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Case Report

Ocular Rosai-Dorfman disease and juvenile xanthogranuloma present at the same time in a young girl. A clinicopathologic report

Alberto Luis Díaz, MD *
Augusto José Gómez, MD **
Carolina Serrano-Calderón, MD ¶
Vanessa C. Fuentes, MD ¶

Summary

Purpose: To report the case of two histiocytic disorders, both present together in a young patient, as the Rosai-Dorfman Disease (RDD) and the Juvenile Xanthogranuloma.(JXG). **Methods**: We examined a 15-year-old girl with multiple, bilateral epibulbar conjunctival masses without apparent nodal involvement on whom we made a partial surgical split. In order to reach a proper diagnosis we perform a complete medical history and physical examination. **Results**: Physical examination was normal and there was no evidence of lymphadenopathies. The diagnosis was confirmed by histopathology and immune histochemical analysis of the split lesions. The rest of the tumours responded satisfactorily to steroid therapy. There was no evidence of RDD or JXG elsewhere. **Conclusion**: RDD and JXG may appear together as epibulbar tumours without any other systemic involvement. [*Díaz AL, Gómez AJ, Serrano-Calderón C, Fuentes VC. Ocular Rosai-Dorfman disease and juvenile xanthogranuloma present at the same time in a young girl. A clinicopathologic report. MedUNAB 2007; 10:130-132*].

Key words: Rosai-Dorfman disease, Juvenile xanthogranuloma, Histiocytosis, Conjunctival neoplasms, Eye.

Resumen

Propósito: Informar el caso de una paciente joven con dos trastornos histiocíticos simultáneos: enfermedad de Rosai-Dorfman y xantogranuloma juvenil. **Métodos:** Examinamos una adolescente de 15 años con múltiples masas de la conjuntiva epibulbar de ambos ojos sin aparente compromiso nodal y en quién se realizó resección parcial de las lesiones. **Resultados:** El examen físico fue normal, sin evidencia de linfadenopatías. El diagnóstico fue confirmado con histopatología y análisis histoquímico de las lesiones removidas. Las demás masas respondieron satisfactoriamente a la terapia con esteroides. No se ha encontrado evidencia posterior de ninguna de las dos entidades. **Conclusión:** Tanto la enfermedad de Rosai-Dorfman como el xantogranuloma juvenil pueden presentarse juntos como tumoraciones epibulbares sin cualesquiera otro compromiso sistémico. [*Díaz AL, Gómez AJ, Serrano-Calderón C, Fuentes VC. Enfermedad de Rosai-Dorfman y xantogranuloma juvenil ocular simultáneas en una adolescente. Un informe clinicopatológico. MedUNAB 2007; 10:130-132*]**.**

Key words: Enfermedad de Rosai-Dorfman, Xantogranuloma juvenil, Histiocitosis, neoplasias conjuntivales, Ojo.

* Department of Oculoplastic and Orbital Surgery, Fundación Oftalmológica de Santander Clínica Carlos Ardila Lülle, Bucaramanga, Colombia.
** Department of Anterior Segment, Fundación Oftalmológica de Santander Clínica Carlos Ardila Lülle, Bucaramanga, Colombia.
¶ Resident in Ophthalmology, Universidad Industrial de Santander; Fundación Oftalmológica de Santander Clínica Carlos Ardila Lülle, Bucaramanga, Colombia.

Correspondence: Dr. Serrano, Fundación Oftalmológica de Santander, Urbanización el Bosque. Floridablanca, Colombia.
E-mail address: carolinaserranoc@gmail.com

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Introduction

The histiocytosis are an important group of diseases that involve the mononuclear phagocytic system and consist of proliferation of macrophages in different organs and systems.[1] Rosai-Dorfman disease (RDD) and Juvenile Xanthogranuloma (JXG) are two different histiocytosis that share several characteristics including their benign course. But they rarely present together in the same patient, at the same time, compromising just the epibulbar conjunctival tissue. As far as we are aware this combination has not been previously reported.

