



Rosai-Dorfman Disease of Extranodal Site

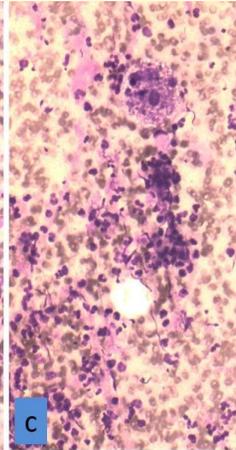
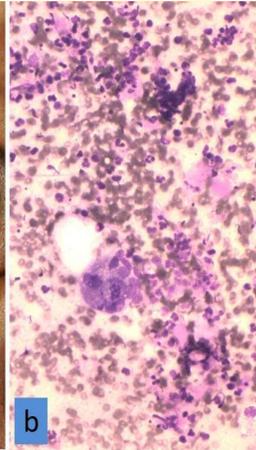
Dr. Seema ,Dr Atin Singhai ,Dr Suresh Babu

Department of Pathology KING GEORGES MEDICAL UNIVERSITY LUCKNOW,UTTAR PRADESH,INDIA



BACKGROUND

- Rosai-Dorfman disease or sinus histiocytosis with massive lymphadenopathy (SHML), is a benign self-limiting disorder that commonly involves the lymph nodes and can metastasize to other parts of body via lymphatic system.
- There are only few cases on the fine-needle aspiration (FNA) cytologic features of this entity.
- It mainly affects the younger population with slight male predominance.
- Extranodal sites is seen in about one fourth of all cases. Extranodal sites include skin, subcutaneous tissue, bone, skeletal muscle, thyroid, liver, kidney, heart, uterine cervix, nasal cavity, submandibular gland, breast, parotid gland, larynx, temporal bone, pterygoid fossa, meninges and orbital region.



DISCUSSION

- Rosai-Dorfman disease is a rare but well-defined histiocytic proliferative disorder of unknown etiology and is characterized by non-malignant proliferation of distinctive histiocytic/phagocytic cells within lymph node sinuses and the lymphatic system in extranodal sites..
- Extranodal disease is documented in 30–40% of patients. The most commonly affected extranodal sites include the skin and soft tissue, the upper respiratory tract and bone followed by the genitourinary tract, the lower respiratory tract, the oral cavity, the gastrointestinal tract, the orbit and the testes.
- The hallmark of these histiocyte is the presence of a variable number of intact lymphocytes within the cytoplasm of the cell, which is a phenomenon referred to as lymphophagocytosis or emperipolesis. The histological differential diagnosis includes Langerhans cell histiocytosis, histiocytic sarcoma, storage disorders such as Gaucher's disease, classical Hodgkin's lymphoma, metastatic melanoma and carcinoma, as well as infections caused by Histoplasma and mycobacteria that involve the lymph nodes.

CASE REPORT

- We present a case of 28 year old male with history of recurrent left preauricular swelling which had persisted for one year. (Figure 1a) The patient had no history of fever, tuberculosis or loss of body weight.
- On examination diffuse, tender and movable 3x3 cm preauricular ill defined swelling was found. There was a cystic, raised reddish area over the centre of swelling. There was no lymphadenopathy in the cervical or other sites. Fine needle aspiration cytology (FNAC) was performed one year before and diagnosis of pyogenic abscess was made.
- Sonography of region showed solid hypoechoic, hypervascular lesion with few central area showing necrocystic changes with dense internal echoes. Aspiration was re-performed from the same site and smears revealed high cellularity with prominent mixed inflammatory infiltrate comprising of polymorphs, lymphocytes, plasma cells as well as few eosinophils. There was also evidence of fair number of singly scattered as well as multinucleated histiocytes with abundant pale blue cytoplasm containing lipid droplets and vesicular nuclei. These histiocytes also demonstrated emperipolesis in form of phagocytosis of lymphocytes, polymorphs and plasma cells at places. (Figure 1b,c) Acid fast bacilli and Gram's staining were also performed which showed negative results.

TAKE HOME MESSAGE

- Due to fewer number of cases reported in the literature this entity should be considered in the differential diagnosis with Langerhans cell histiocytosis, histiocytic sarcoma, storage disorders such as Gaucher's disease, classical Hodgkin's lymphoma, metastatic melanoma and carcinoma, as well as infections caused by Histoplasma and mycobacteria that involve the lymph nodes.

REFERENCES

1. Deshpande V, Verma K. Fine needle aspiration (FNA) cytology of Rosai-Dorfman disease. *Cytopathology* 1998;9:329–335.
2. Das DK, Gulati A, Bhatt NC, Sethi GR. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Report of two cases with fine needle aspiration cytology. *Diagn Cytopathol.* 2001 Jan 1;24(1):42-5.
3. Kumar B, Karki S, Paudyal P. Diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) by fine needle aspiration cytology. *Diagn Cytopathol.* 2008 Oct 1;36(10):691-5.
4. Foucar E, Rosai J, Dorfman R: Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. *Semin Diagn Pathol* 1990;7:19–73.