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Case Report

Sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman Disease) - A rare case of isolated axillary lympadenopathy diagnosed on FNAC

Vineet Banga [1,*]

[1] *Dept. of Pathology, Acharyashree Bhikshu Government Hospital, Moti Nagar, New Delhi, India*

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ABSTRACT

Rosai-Dorfman disease (Sinus Histiocytosis with Massive Lymphadenopathy) is a rare disorder characterized by overproduction (proliferation) and accumulation of histiocytes in the lymph nodes of the body causing lymphadenopathy, most often involving neck. In some cases, abnormal accumulation of histiocytes may occur in other areas of the body besides the lymph nodes (extra nodal). These areas include the skin, central nervous system, kidney, and digestive tract. The case is being reported as a rare case involving isolated axillary lymph node and was diagnosed on FNAC.

1. Introduction

Sinus histiocytosis with massive lymphadenopathy (SHML) also known as Rosai-Dorfman disease (RDD), is a rare histiocytic proliferative disorder of unknown etiology initially described by Rosai and Dorfman in 1969.[1] Many characteristics of this disease remain unknown, restricting early diagnosis and proper treatment. Various theories have been implicated as an etiology; however, there is no strong evidence for any of them.[2]

2. Case Report

A 23 year old woman visited surgery OPD with 6 months history of a painless swelling in right axillary region. The lesion progressively increased in size and was about 1.5 × 1.5 cms. There was no history of fever, weight loss, pallor. On examination there was no other lymphadenopathy.

The patient was advised FNAC. FNAC smears were stained with Giemsa stain. The Giemsa stained smears showed a polymorphous picture with presence of predominantly histiocytes admixed with a variable population of lymphocytes, plasma cells, neutrophils and few eosinophils. The histiocytes had moderate to abundant cytoplasm, large vesicular nuclei and showed phagocytosis (emperipolesis) of lymphocytes. At places these histiocytes were binucleated and multinucleated. There was no atypia. (Figures 1, 2, 3 and 4)

The final Cytological diagnosis was given as Sinus Histiocytosis with massive Lymphadenopathy and patient was asked to remain in follow up. Patient was not given any treatment and lymph node started decreasing in size on its own and disappeared in three months.

3. Discussion

Sinus histiocytosis with massive lymphadenopathy (SHML), also known as Rosai-Dorfman disease (RDD) is a rare self-limited disease. The etiology of the disease is unknown but several theories have been suggested. Some infectious agents have been suspected, including Epstein-Barr virus, Parvovirus B19, Herpes virus type 6 and 8, and Polyomavirus. A relationship with Klebsiella, Brucella and Cytomegalovirus was also suggested, but any

* *Corresponding author.*
E-mail address: drvineetbanga@rediffmail.com (V. Banga).

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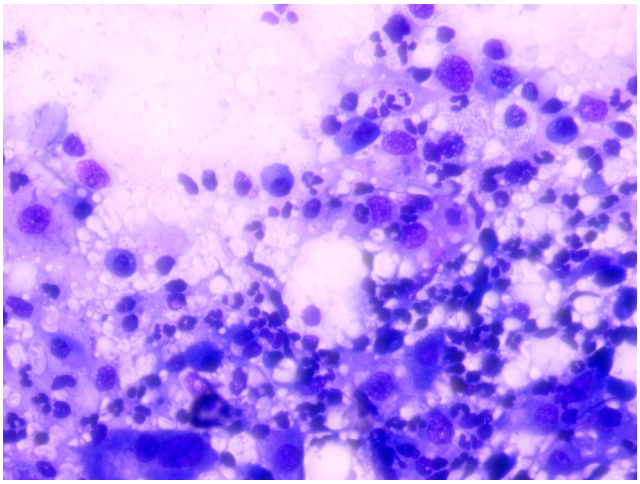

Fig. 1: Microphotograph showing polymorphous picture with presence of predominantly histiocytes admixed with a variable population of lymphocytes, plasma cells and neutrophils. (Giemsa stain, X 10 Magnification)

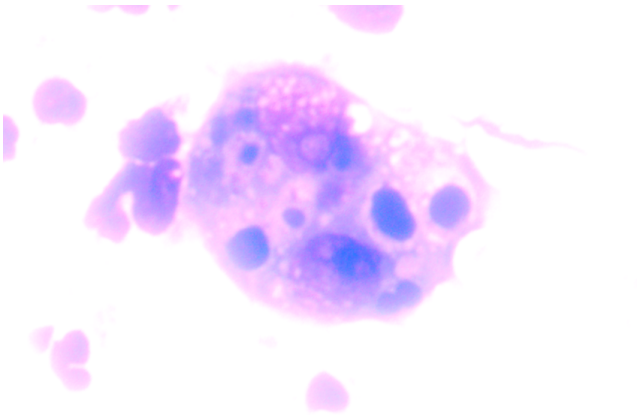

Fig. 2: Microphotograph showing Emperipolesis. (Giemsa stain, X 40 Magnification)

