Pediatric Congenital Hematologic
Disorders Research Center
مرکزتحقیقات بیماری های خونی مادرزادی کودکان

In the name of God

Sinus histiocytosis

with

massive lymphadenopathy

(Rosai Dorfman disease)

BIBI SHAIN SHAMSIAN

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Rosai Dorfman disease

- **Rosai-Dorfman Disease (RDD) or Sinus Histiocytosis with massive lymhadenopathy (SHML):**
- **Rare, benign proliferative disorder of histiocytes,**
- **sometimes showing familial incidence**

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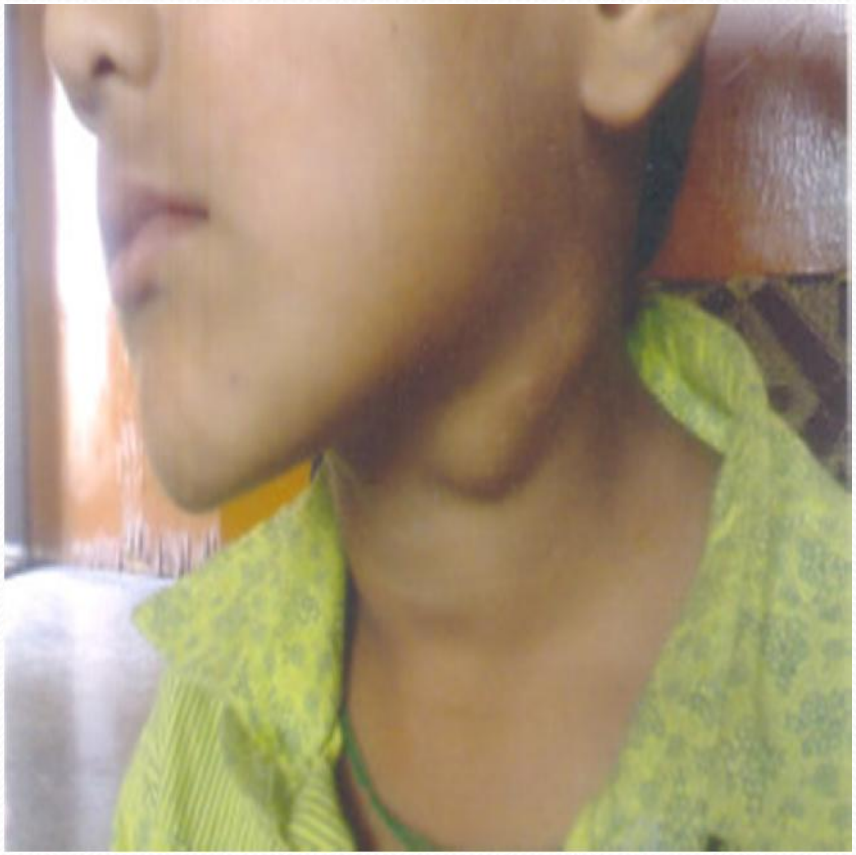
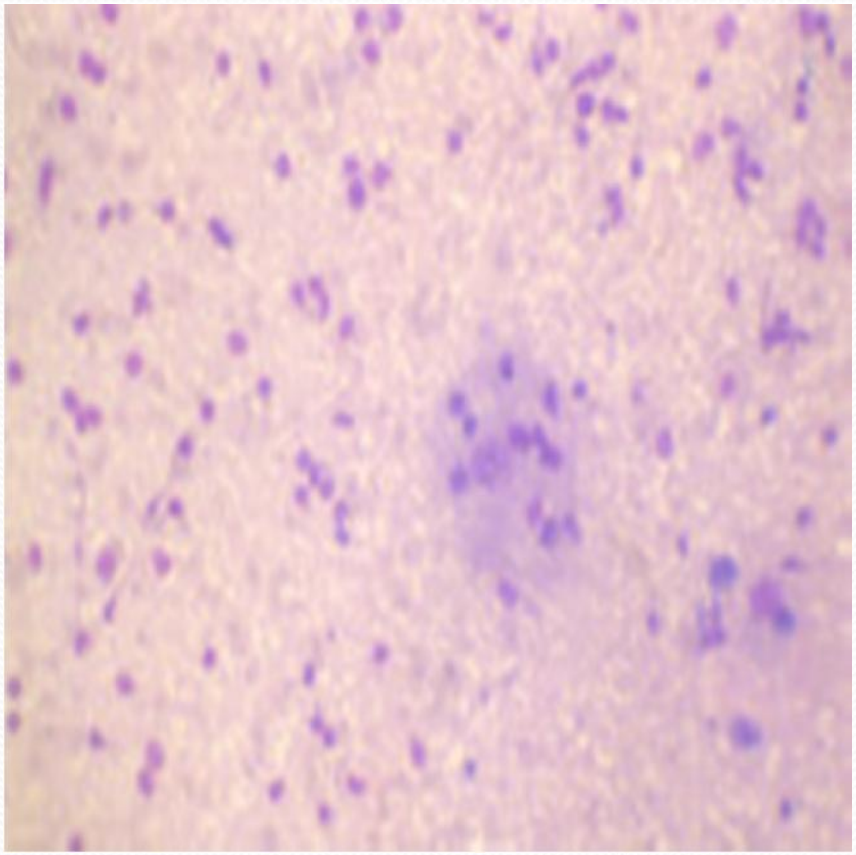
Clinical Photograph of a Child Showing Massive Cervical Lymphadenopathy