Case report

We report a case of a female patient that came up with isolated involvement of the conjunctiva, who developed a rapid growth (4 months) of bilateral and multiple epibulbar pink salmon tumors of different size, almost all about 2 to 4mm., without cornea involvement (figure 1). A complete physical examination did not reveal lymphadenopathy or any other organs and systems involved. She gave a history of atopy, erythema nodosum and scarlet fever at early age without any other past history.

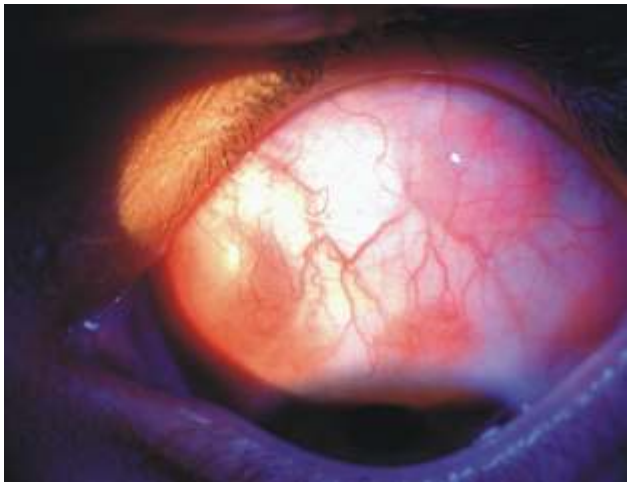

Figure 1. Macroscopic image of salmon colour tumours in the epibulbar conjunctiva.

Our initial diagnosis was a lymphoma and for this reason we made a partial surgical excision on both eyes of the four conjunctival lesions. The diagnosis was made by histological examination showing in the left eye proliferated histiocytosis and infiltration of plasma cells and lymphocytes in a special pattern of emperipolesis doing the diagnosis of RDD (figure 2), and in the right eye a similar pattern with foamy histiocytes but without emperipolesis such as JXG. On immunohistochemical staining histiocytes were reactive for S-100 and CD68 proteins in RDD but just reactive for CD68 in JXG (figure 3). Lymphocytes were

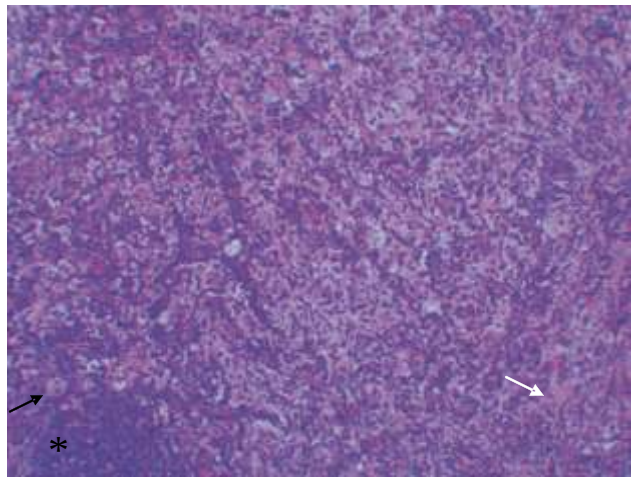

Figure 2. Histiocytes demonstrating emperipolesis (black arrow) often overshadowed by extensive lymphoplasmacytic infiltrates (asterisk) and fibrosis (white arrow) in the background. (H and E, 100x).

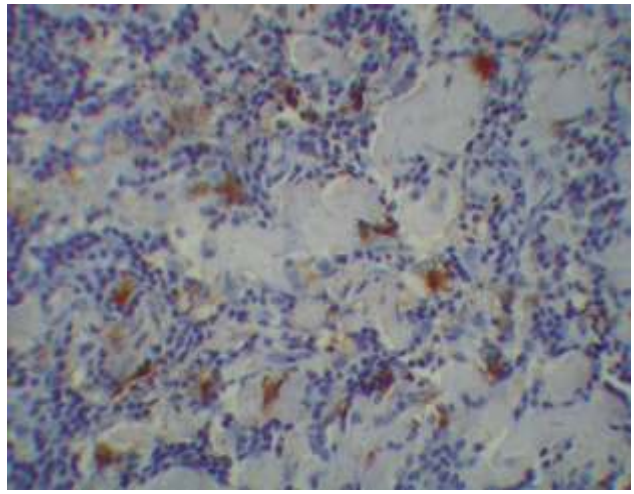

Figure 3. Immunohistochemical staining showing the characteristic histiocytes positive for S100 and CD68 protein in RDD and JXG (400x).

