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Key Words:

Rosai-Dorfman-Destombes, Histiocytosis, Orbito-Palpebral Pseudotumor, Corticosteroid Therapy.

Orbito-Palpebral Pseudotumors Revealing Rosai-Dorfman-Destombes Disease: A Case Report

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Abstract

We report the case of a young woman presenting with bilateral orbito-palpebral pseudotumors and bilateral parotidomegaly, a condition that had been slowly progressive over eight years. The diagnosis was confirmed by histopathology, which revealed the presence of large, foamy histiocytes (emperipolesis) that were S100-positive and CD1a-negative on immunohistochemistry. Corticosteroid therapy led to rapid clinical improvement within three months. This clinical presentation highlights the importance of maintaining a high index of suspicion regarding any unusual orbito-palpebral or parotid pseudotumoral mass.

Introduction

Rosai-Dorfman-Destombes disease (RDD) is a "group R" histiocytosis characterized by the abnormal accumulation of activated histiocytes within affected tissues, leading to clinical manifestations such as painless lymphadenopathy, systemic symptoms, and/or symptoms specific to the involved anatomical site [1,2]. The estimated annual incidence of RDD is approximately 1 case per 200,000 individuals in the United States, whereas its global incidence remains poorly defined [1,2]. In Africa, Laudin et al. reported 149 cases in their meta-analysis of African cases published in 2023 [3]. Reviewing clinical, radiological, laboratory, and pathological data is essential for establishing an accurate diagnosis of RDD, given the disease's rarity, the heterogeneity of clinical presentations, and the variability of the prognosis. Key histopathological features of RDD include: large histiocytes with round nuclei, prominent nucleoli, and abundant, pale cytoplasm; emperipolesis (the presence of intact hematopoietic cells within the histiocyte cytoplasm); and a plasma cell-rich background, with histiocytes testing positive for the S100 marker and negative for CD1a via immunohistochemistry [1,2]. Therapeutic options for RDD vary, and clinical management is tailored to the specific clinical picture [3-7]. This atypical presentation of RDD highlights the importance of maintaining a high index of suspicion when encountering any unusual, slowly progressive orbital or parotid mass.

Case Report

We report the case of a 21-year-old female student with no significant medical history other than an atopic background (chronic allergic rhinitis, atopic dermatitis, family history of atopy), who had been under follow-up since the age of 13 for bilateral orbito-palpebral pseudotumors and slowly



Figure 1: Left orbito-palpebral pseudotumor (L): firm, painless mass filling the palpebral fissure, associated with contralateral (right) ptosis.

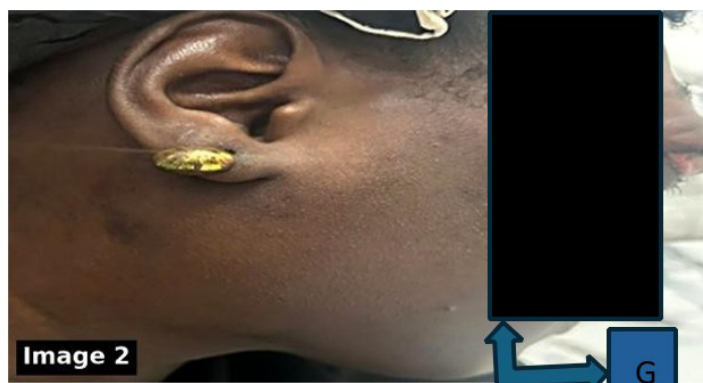


Figure 2: Profile view: swelling of the parotid/subauricular region, corresponding to the described bilateral parotidomegaly



Figure 3: Complete resolution of parotidomegaly and cervical lymphadenopathy after 3 months of corticosteroid therapy.