Case Report

- **Male, 14 years old**
- CC: bilateral large cervical mass, low grade fever
- PH: **Massive bilateral cervical lymphadenopathy, size more than 5× 5cm**
- Lab tests: **WBC: 14 800** Hb: 13 MCV: 86
- Plat: 301000
- PMN:86% L: %7 Mono: 1 5 E: %1
- **ESR: 45**

Case Report

- U/acid: 4.2
- **LDH: 283**
- **Ferritin: 80**
- LFT: Nl
- Echo : Nl
- virology :Nl **EBV IgG: 59** **IgM:0.2**
- Bio : NL
- Toxo titer IgG: 0.5 IgM: 0.3
- CXR: NL
- Abdominal Sono: Nl Spleen : 84 mm Liver: 105 mm

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Case Report

- Bone survey: NL, adenoied is prominant
- **CT: enlargment LN are seen in bilateral all neck chains(Sub mandibular, supraclavicular, posterior triangle)**
- ECHO: EF: 60%
- **Left cervical ,LN Biopsy: 19.1.92**
- **Pathology Rosi Dorfman syndrome (27.1.92)**
-
- **IHC: is not ready**

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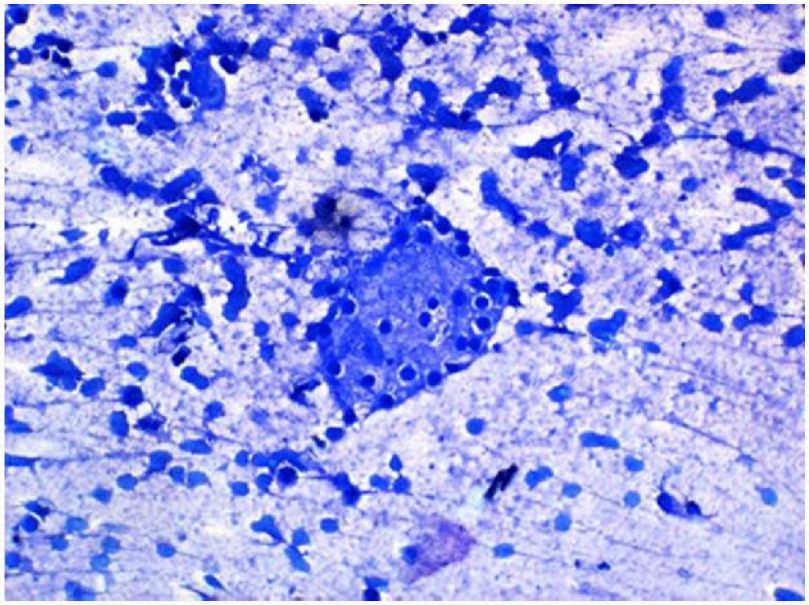
Smear showing histiocyte with engulfed intact lymphocytes (emperipolesis) –Giemsa, X400



Madhusmita Jena. **Diagnosis of Rosai-Dorfman Disease by Fine Needle Aspiration Cytology in a Case with Cervical Lymphadenopathy and Nasal Mass.** ***Journal of Health and Allied Sciences.*** **India, 2011. Case 2**

- 46 y /male : blockade of nose on the left side since 6 months & Multiple bilateral large cervical lymph nodes, The largest one measured 1.0x0.5 cms.
- FNA , then excisonal biopsy : A diagnosis **of reactive lymph node** was made initially.
- However a review of the Pap stained smears : few histiocytes with vesicular nuclei and abundant pale cytoplasm. The cytoplasm of the histiocytes exhibited intact engulfed lymphocytes (emperipolesis) within them.
- **Diagnosis: RD**

SS Bist.Rosai Dorfman Syndrome with Extranodal Manifestations.
India .2007. Case 3

