Pediatric Hematology Oncology Journal 4 (2019) 94–96

Contents lists available at ScienceDirect

Pediatric Hematology Oncology Journal

journal homepage: https://www.elsevier.com/journals/pediatric-hematology-oncology-journal/

Unilateral inguinal lymphadenopathy in a 6-year-old girl: An unusual presentation of Rosai-Dorfman disease

Keywords:
Child
Rosai Dorfman disease
Unilateral lymphadenopathy

A B S T R A C T

Rosai Dorfman disease (RDD) commonly presents as bilateral cervical lymphadenopathy in children and young adults. We herein report a young girl with unilateral inguinal lymphadenopathy as a rare presentation of RDD.

1. Introduction

Rosai Dorfman disease (RDD) commonly presents as bilateral cervical lymphadenopathy in children and young adults. The first clinical description of this entity was published in 1969 by Rosai and Dofman, though Destombes had described it in 1965 [1]. It can involve other lymph node groups as well as extranodal sites. We herein describe a young girl with unilateral inguinal lymphadenopathy as a rare presentation of RDD.

2. Case report

A 6-year-old premorbidily normal girl presented with painless swelling over left groin of one month duration. She did not have fever, weight loss, swelling or wound over left lower limb, or genital ulcer. There was no history of contact with tuberculosis. Developmental history was unremarkable. Examination revealed normal anthropometry; enlarged left superficial inguinal nodes, largest measuring 3 cm x 2 cm, non-tender, firm consistent with no fluctuation [Fig. 1]. Examination of genitalia and left lower limb was normal. Systemic examination was unremarkable. Laboratory investigations showed hemoglobin of 126 g/L, leukocytosis (11.6×10^9/L) with differential count 60% neutrophils, 33% lymphocytes, 6% monocytes and 1% eosinophils, and thrombocytosis (462×10^9/L). Serum transaminase levels were normal (Alanine transaminase: 25 U/L, Aspartate transaminase: 37 U/L). Chest X-ray and ultrasound abdomen were normal. Tuberculin skin test was non-reactive and gastric lavages for acid fast bacilli were negative. She received 2 weeks of oral amoxicillin-clavulanic acid without any improvement following which final needle aspiration cytology (FNAC) was performed from left inguinal lymph node. Cytology showed reactive lymphoid cells with follicles and lymphohistiocytic tangles. There were numerous histiocytes, which showed emperipolesis of lymphocytes which is consistent with Rosai Dorfman disease (RDD) [Figs. 2 and 3]. No acid-fast bacilli or malignant cells were identified in the cytology smears. Positron Emission Tomography-Computed Tomography of whole body revealed FDG avid left inguinal and left external iliac lymph nodes. Serologies for Human Immunodeficiency Virus, Epstein Barr virus, and cytomegalovirus were non-reactive. Immunoglobulin (Ig) profile revealed normal IgG- 10.35 g/L (normal: 5.4–16.10 gL) and IgA - 0.74 g/L (normal: 0.50–2.40 g/L) but low IgM- **0.48** g/L (normal: 0.50–1.80 g/L). Direct Coombs test and antinuclear antibody by indirect immunofluorescence were negative. Assessment of double negative T cells by flow cytometry was normal. Parents were reassured and no specific treatment was proffered in view of isolated lymphadenopathy without any systemic involvement. She was noted to have complete resolution of left inguinal lymphadenopathy at 6 months of follow-up. Currently at 2 years of follow-up, she is doing well.

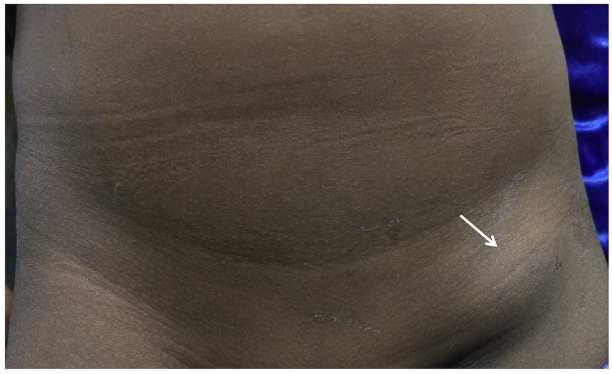

Fig. 1. Left inguinal swelling.

