Non surgical management of unilateral lacrimal gland swelling in a paediatric patient of Rosai-Dorfman disease: A case report

Sailie Shirodkar, Prachi Patil, Nagendra Shah

A 10-year-old girl presented to us with inferonasally displaced, non-axial proptosis of oculus dexter (OD), with ipsilateral, painless massive cervical lymphadenopathy of ten weeks duration. After a detailed hematological work-up and imaging, the histological evidence obtained on cervical lymph node biopsy established the diagnosis of Rosai-Dorfman disease. Although, Rosai-Dorfman disease of unilateral lacrimal gland is extremely infrequent, a high degree of suspicion is warranted in a case of young patient presenting with unilateral isolated lacrimal gland swelling and associated lymphadenopathy, wherein no other cause is found. Treatment protocol should be individualized as per the extent of systemic involvement and functional disability.

Key words: Lacrimal gland swelling, Rosai-Dorfman disease, sinus histiocytosis

Rosai-Dorfman disease or sinus histiocytosis is an extremely rare disorder, originally described by Destombes in 1965 and later identified as a distinct clinicopathological entity in 1969 by Rosai and Dorfman.[1] The most classical manifestation in 90% cases, is chronic, painless cervical lymphadenopathy with pyrexia, leukocytosis, hypergammaglobulinemia etc. However, this presentation is not mandatory for diagnosis. 43% of patients have reported with extranodal manifestations. These extranodal sites include the skin, soft tissue, nasal cavity, eye, orbit, ocular adnexa, oral cavity, central nervous system etc. The ophthalmic manifestations include eyelid or orbital masses, and very rarely uveitis.[1,2]

Case Report

A 10-year-old girl presented to us with painless progressive swelling of the upper and outer quadrant of oculus dexter (OD) of six weeks duration [Fig. 1a], along with painless swelling of the right cervical area of eight weeks duration, failing to respond to persistent antibiotic treatment. She also complained of diplopia on dextroelevation for the past four weeks. There was no history of systemic disease, surgery, trauma or associated visual or ocular complaints such as redness, discharge, lacrimation etc.

Her uncorrected visual acuity was 6/6, N6 with normal color vision and an intraocular pressure of 14 mm of Hg in both eyes. OD showed non-axial inferonasal proptosis of 4 mm with restriction of dextroelevation. There was no evidence of discoloration of the lid skin, vascular prominence or change in size or shape of swelling with posture changes or Valsalva maneuver. On palpation, OD revealed a well-defined, non-tender, firm orbital swelling of approximately 2.0 × 1.0 cm, beneath the anterior orbital rim in the superotemporal quadrant, with no definite palpable posterior margin and associated irregular anterior orbital margin. Anterior segment examination was normal, while fundus examination showed retinochoroidal folds in the superotemporal quadrant. Oculus sinister (OS) was normal.

On systemic examination, multiple massive, confluent, non-tender and rubbery enlarged anterior cervical lymph nodes were noted, the largest one being approximately 2.5 × 2.0 cm. No other organ involvement was observed.

Blood picture demonstrated microcytic hypochromic anemia with leukocytosis, elevated erythrocyte sedimentation rate (ESR) with hypergammaglobulinemia and marginally elevated serum Angiotensin Converting Enzyme (ACE) level. Rest of the investigations were normal. Fine needle aspiration cytology (FNAC) of the cervical lymph nodes was done, which showed non-specific lymphadenitis. Excisional biopsy of the cervical lymph was done [Fig. 2a and b], based on which a confirmatory diagnosis of Rosai-Dorfman disease was made.

To assess the extent of disease, PET scan was also reported before and after treatment [Fig. 3a and b]. Computerized Tomography (CT) of the head and orbit demonstrated a homogenously enhancing soft tissue mass of the right orbit, approximately 22 mm cranio-caudally, 31 mm antero-posteriorly, 16 mm transversely in size, causing external compression over right globe with remodelling of anterior orbital rim of lateral wall and displacement of superior and lateral rectus [Fig. 4].

After consulting the paediatric oncologist, medical management was started with intravenous dexamethasone for five days, followed by oral steroids and 6-mercaptopurine for eight weeks with excellent resolution of lacrimal gland swelling clinically [Fig. 1b and c]. Over the next three years, there was no reported recurrence of the disease in the orbit or any focus of sinus histiocytosis elsewhere in the body.