Cd3 and CD20 positive and plasma cells were polyclonal for Kappa and Lambda chains.

The analysis of serum levels of C-reactive protein, total proteins, albumin, globulin, calcium, phosphorus, creatinine, urea and lactate dehydrogenase were normal. Blood and urine test, RA test and erythrocyte sedimentation rate were normal as well. Elevated levels of Ig E (226.3 IU/mL) were documented with normal levels of Ig G and Ig M. Antinuclear antibodies were reactive 1:40. Torax X-ray did not show any relevant feature. CT of the orbit, abdomen and pelvis were all normal. The tumours responded satisfactorily to steroid therapy. There was no evidence for RDD or JXG elsewhere.

Discussion

The Histiocytes International Society classifies the histiocytosis in three large groups: Langerhans´ cells histiocytosis, monocytes/macrophages cells histiocytosis different from Langerhans´ cells (among them the RDD and JXG) and the harmful histiocytic disorders.[1]

The sinus histiocytosis with massive lymphadenopathy (SHML) or RDD is a rare histiocytic disorder of unknown origin which shares several cell markers with JXG. Although Rosai-Dorfman cells exhibit an aberrant immunophenotype, the indolent clinical course of SHML suggests a reactive disorder rather than a neoplastic process.[2] Its clinical features include bilateral cervical lymphadenopathies (in a 90%) of great size, painless, that also can affect any other ganglionar region with extranodal signs by extension to another corporal region in a 43% of the cases.[3] Extranodal infiltrates are often found in the absence of lymphadenopathy, in this case the term SHML would be inappropriate, being more correct the eponym RDD. The compromise of the ocular globe and orbit is uncommon with just 11% [2] reported in the reviewed literature and the conjunctiva is a tissue of unusual infiltration that has been reported in just seven cases in the whole world. Histologically RDD is characterized by the features described above.[4] Lymphoma shares, both histological and inmunohistochemical patterns with RDD except the reactivity for S-100 protein.

At present, seven previous cases of ocular histiocytosis with bulbar affectation have been reported in the literature. In 1988, the first two cases of SHML were known with epiescleral mass and cervical and inguinal lymphadenopathy in patients less than 20 years. Wong reported in 1989 a patient of 40 years old with cutaneous manifestations and epibulbar mass. Subsequently, Ireland in 1999 showed a case of RDD in a 4 year-old girl with bilateral epibulbar masses, as only demonstration of the disease. Tan published in 2002, 63 year-old patient with SHLM diagnostic by ocular epibulbar affectation and chronic uveitis without lymphadenopathy. Finally, Albini in 2005 reported two cases of histiocytosis in older adults, with epibulbar tumors without ganglionar compromise.[5]

JXG is a cutaneous granulomatous disease occurring primarily in infants of less than one year and less commonly in older children and adults. Blank et al in 1949, reported the first case of ocular JXG in a 4-month old male infant who presented with an iris mass and secondary glaucoma.[6] Few cases of conjunctival involvement have been reported. Histologically, the typical appearance of JXG is a mixture of foamy and epitheloid histiocytes with scattered lymphocytes, eosinophils, and occasional plasma cells; the classic Touton giant cell with its ring of nuclei is often seen, especially in mature lesions. The cells are positive for CD68 and negative for S-100 protein and CD1a.[6]

The differential diagnosis of RDD and JXG should be made with ocular lymphoma. This patients should be evaluated integrally by internal medicine, ophthalmology, hematology and histophatology for the good management and resolution of the disease.

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