Peer review under responsibility of Pediatric Hematology Oncology Chapter of Indian Academy of Pediatrics.

https://doi.org/10.1016/j.phoj.2019.12.005

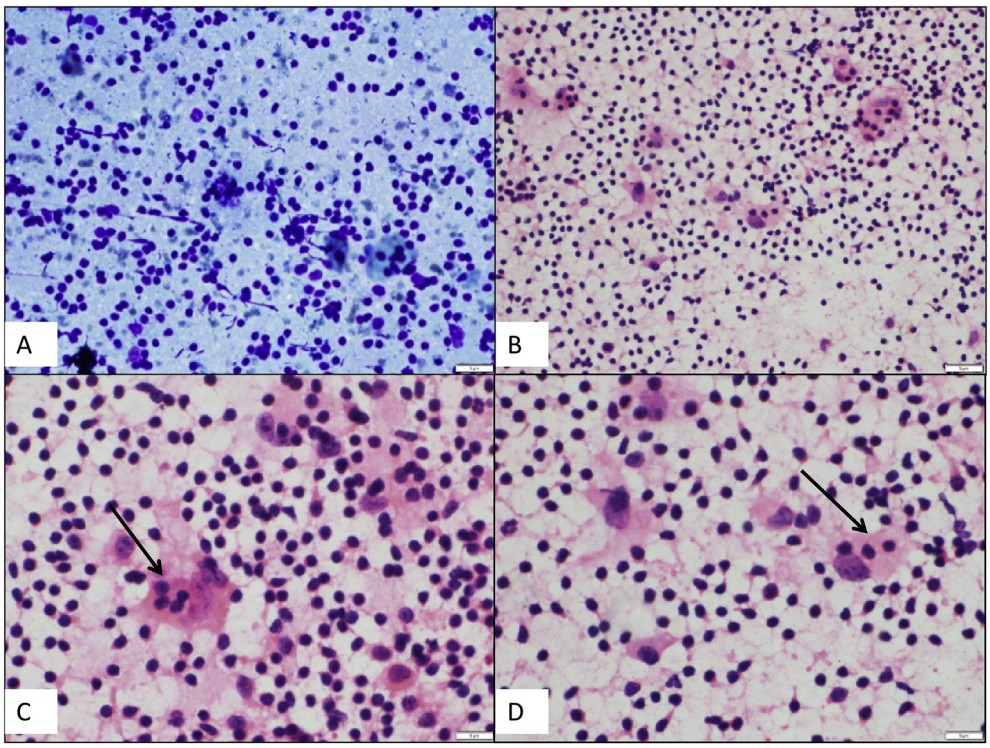

Fig. 2. A, B- MGG and H&E stained slides (20x view) showing numerous lymphocytes, histiocytes and lymphohistiocytic tangles. C,D- H&E stained slides- Histiocytes showing presence of intact lymphocytes with in their cytoplasm (Emperipolesis).

3. Discussion

RDD commonly presents as bilateral cervical lymphadenopathy in children and young adults. The first clinical description of this entity was published in 1969 by Rosai and Dofman, though Destombes had described it in 1965 [1]. It can involve other lymph node groups as well as extranodal sites. However, unilateral inguinal lymphadenopathy as presentation of RDD is unusual. We