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Department of Ophthalmology, Taparia Institute of Ophthalmology, Bombay Hospital and Institute of Medical Sciences, New Marine Lines, Mumbai, Maharashtra, India

Correspondence to: Dr. Nagendra Shah, Department of Ophthalmology, Taparia Institute of Ophthalmology, Bombay Hospital and Institute of Medical Sciences, 12, New Marine Lines, Mumbai - 400 020, Maharashtra, India. E-mail: kanudans@gmail.com

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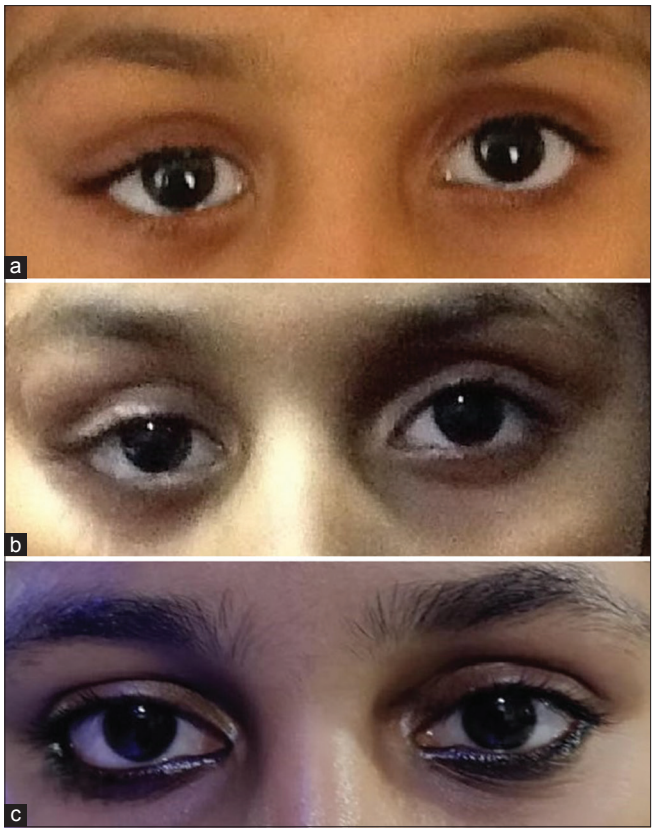

Figure 1: (a) Patient with right eye showing marked inferonasal non-axial proptosis of 4 mm. (b) Patient with mild reduction of proptosis, 3 days after initiation of intravenous steroids. (c) Complete resolution of proptosis, 1 month after the initiation of treatment

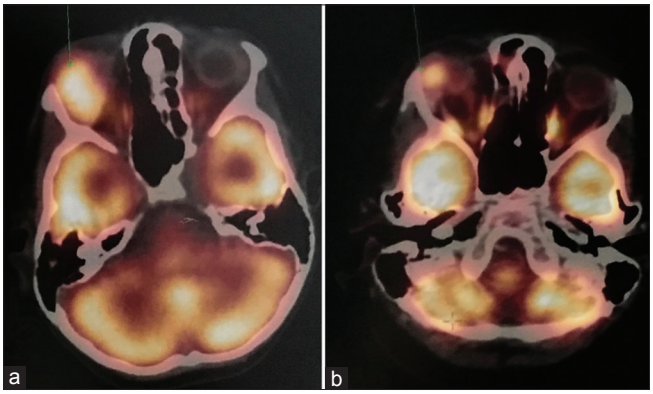

Figure 3: (a) PET scan: showing increased metabolic activity and enlargement (3 × 2 × 1.8 cms) of the right lacrimal gland. (b) PET scan: showing approximately 20% reduction in metabolic activity and regression of size

Discussion

Rosai-Dorfman disease (RDD), or sinus histiocytosis is an extremely rare disorder, wherein there is non-neoplastic proliferation of histiocytes. In a study of 423 patients, 43% cases were noted to have at least one extranodal site of involvement in addition to lymph node affectation, while 23% were found to show extranodal involvement alone. Around 9 to 11% of the cases were found to show involvement of the eye, orbit or ocular adnexa.[1,2]

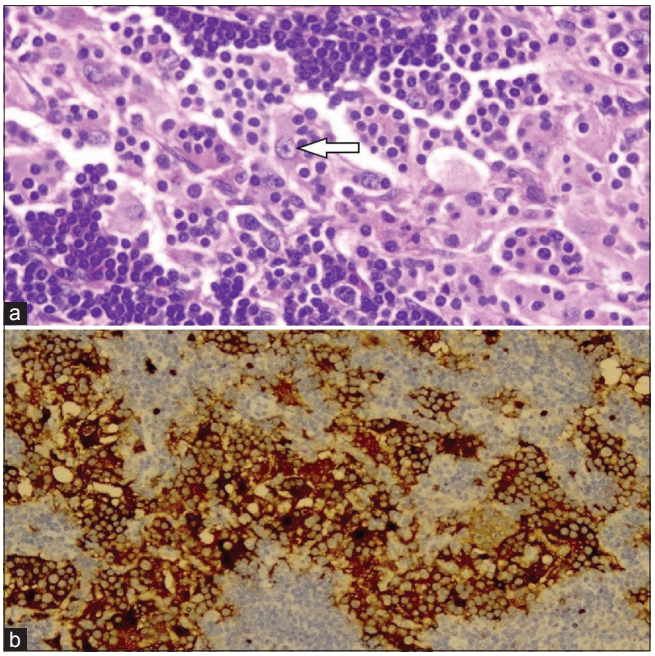

Figure 2: (a) Excisional biopsy of cervical lymph node: Histological section stained with H and E stain (400 × magnification) demonstrating dilated sinusoids containing numerous large histiocytic cells with pale staining round nuclei and abundant cytoplasm, and emperipolesis. (b) Excisional biopsy of cervical lymph node: CD68 stain (400 × magnification) showing cytoplasmic positivity in histiocytes