progressive bilateral parotid enlargement. The condition began eight years earlier with unilateral eyelid swelling that gradually became bilateral and was subsequently accompanied by bilateral parotid swelling. There were no reports of fever, constitutional symptoms, or night sweats. Clinical examination revealed firm, non-tender, asymmetric bilateral orbito-palpebral masses and total vision loss in the left eye, along with bilateral parotid swelling (without associated facial paralysis) and non-inflammatory right-sided cervical lymphadenopathy. Dermatological examination showed pruritic, erythematous-scaly lesions in the skin folds (consistent with her known atopic dermatitis) and abdominal macules, as well as congenital-type

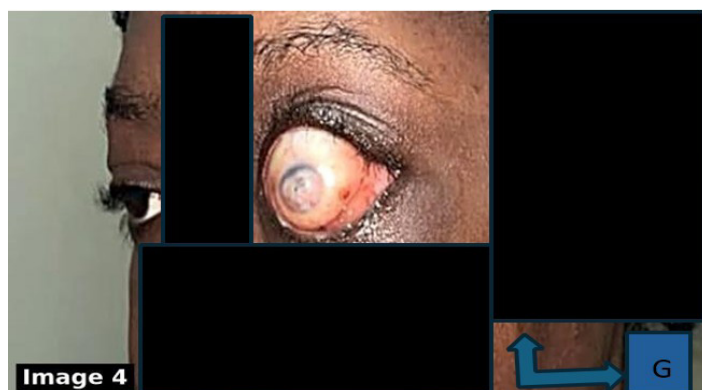
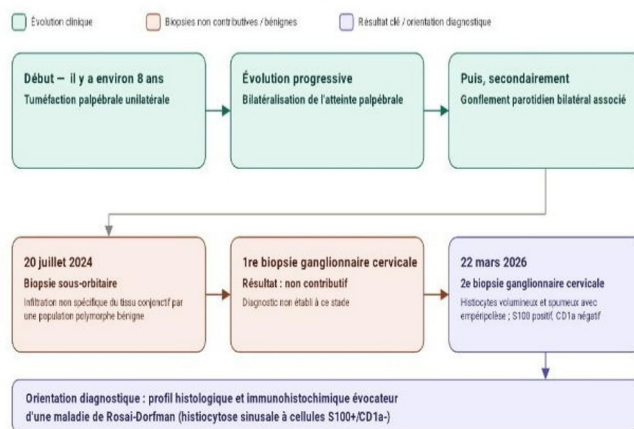


Figure 4: Regression of the orbito-palpebral pseudotumor after 3 months of corticosteroid therapy, with exposure of the left eyeball and residual proptosis; residual corneal opacity is visible.

Figure 5: Clinical and histological timeline.

Frise chronologique – Évolution clinique et diagnostique

*Tumefaction palpébrale unilatérale puis bilatérale, gonflement parotidien, et biopsies successives sur 8 ans



melanoderma of the cheeks and palms, but no specific skin lesions suggestive of primary cutaneous Rosai-Dorfman disease (RDD). The remainder of the general physical examination was unremarkable.

Laboratory tests revealed moderate normochromic normocytic anemia (hemoglobin 10.1 g/dL) without leukocytosis, accompanied by thrombocytosis (platelets 500,000/ μ L) and signs of chronic inflammation (CRP 46 mg/L). Renal function was preserved (urea 0.11 g/L, serum creatinine 6.4 mg/L, glomerular filtration rate 100.15 mL/min), and the serum electrolyte panel was normal (sodium 145 mmol/L, potassium 4.5 mmol/L, chloride 102 mmol/L); the albumin-to-creatinine ratio was 11.20 mg/mmol, and 24-hour urinary calcium excretion was 142.80 mg/24h. Viral serology (HIV, hepatitis), toxoplasmosis, and syphilis tests were negative. The autoimmune workup was negative (antinuclear antibodies, ANCA, angiotensin-converting enzyme, serum IgG4). MRI of the brain and facial region showed a diffuse, infiltrative tissue process involving the parotid gland, an extensive left orbital process, a localized lesion on the right, and calcified foci within the opacified

ethmoid sinuses.

