

Assess the Utility of Minimally Invasive Technique Fine Needle Aspiration Cytology for Early Diagnosis of Cases of Rosai Dorfman Disease

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Abstract

Aims: The objective here is to assess the utility of minimally invasive technique fine needle aspiration cytology for early diagnosis of cases of Rosai Dorfman disease.

Material and Methods: The cytology of 10 cases of Rosai Dorfman disease.

Results: Our study included 10 cases out of which 6 were of nodal Rosai Dorfman whereas 4 of them showed extranodal disease.

Conclusions: FNAC is a simple, primary, minimally invasive technique which is a reliable first line investigation in the diagnosis of Rosai Dorfman disease and very useful in prevention of overdiagnosis and overtreatment of a self-resolving disease.

Keywords: Emperipolesis, Rosai Dorfman disease, sinus histiocytosis with massive lymphadenopathy, extranodal RosaiDorfman disease.

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Introduction

Sinus histiocytosis with massive lymphadenopathy or Rosai Dorfman disease is a rare self-limited and benign disease and was first described in 1969[1]. Patients present classically with painless enlargement of cervical lymph nodes often accompanied by fever, leukocytosis, anemia and polyclonal hypergammaglobulinemia. However, other lymph nodes as well as single or multiple extranodal involvement is also fairly common. A distinctive feature is emperipolesis the presence of lymphocytes, red blood cells, and few plasma cells within vacuoles in the cytoplasm of many histiocytes. Clinically it mimics various

neoplastic and non-neoplastic lesions but has a self-limiting nature. Fine needle aspiration cytology reveals distinct morphology and can help in primary diagnosis by its typical cytological features and prevent consequences of overdiagnosis.

Material and Methods

We studied cytological features of 10 cases of Rosai Dorfman disease from May 2017 to December 2021. Patients were clinically examined and sent for FNAC as primary investigation. FNAC was performed using 22 gauge needle and slides were further fixed in isopropyl alcohol. On primary diagnosis of

Rosai Dorfman disease, these cases were followed and histopathological examination

was done by routine fixing and staining and the findings were correlated.

Results

Table 1: Clinical findings of Nodal Rosai Dorfman patients

S.No	Age/ Sex	Site of involvement	Duration of symptoms	Other symptoms
1	40/Male	Bilateral cervical lymphadenopathy	1 Year	Fever, malaise
2	45/Male	Bilateral inguinal, cervical lymphadenopathy	4 months	Asymptomatic
3	50/ Male	Bilateral axillary, cervical lymphadenopathy	6 months	Malaise
4	12/Male	Right sided inguinal lymphadenopathy	8 months	Fever, weakness
5	43/Male	Bilateral cervical lymphadenopathy	1.5 years	Fever
6	38/Female	Bilateral cervical, axillary lymphadenopathy	1 year	Asymptomatic

Table 2: Clinical findings of Nodal Rosai Dorfman patients

S.No	Age/sex	Site of involvement	Duration of symptoms	Other symptoms	Initial clinical diagnosis
7	40/Male	Left lower back	2 years	Recurrent similar lesions	Lipoma
8	28/Female	Left breast	8 months	Asymptomatic	Fibroadenoma
9	44/Male	Right thigh	4 months	Asymptomatic	Lipoma
10	32/Male	Left forearm	6 months	Itching	Infected cystic lesion

Case 1:

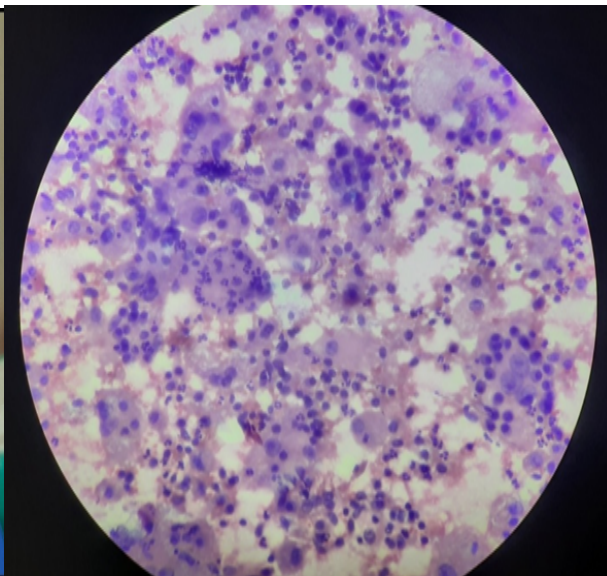
40 years male presented to the ENT OPD with multiple bilateral painless enlargement of cervical lymph nodes with on and off fever and weakness. Patient was sent to us for FNAC. USG findings revealed enlarged cervical lymph nodes largest measuring around 41x22mm with suspected internal degeneration. Cytological findings revealed sheets of large foamy histiocytes which were engulfing lymphocytes, plasma cells, red blood cells and neutrophils. Few histiocytes