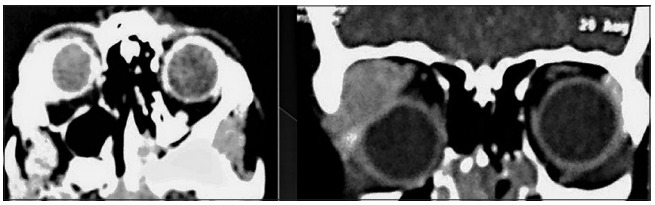

Figure 4: Computerized Tomography (CT) of the Head and Orbit: homogenously enhancing soft tissue mass of the right orbit, approximately 22 mm cranio-caudally, 31 mm antero-posteriorly, 16 mm transversely in size, causing external compression over right globe

The etiology is unknown. Some have speculated it to be an exaggerated immune response against infectious agents or antigen. Human herpesvirus-6, Epstein-Barr virus, Parvovirus B19 etc., have been suspected to play a role in the pathogenesis.[1,2] Current studies also suggest that the defect lies in the Fas/FasL signaling leading to altered apoptosis whereby uncontrolled histiocytic proliferation is triggered.[1]

The differential diagnoses of lacrimal gland swelling associated with cervical lymphadenopathy in young patients include dacryoadenitis, Wegener's disease, sarcoidosis, lymphoreticular malignancies etc., As most of these are responsive to steroids and show comparable CT scan patterns, the diagnosis is challenging and is mainly based on clinical features, histopathological examination and immunohistochemistry.[3,4]

Histopathology shows diffuse histiocytosis with bland nuclei, well-defined nuclear membrane and a single small nucleolus. The cells show abundant eosinophilic to amphophilic

cytoplasm. The hallmark of the disease is 'emperipolesis', indicating presence of multiple phagocytosed lymphocytes in intracytoplasmic vesicle.[1-5] Immunochemistry shows histiocytes to be positive for CD68 protein, while they are negative for CD1a. This helps distinguish the condition from Langerhans cell histiocytosis.[5]

Only 50% of cases require definitive management, as the condition may be self-limited. There is no definite treatment protocol. Management options include observation for mild manifestations with no cosmetic or functional abnormality; surgical excision or debulking for lesion in surgically accessible locations and/or systemic corticosteroids, chemotherapy (vinca alkaloids, alkylating agents etc.) or radiotherapy in patients with severe symptoms where vital organ function is compromised causing severe handicap.[6-8]

Conclusion

In our case, in view of the young age of the patient, medical management was adhered to, so as to avoid surgical trauma and post-operative scarring due to debulking. Considering the rapid and sustained response to corticosteroids and chemotherapy, in addition to prevention of residual disease or recurrence, the line of management seems to be a promising approach in the management of RDD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Sujata G. Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: An overview. Arch Pathol Lab Med 2007;131:1117-21.
2. Vemuganti GK, Naik MN, Honavar S. Rosai dorfman disease of the orbit. J Hematol Oncol 2008;1:7.
3. Azoulay R, Brisse H, Fréneaux P, Ferey S, Kalifa G, Adamsbaum C. Lacrimal location of sinus histiocytosis (Rosai-Dorfman-Destombes disease). AJNR Am J Neuroradiol 2004;25:498-500.
4. Pinto DCG, Vitigal TA, Castro B, Santos BH, DeSousa NJA. Rosai- Dorfman disease in the differential diagnosis of cervical lymphadenopathy. Braz J Otorhinolaringol 2008;74:632-35.
5. Mantilla JG, Goldberg-Stein S, Wang Y. Extranodal Rosai-Dorfman disease: Clinicopathologocail series of 10 patients with radiologic correlation and review of the literature. Am J Clin Pathol 2016;145:211-21.
6. Albini TA, Evans M, See R, Rao NA, Marback E, de Souza MM. Rosai-Dorfman disease: Isolated epibulbar masses in two adult patients. Br J Ophthalmol 2005;89:241-3.
7. Khan R, Moriarty P, Kennedy S. Rosai Dorfman disease or sinus histiocytosis with massive lymphadenopathy of the orbit. Br J Ophthalmol 2003;87:1054.
8. Dalia S, Sagatys E, Sokol L, Kubal T. Rosai-Dorfman disease: Tumour biology, clinical features, pathology and treatment. Cancer Control 2014;21:322-7.