Histopathological analysis of the infraorbital biopsy performed on July 20, 2024, revealed nonspecific connective tissue infiltration by a benign polymorphic cell population; the initial biopsy of the cervical lymph nodes was non-contributory. A second biopsy of the cervical lymphadenopathy, performed on March 22, 2026, revealed the presence of large, foamy histiocytes (emperipolesis), with S100 positivity and CD1a negativity on immunohistochemistry (Figure 1).

The patient received a corticosteroid bolus (methylprednisolone at 120 mg/day, i.e., 5 mg/kg/day for a body weight of 70 kg) for three days, followed by a switch to oral therapy at 1 mg/kg/day with a progressive dose reduction of 10% every two weeks. Currently, the patient is taking 10 mg daily. The clinical course was favorable after three months of treatment, marked by the complete resolution of parotidomegaly and lymphadenopathy (Image 3) and significant regression of the orbito-palpebral pseudotumors (Image 4).

Discussion

Rosai-Dorfman-Destombes disease (RDD) is a rare histiocytosis with extremely diverse clinical manifestations [3-7]. The case we report illustrates an unusual presentation involving orbital-palpebral pseudotumors and bilateral parotidomegaly, mimicking a malignant tumor. This extranodal involvement is rarely described in the literature, where the most common sites of disease include cervical lymph nodes, skin, the respiratory tract, the digestive system, and the central nervous system [3-7]. The diagnosis of RDD relies on histopathology and immunohistochemistry. Typical features include the presence of large histiocytes that are S100-positive and CD1a-negative, exhibiting emperipolesis [3-7]. In our case, initial biopsies were non-specific, highlighting the diagnostic challenge and the need for repeat sampling. Confirmation was achieved through the histopathological identification of large, foamy histiocytes (showing emperipolesis) alongside S100 positivity and CD1a negativity on immunohistochemistry. There is no standardized therapeutic approach for RDD. Options include observation, surgery, radiotherapy, and various systemic treatments (corticosteroids, immunosuppressants, targeted therapies). The choice depends on the location, extent, and severity of symptoms. In our case, corticosteroid therapy led to rapid clinical improvement, confirming its efficacy in certain inflammatory forms. This result aligns with several observations reported in the literature, where corticosteroids are considered a first-line treatment option for symptomatic cases [8-11]. The clinical variability of RDD can mimic various conditions, particularly IgG4-related diseases, chronic infections, and other histiocytoses. This

highlights the need for a multidisciplinary approach—involving clinicians, radiologists, and pathologists—to achieve a reliable diagnosis, as well as the value of corticosteroid therapy, which can serve as an effective treatment alternative for symptomatic extranodal disease. Thus, this case adds to the literature on atypical RDD presentations and underscores the appropriate management approach for unusual orbital-palpebral or parotid masses. 4. Conclusion Rosai-Dorfman-Destombes disease remains a rare and complex entity characterized by significant clinical and histopathological heterogeneity. The case presented illustrates an atypical orbital-palpebral and parotid localization that mimicked a malignant tumor. The diagnosis was confirmed by the histopathological identification of large, foamy histiocytes (showing emperipolesis) and positive S100 immunohistochemical staining—findings that allowed for the exclusion of the main differential diagnoses. Rapid clinical improvement with corticosteroid therapy confirms the value of this treatment for certain inflammatory forms, while underscoring the need for management tailored to the location, extent, and severity of symptoms. This case thus adds to the literature on extranodal presentations of the disease and highlights the importance of a multidisciplinary approach for reliable diagnosis and appropriate management.

Conflicts of interest: The authors declare no conflicts of interest.

Author contributions:

*Abdoulaye SOW: Writing, review and editing, visualization.

*Omar Boun KHATAB: Supervision, validation.

All other authors: validation.

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Citation: Dr. Abdoulaye SOW , Orbito-Palpebral Pseudotumors Revealing Rosai-Dorfman-Destombes Disease: A Case Report . *Jour of Clin Cas Rep, Med Imag and Heal Sci* 14 (5)-2026.

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