Destombes–Rosai Dorfman Disease: A Rare Case Report

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Abstract Destombes Rosai Dorfman disease and sinus histiocytosis with massive lymphadenopathy is a rare benign idiopathic proliferative disease of phagocytic histiocytes. Approximately 80% of patient present with painless massive cervical lymphadenopathy. It rarely involves other sites for example CNS, eyes, upper respiratory tract and skin. A case of Destombes Rosai Dorfman disease is reported with history of gradual protrusion of left eye since many years. The histological features were consistent with the diagnosis.

Keywords Orbit · Rosai dorfman disease · Management

Introduction

Rosai and Dorfman [1] described the rare disease entity in 1969. Rosai Dorfman disease is a rare benign histiocytosis predominately affects children and young adults. The clinical presentation is most often a bilateral cervical painless lymphadenopathy. Extra nodal manifestation of the disease occur in up to 43% of patients, the most common sites affected are soft tissue of head and neck, paranasal sinuses and nasal cavity, because of the rarity of the diagnosis, the extensive differential consideration, the multiple site of involvement, the diagnosis of sinus histiocytosis with massive lymphadenopathy (SHNL) is often not consider, which leads to in appropriate treatment of benign but often progressive disease. Most of the patients are in good health and no significant symptoms relating to this disease which

complicate the diagnosis [2]. We report an unusual location of Destombes Rosai Dorfman disease.

Case Report

A 60-year-old female presented with history of gradual protrusion of left eye since 5 years, initially diagnosed as orbital pseudotumor.

Physical examination revealed no lymphadenopathy, no weight loss or fever noticed. Patient presented with moderate pallor, indicative of anaemia. The eye examination revealed limited extraocular muscle movements laterally and eccentric proptosis with superior displacement. Visual acuity, fundoscopy was within normal limits. In local examination two swelling were present on lower lid and distal wall of eyeball non tender 3×3 cm.

Laboratory investigations were done and ultrasonography showed intraconal, hyperechoic lesion involving lateral rectus muscle. MRI demonstrated well defined isodense lobulated soft tissue mass in intraconal retro orbital region measuring $5 \times 3 \times 3$ cm and pushing left eyeball anteriorly (Fig. 1). The patient underwent a debulking procedure to excise the tumour of the left orbit whose biopsy (excisional biopsy) yielded firm, tan, slightly fibrous tissue demonstrating a mixed histiocytic and lymphocytic infiltrate and few phagocytes (Fig. 2). Initiation of short term steroid (dexamethasone 8 mg twice daily for 5 days) regimen resulted with subsequent improvement.

Discussion

Since 1969, when Rosai and Dorfman first described a rare benign lymphoproliferative disease, then referred to as

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Fig. 1 CT scan

Fig. 2 Histopathological picture

sinus histiocytosis with massive lymphadenopathy and now known primarily as Rosai Dorfman disease [1]. More than 600 cases have been registered [3].

The orbit, globe and eyelid are well known sites of involvement in Rosai Dorfman disease, occurring in 7–10% of patients [2]. Amongst the registered patients, 40 cases had some evidence of orbital or eyelid involvement. Only 8 of these 40 revealed no evidence of lymphadenopathy, including our case (Fig. 3).

Most patients presented with proptosis, limited eye movements, eyelid oedema and decreased visual acuity, orbital involvement is usually unilateral [4]. Bilateral is rare [5]. Appearance of orbital involvement during imaging has been reported in a few cases [4–6]. The lesion usually

Preoperative photograph of the patient

Postoperative photograph of the patient

Fig. 3 a Pre op. b Post op

appears as an infiltrative soft tissue mass with variable contrast enhancement. Fatty infiltration within the orbital mass is occasionally seen. Some cases have both eyelid and orbital involvement with extensive pre-septal and non-septal masses, but lesion can be purely retrobulbar.

Lacrimal gland involvement also has been reported, as has direct intra cranial extension through the orbital apex to the middle cranial fossa.

In one case report describing the appearance of this lesion, the lesion was noted as being isodense to gray matter [7] with homogenous contrast enhancement. The differential diagnosis is broad and includes optic nerve sheath tumour, leukaemia, inflammatory pseudotumor, sarcoidosis, Wegener's granulomatosis and infectious processes [2].

Histopathological features showed a mixed histiocytic and lymphocytic infiltration with few phagocytosis. Various clinical and laboratory findings include fever, anaemia, neutrophilia and elevated ESR [1].

Ophthalmic manifestation is rare and seen in 9–11% of cases. The most frequent ophthalmic manifestation are orbital soft tumour mass with proptosis of eyelid [4, 7, 8], as in our case. Very few articles have been reported for ophthalmic involvement.

Most 95% patient present with cervical lymphadenopathy, the extent of lymphadenopathy is often striking with nodes as large as 6 cm noted, in some cases leading to respiratory compromise [9] laboratory abnormalities are non specific and include anemia, hyper gamma globulinemia and increased ESR and WBC, the pathogenesis of this disease is poorly understood although some have suggested as either infection or immuno deficiency might play a role [9, 10]

Sinus histiocytosis is characterized histologically by nodal sinus dilation with non neoplastic proliferation of histiocytes. These histiocytes typically demonstrate a foamy or vacuolated eosinophilic cytoplasm and characteristically contain small mature lymphocytes, a condition called lymphocytophagocytosis. However, more fibrosis is evident and lymphophagocytosis may be often less demonstrated. Although many patients experience spontaneously regressing course, some will have disease presence in more extensive form requiring treatment to prevent functional loss.

The reported mortality rate is 7% with many patients who have fatal outcome having concomitant immune dysfunction. There have been some reports of success in treating Rosai Dorfman disease with combination of cytotoxic agent such as alkylating agents, vinca alkaloid and prednisone [11].

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