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Case Report

Rosai Dorfman Disease of the Orbit

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Abstract

Introduction: Rosai-Dorfman disease or Sinus Histiocytosis with massive lymphadenopathy (SHML) is a rare, benign and self-limited histiocytic proliferative disorder which affects mainly lymph nodes and very rarely extranodal sites.

Presentation of case: A 60 year old lady with SHML of orbit is described. She underwent excision of the mass, received steroids for 6 weeks and is in remission at 2 years.

Conclusion: Though the orbit is a rare site for SHML, the disease should be considered in the differential diagnosis of orbital swellings.

Keywords: SHML; Rosai Dorfman; Orbit
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Introduction

Rosai-Dorfman disease or Sinus Histiocytosis with massive lymphadenopathy (SHML) is a rare, benign and self-limited histiocytic proliferative disorder which affects mainly lymph nodes and very rarely extranodal sites. We present the case of a lady with SHML involving the orbit.

Presentation of case

A 60 year old lady presented to us with blurring of

vision of right eye, diplopia and a swelling in the superotemporal part of right orbit. Computerised tomogram (CT scan) of orbits showed a well defined hyperdense soft tissue lesion in the superior aspect of right orbit and involving the lacrimal gland (Figure 1). She did not have disease elsewhere. She underwent lateral orbitotomy and excision biopsy. Histopathology showed sheets of histiocytes with sinusoidal pattern showing emperipolesis (Figure 2) and were positive for CD68 and S100 (Figure 3, and 4). The diagnosis was SHML or Rosai Dorfman disease. She received steroids for 6 weeks and is asymptomatic at 2 years.

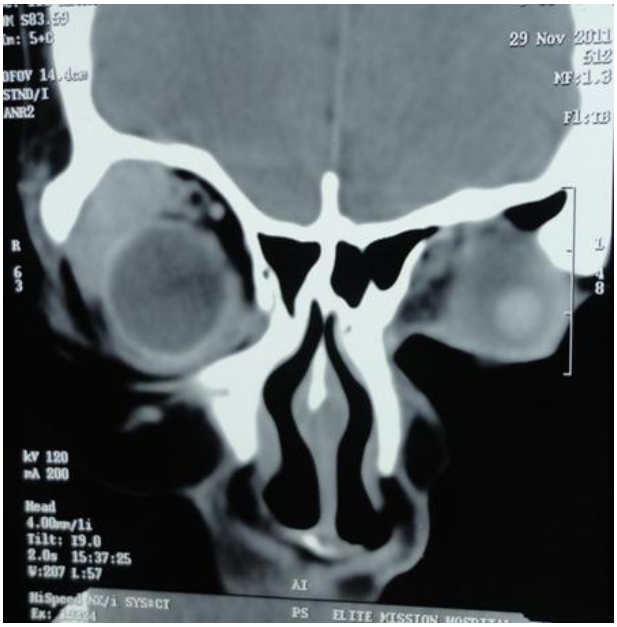

Figure 1 Coronal CT image of orbits showing a well defined soft tissue lesion in the superior aspect of right orbit

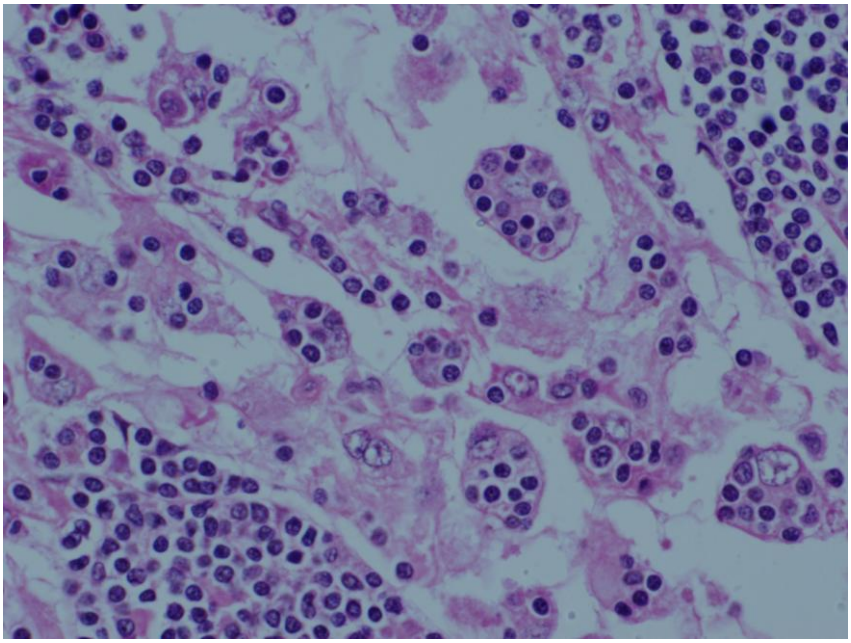

Figure 2 Histiocytes showig emperipolesis (H&E x400)