- 22-y/ male :bilateral, massive, painless neck swelling for past six months. & gradually increasing nasal obstruction on right side with occasional episodes of epistaxis for past 3 mo.
- surgery for nasal mass / general anesthesia /mass was excised endoscopically.
- oral prednisolone 60 mg / day /8 weeks and then tapered off

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Rosai-Dorfman disease: unusual cause of diffuse and
case 4.2005**K S SODHI**.massive retroperitoneal lymphadenopathy

- 34-y/ male diffuse non colicky abdominal pain, with bilateral, cervical lymphadenopathy and fever for 2 months.
- bilateral large, mobile and non tender-cervical lymphadenopathy, along with axillary and inguinal lymphadenopathy.
- The abdomen was soft with mild hepatomegaly.
- CT :multiple enlarged homogeneously enhancing lymph nodes in the pre-vascular space paratracheal regions, aortopulmonary window, and subcarinal region
- , mild hepatomegaly with multiple enlarged homogeneous lymph nodes.
- FNA cytology from a retroperitoneal lymph node

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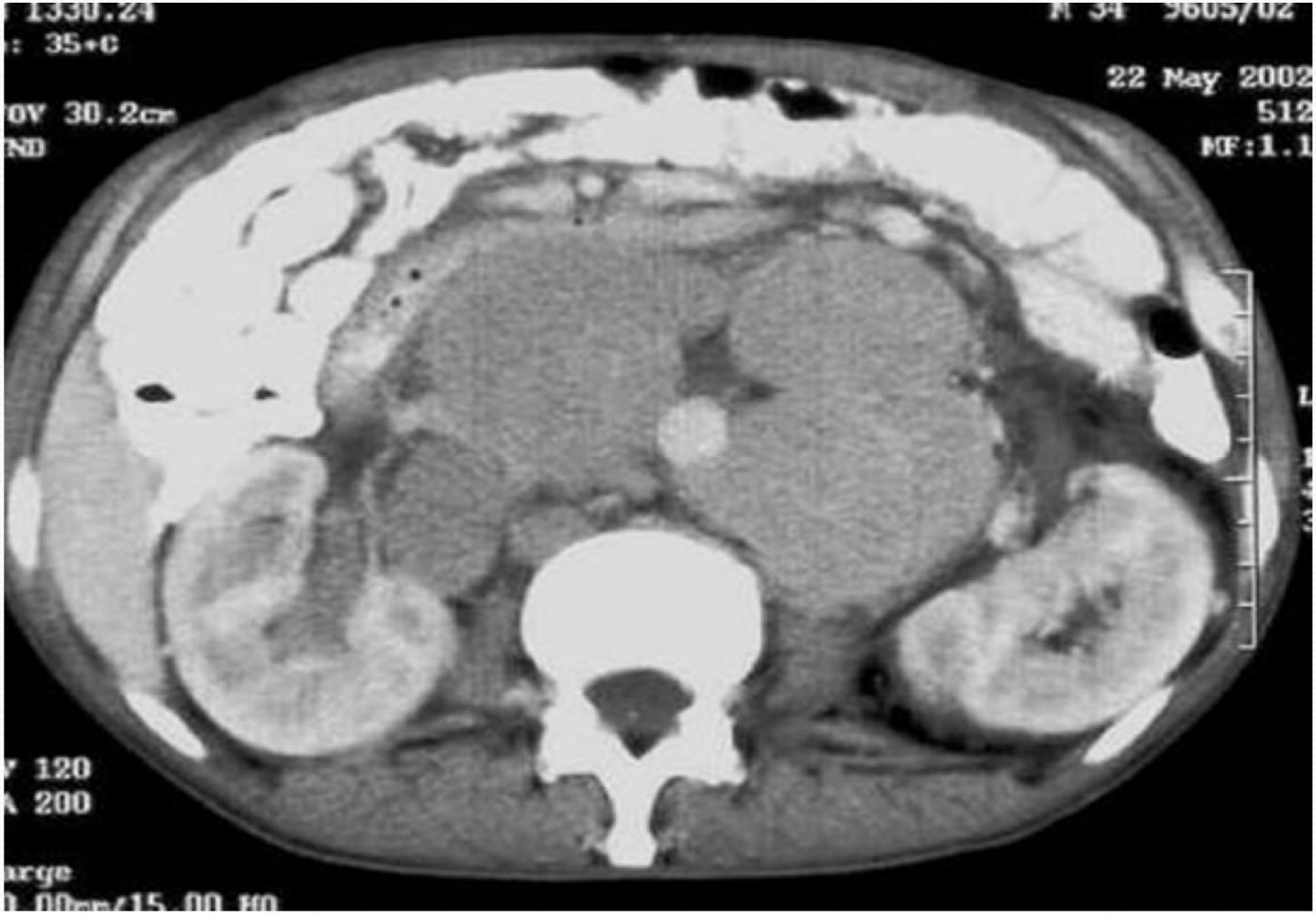