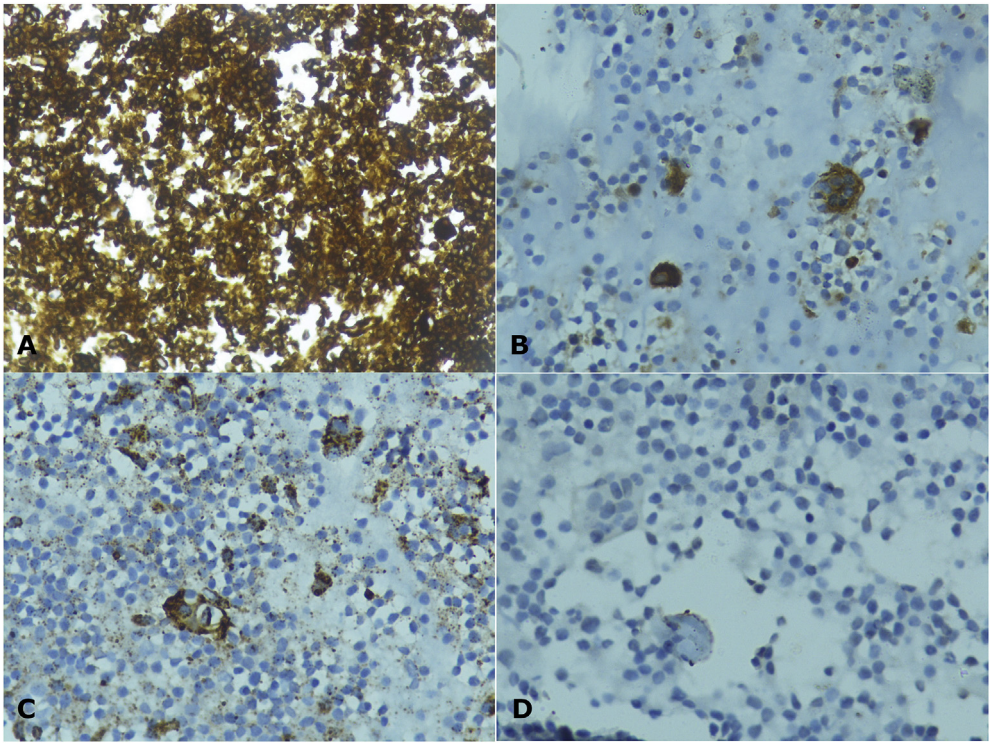

Fig. 3. Immunocytochemistry on FNA cell block section. A, CD45 immunostain showing membrano-cytoplasmic positivity in all the leukocytes. B, C, S100 and CD68 immunostain showing membrano-cytoplasmic positivity in the histiocytes. The lymphocytes are seen with in the cytoplasm of these histiocytes. D, CD1a immunostain showing negativity in the histiocytes.

herein describe a young girl with unilateral inguinal lymphadenopathy as a rare presentation of RDD. RDD is a proliferative disorder of histiocytes which usually manifests as rapidly progressive lymphadenopathy, hence the acronym sinus histiocytosis with massive lymphadenopathy. The most common presentation is enlargement of cervical lymph nodes [2]. However, lymphadenopathy involving inguinal, axillary, mediastinal and other group of lymph nodes may also occur [3]. Lymph node involvement is often bilateral and painless. Our patient had unilateral inguinal involvement which has rarely been reported in RDD.

Disease can involve extra-nodal sites including skin, soft tissue, upper and lower airway, sinuses, salivary glands, bones, orbits and urogenital system [2,3]. Fever may be seen in 30% of patients but is often not the presenting complaint. Laboratory abnormalities include anemia, neutrophilic leukocytosis and hypergammaglobulinemia. Etiology remains unknown, though an aberrant immune response to infection with Epstein Barr virus, Cytomegalovirus, and Klebsiella has been reported [4,5]. It is a self limiting condition unless the course is complicated by extranodal involvement [6]. Disease may often be confused with other forms of histiocytosis like Langerhan cell histiocytosis [7]; malignancy like Hodgkin's lymphoma; and tuberculosis [2]. RDD can coexist with autoimmune disorders in approximately 10% cases like systemic lupus erythematous, juvenile idiopathic arthritis, autoimmune hemolytic anemia, and rarely autoimmune lymphoprolifertaive disease [2]. Investigations for these etiologies were negative in our case. Selective IgM deficiency is a defined as isolated deficiency of IgM (<−2 SD of mean) and normal IgG, and IgA and T cell functions. Index case had isolated IgM deficiency during presentation which has resolved in follow-up. Selective IgM deficiency has been associated with various autoimmune disorders [8]. However, significance of this finding in context of RDD is unknown but interesting.

Diagnosis is confirmed on histopathological examination of the specimen which shows predominant histiocytic cells with variable number of plasma cells and lymphocytes. The most characteristic finding is emperipolesis, which refers to the presence of histiocytes with phagocytosed cells, predominantly lymphocytes [2,3]. Patients generally do well with symptomatic management and, disease often warrants no specific treatment in view of benign course and self limiting nature. However, patients with airway or vital organ involvement may require corticosteroids, immunomodulators (alkylating agents and antimetabolites), and surgical resection [2].

Source of funding

None.

Declaration of competing interest

None.

Acknowledgements

Nil.

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Rakesh Kumar Pilania
Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Aman Gupta
Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Zaidi Ariba
Department of Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Pandiarajan Vignesh*
Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India

* Corresponding author. Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, 160012, India.
E-mail address: vigimmc@gmail.com (P. Vignesh).

9 November 2019
Available online 19 December 2019