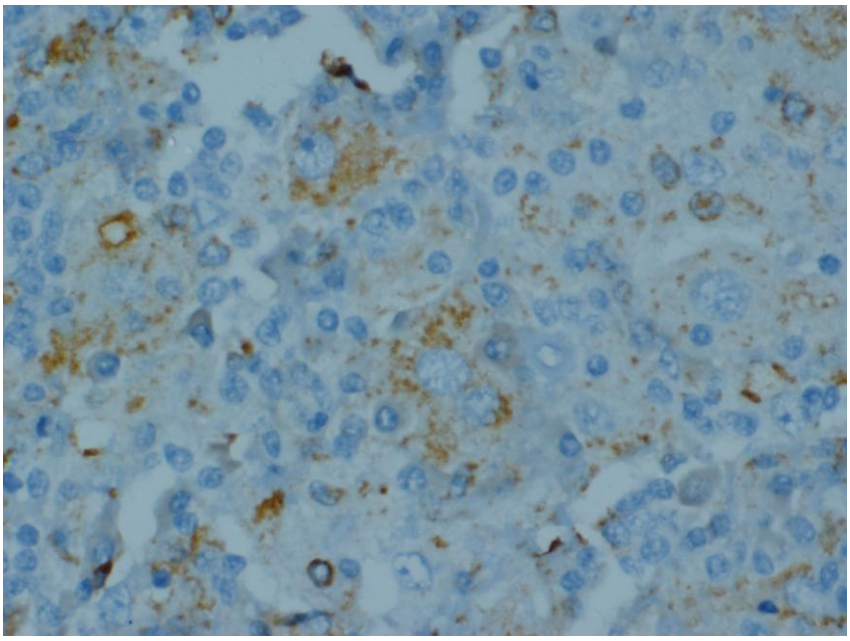

Figure 3 Histiocytes are CD68 (KP-1) positive (IHCx400)

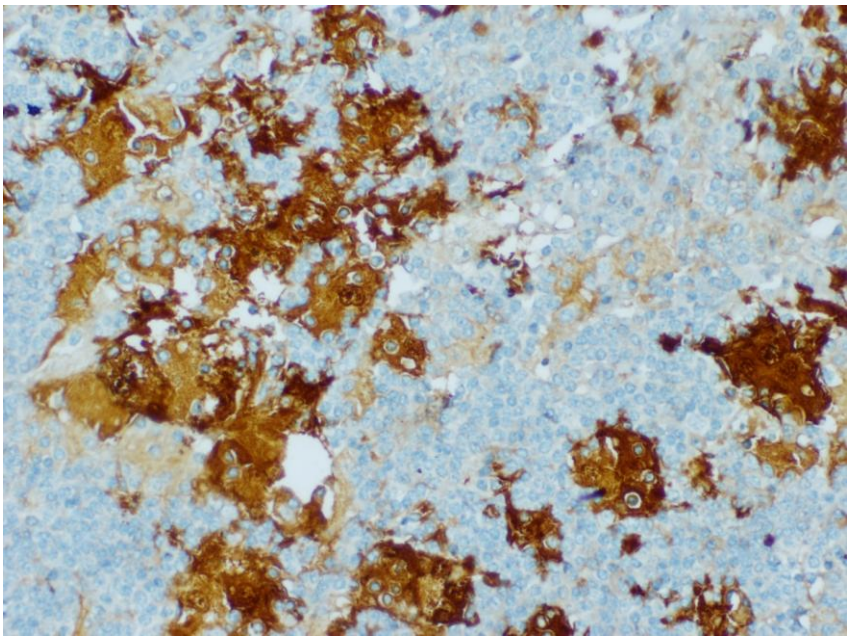

Figure 4 Histiocytes are S100 positive (IHCx200)

SHML otherwise Rosai Dorfman disease, introduced in 1969 by Rosai and Dorfman, is a rare disease of unknown etiology that commonly involves the cervical lymph nodes. SHML commonly presents as massive painless bilateral neck nodal mass. Rarely extranodal sites are involved in 30-40% cases, the common sites being head and neck, central nervous system, eyes, respiratory tracts and skin. The cause is unknown although a viral etiology is suspected. Characteristic histologic findings include emperipolesis (engulfment of lymphocytes) and S100 protein positivity.

There are only few reports of SHML of the orbit. A 12-year-old boy with Rosai-Dorfman disease presenting with bilateral relapsing uveitis and

papilledema that appeared four years before the onset of lymphadenopathy was described [1]. A 38-year-old woman had compressive optic neuropathy of the right eye caused by orbital involvement with sinus histiocytosis and received cyclophosphamide, vincristine, and prednisone [2]. A 20-year-old man with ocular involvement with uveitis and deep marginal corneal infiltrates in association with cervical lymphadenopathy was reported [3]. A 57 year old lady with SHML of orbit has been described, the tumor was excised and she was alive without recurrence at 3 year [4].

Conclusion

The disease resolves spontaneously in most patients and treatment is required only in organ threatening situations where steroids or chemotherapy has been tried. Though the orbit is a rare site for SHML, the disease should be considered in the differential diagnosis of orbital swellings.

References

1. Pivetti-Pezzi P, Torce C, Colabelli-Gisoldi RA, Vitale A, Baccari A, Pacchiarotti A. Relapsing bilateral uveitis and papilledema in sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). ***Eur J Ophthalmol***. 1995, 5:59-62
2. Goldberg S, Mahadevia P, Lipton M, Rosenbaum PS. Sinus histiocytosis with massive lymphadenopathy involving the orbit: reversal of compressive optic neuropathy after chemotherapy. ***J Neuroophthalmol***. 1998, 18:270-275
3. Rumelt S, Cohen I, Rehany U. Marginal corneal infiltrates: a possible new manifestation of sinus histiocytosis with massive lymphadenopathy. ***Cornea***. 2000, 19:857-858
4. R Khan, P Moriarty S Kennedy. Rosai Dorfman disease or sinus histiocytosis with massive lymphadenopathy of the orbit. ***Br J Ophthalmol***. 2003, 87:1043-1055