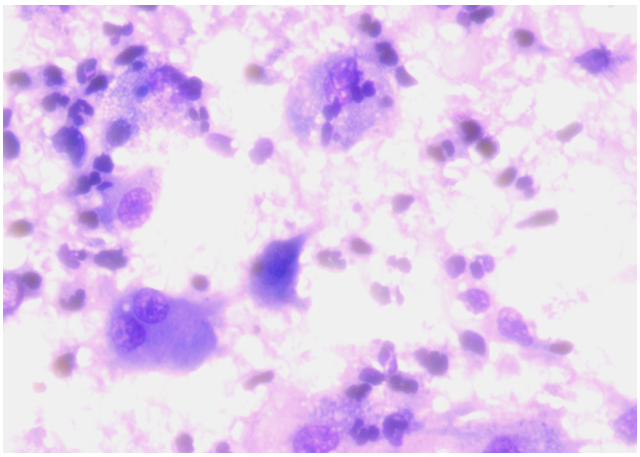

Fig. 3: Microphotograph showing binucleated histiocytes and emperipolesis. (Giemsa stain, X 40 Magnification)

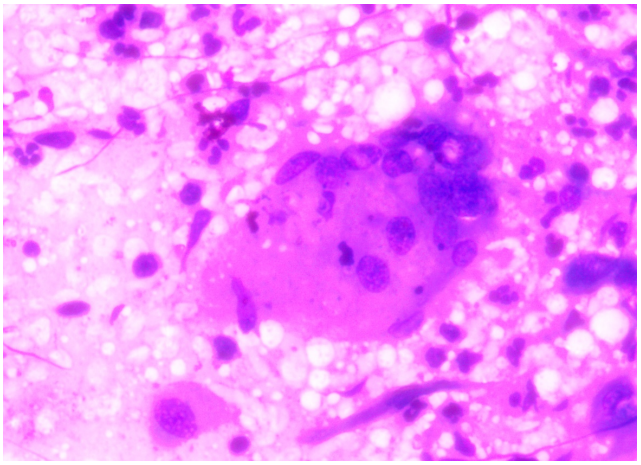

Fig. 4: Microphotograph showing multinucleated histiocytes. (Giemsa stain, X 40 Magnification)

attempt to isolate the organisms consistently failed.[3]

The presentation of the disease also varies with site of involvement. Commonly it presents with massive cervical lymphadenopathy. But extra nodal involvement has also been documented in around 40% of cases, the most common being skin followed by upper respiratory tract and bone.[2,4,5]

Foucar et al[6] reviewed the clinical and pathologic features of 423 examples of sinus histiocytosis with massive lymphadenopathy entered in a case registry, with special emphasis on extra nodal manifestations of the disease. The most common sites of extra nodal involvement are skin, upper respiratory tract, and bone. However, SHML also can occur in a variety of other sites, including the genitourinary system, lower respiratory tract, oral cavity, and soft tissues. Involvement of kidney, lower respiratory tract, or liver was found to be a poor prognostic sign, and patients with associated immunologic disease often fared poorly. In general, prognosis has been found to correlate both with the number of nodal groups and with the number of extra nodal systems involved by SHML.

RDD is notable for its varied clinical presentations which evoke a wide differential diagnosis. Although correlation of clinical presentation with radiologic and laboratory values is very helpful, the pathologic assessment is pivotal in making the diagnosis. The classic histology is characterized by effacement of nodal architecture and dilatation of lymph node sinuses by lymphocytes, plasma cells and numerous characteristic histiocytes with large vesicular nuclei and abundant clear cytoplasm. Many of these histiocytes, also known as RDD cells, contain intact lymphocytes, and sometimes plasma cells and red blood cells, within their cytoplasm. This process whereby cells enter and transit through a cell evading cellular degradation is known as emperipolesis and was first described by Humble et al.[7,8]

Das et al[9] concluded that FNA cytology is a useful tool in the diagnosis of SHML. FNA smears showed

numerous histiocytes with evidence of lymphophagocytosis (emperipolesis) against a background of reactive lymphoid cells. All the classical cytomorphological features described were found in our case.

Kumar et al[10] concluded that FNAC is a useful and reliable tool for the diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) and biopsy can be avoided in these patients, thereby reducing patient inconvenience and health care cost.

Kushwaha et al[11] described that the cytomorphology of Rosai–Dorfman disease is so distinctive that it can be diagnosed by FNAC. FNAC is a reliable and sensitive means to establish conclusive diagnosis, obviating the need for biopsy.

4. Conclusion

Rosai-Dorfman disease must be considered in the differential diagnosis of young patients who exhibit massive or multiple lymphadenopathies, especially when involvement of the cervical area occurs. The disease can be diagnosed by FNAC that is reliable, convenient, sensitive test for diagnosis and prevents surgical procedure therefore reduces patient inconvenience and health care cost.

5. Conflict of Interest

The authors declare no relevant conflicts of interest.

6. Source of Funding

None.

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Author biography

Vineet Banga, Senior Specialist & HOD Pathology https://orcid.org/0000-0002-7412-6007

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