Isolated lacrimal gland involvement in Rosai-Dorfman-Destombes disease

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Rosai-Dorfman-Destombes (sinus histiocytosis with massive lymphadenopathy) disease is an uncommon disease characterized by benign proliferation of histiocytes, with painless lymph node enlargement and frequent extranodal disease. Orbital involvement occurs in 9-11% of cases. However, isolated Rosai-Dorfman-Destombes disease of the lacrimal gland without any systemic involvement is very rare with only three case reports.

We describe here one such young male patient with unilateral lacrimal gland swelling. Excision biopsy revealed almost complete replacement of the lacrimal gland by lymphocytes, plasma cells and large pale histiocytes. The latter exhibited emperipolesis and stained positive for S-100 and CD68 on immunohistochemistry. Patient is well and has no other manifestation or recurrence of the disease during a follow-up of 24 months.

Key words: Lacrimal gland, Rosai-Dorfman-Destombes disease, sinus histiocytosis with massive lymphadenopathy

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Destombes in 1965 reported four cases of “adenitis with lipid excess”, in children and young adults which went unnoticed till Rosai and Dorfman four years later characterized the unique pathological features of this rare lymph nodal disorder which they called sinus histiocytosis with massive lymphadenopathy. Extralymphal involvement of one or multiple tissues and organs by this Rosai Dorfman or more appropriately Rosai-Dorfman-Destombes (RDD) disease can occur. Even though the head and neck region is commonly involved, its occurrence in the lacrimal gland is rare. Isolated involvement of lacrimal gland...
without lymph nodal involvement is even rarer with only three case reports in the literature till date.3,4 Because of this extremely unusual occurrence we present here one such case report of a young male who presented with swelling in the lacrimal gland region.

Case Report

A 38-year-old man presented with a swelling in the left upper eyelid for three years. The mass was painless and slowly progressive. There was no history of fever or any other systemic involvement. Clinical examination of the left eye revealed mild proptosis and downward and inward dystopia. Fundus examination was normal. The other eye was completely normal. Magnetic resonance imaging (MRI) of the orbit showed an extraconal mass in the left lacrimal gland region abutting the left globe and pushing it anteriorly and medially [Figure 1]. The optic nerves, chiasma and rectus muscles revealed normal MRI signals. Comparison with the previous MRI scans done three and two years earlier showed no significant change in size of the lesion. The other orbit was normal. Complete surgical excision of the mass was carried out. A single nodular tissue measuring 2.5 x 2 x 1.5 cm was received for histopathological examination. Its cut surface was homogeneous, smooth and grey white. Routinely stained tissue sections revealed a lesion comprising nodular aggregates of small lymphocytes and diffuse occasional aggregates [Figure 2A]. In between these were many large foamy histiocytes, some of which had lymphocytes and plasma cells entrapped within the cytoplasm [Figure 2B]. Hyalinized collagen bands along with proliferated myofibroblasts were also present. Most of the lacrimal gland was destroyed by the disease except for a few entrapped remnants of lacrimal ducts [Figure 2C]. Immunostaining showed the foamy histiocytes to be strongly positive for S-100 protein [Figure 2D] and CD68. A diagnosis of RDD disease involving the left lacrimal gland was made. A follow-up of about 24 months was available after surgical resection of mass. There has been no recurrence, neither has the patient developed any lymphadenopathy or any other systemic organ involvement.

Discussion

Rosai-Dorfman-Destombes disease or sinus histiocytosis with massive lymphadenopathy (SHML) is a rare non-neoplastic proliferative disorder of histiocytes that affects predominantly children and young adults, with a slight male predominance. Nearly 87% of patients have bilateral cervical painless lymphadenopathy; other nodal groups are less frequently involved.2 In some cases these extranodal manifestations represent the predominant or even exclusive manifestation of the disease. Practically all organ systems have been recorded as being the site of the disease. The most common are eyes and ocular adnexa. Classically, other sites include skin, upper respiratory tract, salivary gland, epidural space, bone and orbital soft tissue.5 The most frequent manifestation is an orbital soft tissue mass with proptosis. To the best of our knowledge, there are only three case reports with isolated lacrimal gland involvement.6,7 In the earlier three case reports along with our present case the lacrimal gland involvement was unilateral. In our present case, the lacrimal gland mass was very well circumscribed with no extension into orbital soft tissues.

Typically, RDD disease undergoes spontaneous remission after a protracted course but may develop recurrences. Rarely, the involvement of vital organ may lead to death. In our cases no further treatment was given after surgical resection of the mass. No recurrence or any lymphadenopathy was noted in a follow-up period of 24 months.

The etiology is still unknown. Viruses like Epstein-Barr virus and Human Herpes virus 6 have been suggested. Molecular studies done on involved tissue have failed to show evidence of clonality in keeping with their presumed reactive nature.

Rosai-Dorfman-Destombes disease presents in its most typical form as lymph node involvement with sinus histiocytosis.3 It is important to be familiar with the clinical and diagnostic histopathologic features of this entity especially when the disease presents extra-nodally in an isolated form. Histopathologic features include the presence of lymphoid aggregates altering with pale-appearing areas composed of histiocytes and plasma cells.8
The diagnosis is based on the identification of large histiocytic cells showing emperipolesis. Emperipolesis is the presence of intact lymphocytes, plasma cells, erythrocytes or polymorphonuclear leukocytes engulfed within the cytoplasm of the histiocytes.

Immunohistochemical stains are useful in differentiating RDD disease from other histiocytic disorders. The RDD histiocytes are strongly positive for S-100 and CD 68 as was observed in our case. No immunoreactivity to CD1a is observed (CD1a is expressed in Langerhans cells).

To summarize, eyes and ocular adnexa, including lacrimal gland involvement represent the most common extranodal areas affected by RDD disease. This disease is not considered as fatal. In many cases, RDD undergoes quick and complete spontaneous resolution. In others, it follows a protracted clinical course for years or decades. This latter is particularly true in cases with widespread extranodal involvement.

References
1. Destombes P. Adenitis with lipid excess, in children or young adults, seen in the Antilles or Mali (4 cases). Bull Soc Pathol Exot Filiales 1965;58:1169-75.
2. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: A newly recognized benign clinicopathological entity. Arch Pathol 1969;87:63-70.
3. Yuen HK, Cheuk W, Leung DY, Tse RK, Chan N. Atypical presentation of Rosai-Dorfman disease in the lacrimal gland mimicking malignancy. Ophthal Plast Reconstr Surg 2006;22:145-7.
4. Quintyn JC, Ranty ML, Courville P, Metayer J, Retout A. Orbital sinus histiocytosis (Rosai-Dorfman disease): A lacrimal gland involvement. Ophthalmologica 2002;216:277-80.
5. Juan Rosai In Rosai and Ackerman's Surgical Pathology. 9th ed. vol 2, Mosby; 2004. p. 1911-3.
6. Wenig BM, Abbondanzo SL, Childers EL, Kapadia SB, Heffner DR. Extranodal Sinus Histiocytosis with massive lymphadenopathy (Rosai Dorfman Disease) of the Head and Neck. Hum Pathol 1993;24:483-92.