were showing large nuclei, prominent nucleoli and some binucleate and multinucleate histiocytes were also seen. Vascular proliferation was also evident. FNAC findings were suggestive of Sinus histiocytosis with massive lymphadenopathy.

On histopathological examination, these cells showed emperipolesis along with mild cytological atypia and variable inflammatory cell infiltrate. Advice for histopathology and IHC was made.



Case 1: clinical picture of 40 years male with cervical lymphadenopathy



Cytology reveals emperipolesis

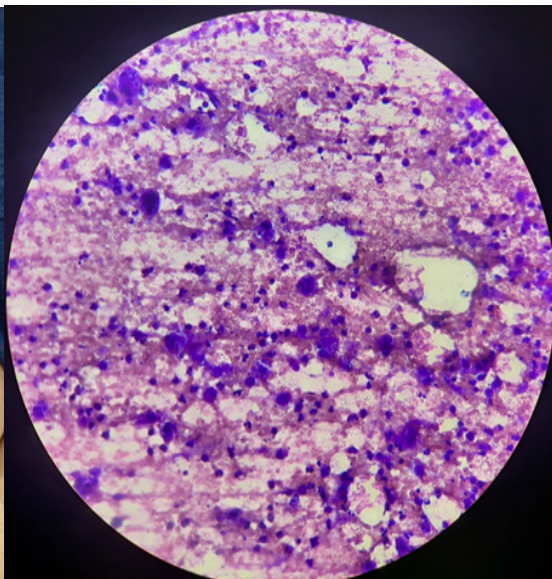
Case 4:

A twelve year male presented to our hospital with complain of fever, weakness since 7-8 months. Patient also complained of right sided inguinal lymphadenopathy. Clinically, lymphoma was suspected and patient was

sent for investigation. On USG right sided lymph node measured 5X3 cm with fatty hilum. On FNAC numerous histiocytes were seen that were engulfing lymphocytes and plasma cells. On histopathology, histiocytes were showing emperipolesis. Mild inflammatory cell infiltrate was also seen.



Case 4: clinical picture 12 year male with inguinal lymphadenopathy



Histiocytes showing emperipolesis

Case 7:

A forty year male presented to surgery OPD with complain of left lower back swelling. He was sent for FNAC to our hospital. Clinically, lesion size was approx. 1.5x1.5 cm, soft mobile and non-tender. Patient gave history of similar self-resolving lesions associated with some itching present on forearm, arm, thigh since 2 years. On RDD is now established as a separate entity. The exact etiology of this disease is unknown. Recent data suggests that it is an exaggerated immune response of the hematolymphoid system. [2,3]

Earlier texts suggested the role of Epstein barr virus, Herpes virus, Parvo virus B19 and Polyoma virus. [4-8]

Autoimmune mechanism is also proposed in some studies. [9,10] The proliferating histiocytes are polyclonal hence they are reactive and not neoplastic. [11]

Cytological findings are characteristic yielding a highly accurate diagnosis. Numerous large histiocytes are seen engulfing other blood cells. Background of lymphocytes, plasma cells and neutrophils are also seen. [12,13]

Nearly all cases diagnosed in our study showed good cellularity showing these histiocytes which were demonstrating emperipolesis. However, extranodal RDD showed less prominent emperipolesis than nodal RDD.

