with multiple CNS lesions involving the orbital region have been described.
- ❖Recognizing RDD is important because RDD is a rare clinical entity, that should be considered in the differential diagnosis of meningioma and other space occupying lesion of the brain.
- ❖The correct diagnosis primarily relies on histological and immunohistochemical characterization.
- ❖The treatment of the disease is controversial: surgery, steroids, chemo and radio therapy have been proposed.
- ❖Surgery represents the best chance of cure for patients with orbital and neurological manifestations as the sole extranodal site of involvement without synchronous nodal disease.
- ❖The case reported would suggest the use of 6-mercaptopurine for treatment of this form of RDD

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