·Letter to the Editor·

Rosai-Dorfman disease of unilateral lacrimal gland in an elderly Chinese male

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Dear Sir,

I am Dr. Jing Li, from the Department of Ophthalmology, Beijing Tongren Hospital, Beijing, China. I want to present a rare case of orbital Rosai-Dorfman disease presenting as unilateral lacrimal gland enlargement without lymphadenopathy involvement.

Rosai-Dorfman disease is also called sinus histiocytosis with massive lymphadenopathy (SHML), which was first described by Rosai and Dorfman in 1969 [1]. This is a rare, benign, idiopathic histiocytic proliferative disorder that occurs predominantly in children and young adults [2]. Its typical manifestation is massive, painless, bilateral cervical lymohadenopathy, fever, leukocytosis, high erythrocyte sedimentation rate and hypergammaglobulinemia [2]. Even though orbital involvement may occur in 11% of patients with Rosai-Dorfman disease, isolated lacrimal gland involvement is very rare. To the best of our knowledge, only four cases in the literature have been reported with isolated lacrimal gland involvement, with age ranging from 7-year-old to 57-year-old [3-5]. Our patient is a 72-year-old Chinese male, who is the first Chinese to be reported and older than all previous reports.

A 72-year-old Chinese man presented to our department of ophthalmology because of a one-month history of right lacrimal region swelling. The mass was slowly increasing in size without any discomfort. He hadn't ever applied any drug by himself. There had been no similar history of this appearance in his family so far. Systemic evaluation including a chest radiograph and cervical lymph nodes was normal. Ophthalmologic examination showed that the right lacrimal gland was enlarged, which was firmed, oval-shaped and removable. The movement of the right eyeball wasn't affected. The remainder of ophthalmologic examination was completely normal. Axial magnetic resonance image (MRI) of the orbit demonstrated enlarged lacrimal gland of the right eye with distinct margin. The mass showed equal T1 (Figure 1A) and T2 (Figure 1B) signal intensity and an evident homogeneous enhancing on dynamic contrast-enhanced magnetic resonance imaging (Figure 1C). No abnormal signal founded in surrounding orbital bone or any other nearby region.

Then, complete surgical excision of the mass was carried out, showing the size of the mass was approximately 1.0cm× 1.5cm ×1.5cm. The pathological examination of frozen section pointed to a benign proliferative disorder. So we performed a simple tumor excision. Postoperatively, the resected specimens were made routine paraffin sections, and then histopathologic and immunohistochemical examinations were performed. Histopathologic examination showed polymorphic plasma cell infiltration with lymphocytes, histiocytes, plasmocytes and a few phagocytes (Figure 2A), and large histiocytes exhibited characteristic emperipolesis (Figure 2B). Immunohistochemically, the mass demonstrated positive for CD68 (Figure 3), S-100, lysozyme, CD20, CD21, CD35, CD34 and negative for CD1a, CD30, CK and P53. Thus, a definite diagnosis of Rosai-Dorfman disease without lymphadenopathy was confirmed. There was no evidence of recurrence or lymphadenopathy involvement at the 2-year follow-up.

In conclusion, Rosai-Dorfman disease can only involves unilateral lacrimal gland. Although very rare, as ophthalmologists, we should recognize the possibility of the occurrence of isolated lacrimal gland involved by Rosai-Dorfman disease. Only in this way, can we consider this disease in the differential diagnosis of unilateral lacrimal gland enlargement and decrease the misdiagnosis rate as much as possible.

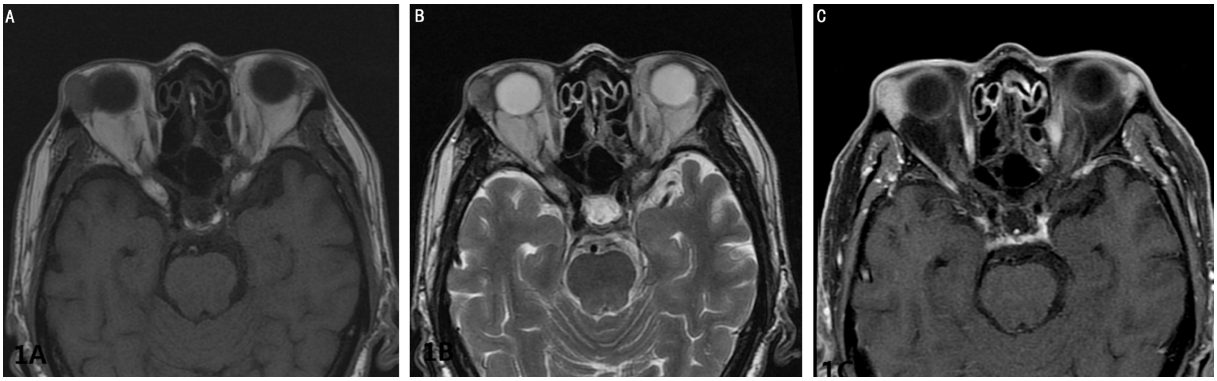

Figure 1 Axial magnetic resonance image of the orbit demonstrating enlarged lacrimal gland of the right eye with distinct margin The mass showed equal T1 (A) and T2 (B) signal intensity and an evident homogeneous enhancing on dynamic contrast-enhanced magnetic resonance imaging(C).

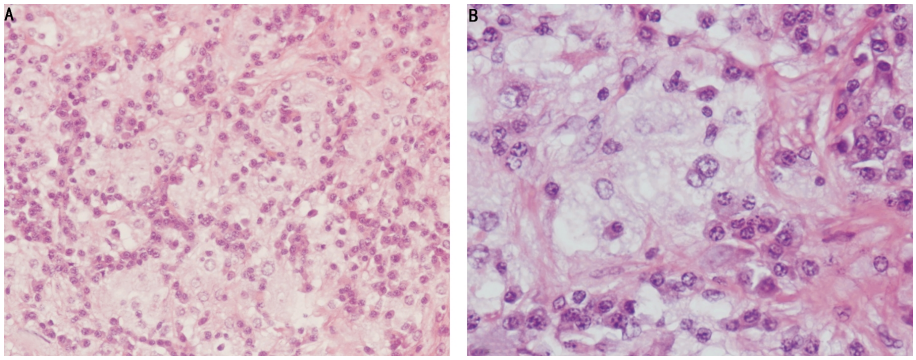

Figure 2 A: Histopathologic examination showing polymorphic plasma cell infiltration with lymphocytes, histiocytes, plasmocytes and a few phagocytes (hematoxylin-eosin, ×10). B: Large histiocytes exhibiting characteristic emperipolesis (hematoxylin-eosin, ×40).

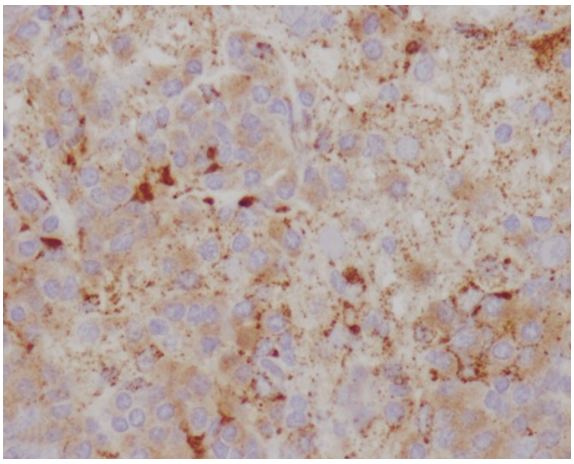

Figure 3 Photomicrograph demonstrating positive for CD68 (immunostain, ×40).

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