Histology reveals dilated lymphatic sinuses occupied by lymphocytes and histiocytes along with phagocytosis of lymphocytes, neutrophils and plasma cells (emperipolesis). [1,14]

On immunohistochemistry histiocytes show strong S100. The differential diagnosis on cytology of nodal RDD includes reactive lymphadenitis, granulomatous lymphadenitis, sinus histiocytosis and LCH. [15]

cytology, sheets of histiocytes were seen engulfing lymphocytes, RBCs and neutrophils. A diagnosis of Rosai dorfman disease was made and histopathology was advised for further confirmation.

Discussion

RDD has excellent prognosis and spontaneous remission hence it is of more importance to diagnose the disease correctly.

References

1. Foucar E, Rosai J and Dorfman R: Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Review of the entity. *Semin Diagn Pathol.* 1990; 7: 19-73.
2. Cai Y, Shi Z, Bai Y. Review of Rosai-Dorfman disease: New insights into the pathogenesis of this rare disorder. *Acta Haematol.* 2017;138:14-23.
3. Yoon AJ, Parisien M, Feldman F, Lee FY. Extranodal Rosai-Dorfman disease of bone, subcutaneous tissue and paranasal sinus mucosa with a review of its pathogenesis. *Skeletal Radiol.* 2005;34:653-7.
4. Tsang WY, Yip TT and Chan JK: The Rosai-Dorfman disease histiocytes are not infected by Epstein-Barr virus. *Histopathology.* 1994; 25: 88-90.
5. Mehraein Y, Wagner M, Remberger K, Füzesi L, Middel P, Kaptur S, Schmitt K and Meese E: Parvovirus B19 detected in Rosai-Dorfman disease in nodal and extranodal manifestations. *J Clin Pathol.* 2006; 59: 1320-1326.
6. Arakaki N, Gallo G, Majluf R, Diez B, Arias E, Riudavets MA and Sevlever G: Extranodal rosai-dorfman disease presenting as a solitary mass with human herpesvirus 6 detection in a pediatric patient. *Pediatr Dev Pathol.* 2012; 15: 324-328.
7. Ortonne N, Fillet AM, Kosuge H, Bagot M, Frances C and Wechsler J: Cutaneous

- Destombes-Rosai-Dorfman disease: Absence of detection of HHV-6 and HHV-8 in skin. *J Cutan Pathol*. 2002; 29: 113-118.
8. Al-Daraji W, Anandan A, Klassen-Fischer M, Auerbach A, Marwaha JS and Fanburg-Smith JC: Soft tissue Rosai-Dorfman disease: 29 new lesions in 18 patients, with detection of polyomavirus antigen in 3 abdominal cases. *Ann Diagn Pathol*. 2010; 14: 309-316.
 9. Grabczynska SA, Toh CT, Francis N, Costello C and Bunker CB: Rosai-Dorfman disease complicated by autoimmune haemolytic anaemia: Case report and review of a multisystem disease with cutaneous infiltrates. *Br J Dermatol*. 2001; 145: 323-326.
 10. Maric I, Pittaluga S, Dale JK, Niemela JE, Delsol G, Diment J, Rosai J, Raffeld M, Puck JM, Straus SE, *et al*: Histologic features of sinus histiocytosis with massive lymphadenopathy in patients with autoimmune lymphoproliferative syndrome. *Am J Surg Pathol*. 2005;29: 903-911.
 11. Paulli M, Bergamaschi G, Tonon L, Viglio A, Rosso R, Facchetti F, *et al*. Evidence for a polyclonal nature of the cell infiltrate in sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). *Br J Haematol*. 1995;91:415-8.
 12. Das DK, Gulati A, Bhatt NC, Sethi RG. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Report of two cases with fine needle aspiration cytology. *Diagn Cytopathol*. 2001;24:42-5.
 13. Deshpande AH, Nayak S, Munshi MM. Cytology of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). *Diagn Cytopathol*. 2000;22:181-5.
 14. Shi Y, Griffin AC, Zhang PJ, Palmer JN and Gupta P: Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease): A case report and review of 49 cases with fine needle aspiration cytology. *Cytojournal*. 2011; 8: 3.
 15. Kumar B, Karki S and Paudyal P: Diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) by fine needle aspiration cytology. *Diagn Cytopathol*. 2008; 36: 691-695.