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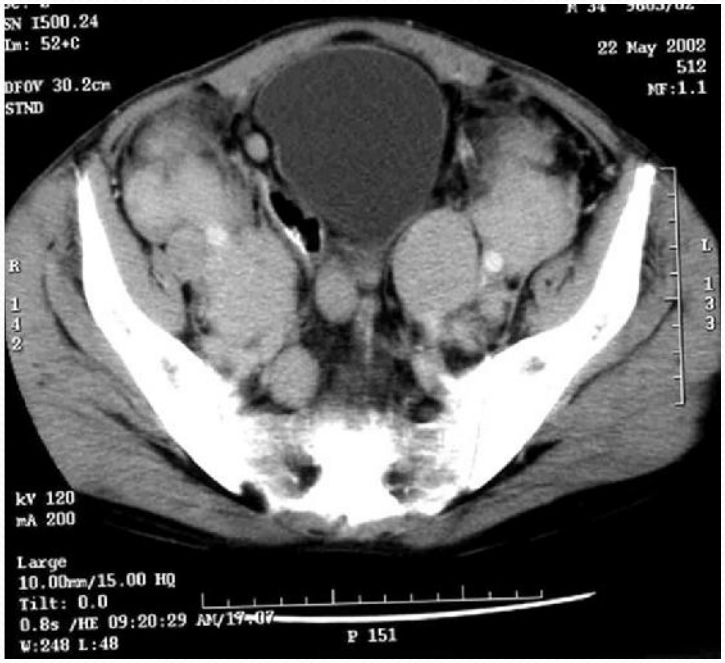
CT of the abdomen (pelvis) showing bilateral enlarged iliac lymph nodes.



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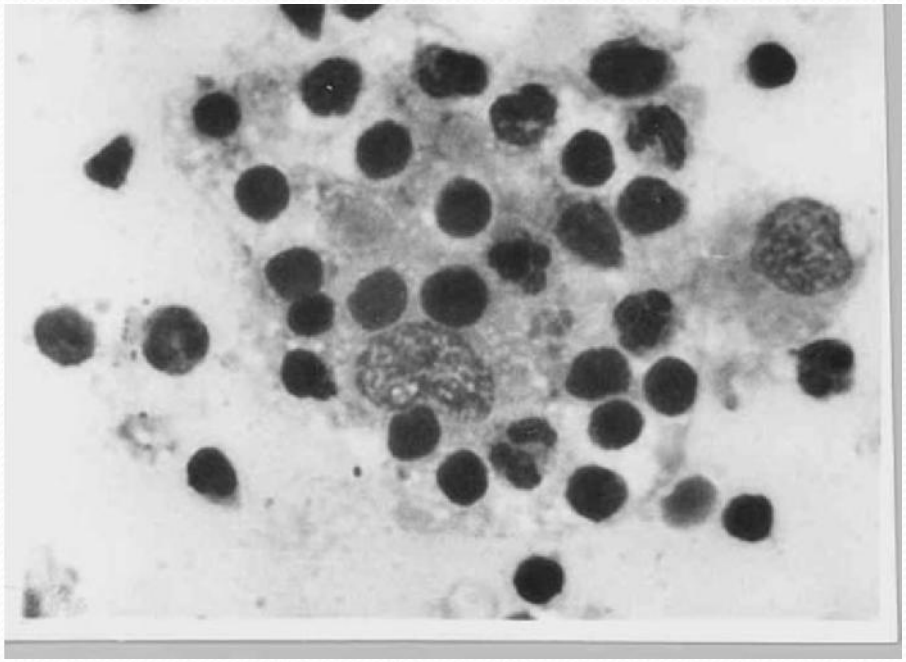
A histiocyte depicts lymphophagocytosis (Emperipolesis)



Rosai-Dorfman Disease Manifesting as Relapsing Uveitis and . Subconjunctival Masses

Case 5. **Case 5.** Hsin-Yuan Tan

- a 63- year-old man with SHML with unusual ophthalmic manifestations of relapsing uveitis and
- bilateral subconjunctival masses.
- PE: bilateral subconjunctival infiltrative lesions were noted
- DD? lymphoma,
- Eye involvement is relatively uncommon (8.5%)
- Uveitis is an even more rare presentation
- conjunctival biopsies as well as systemic investigations

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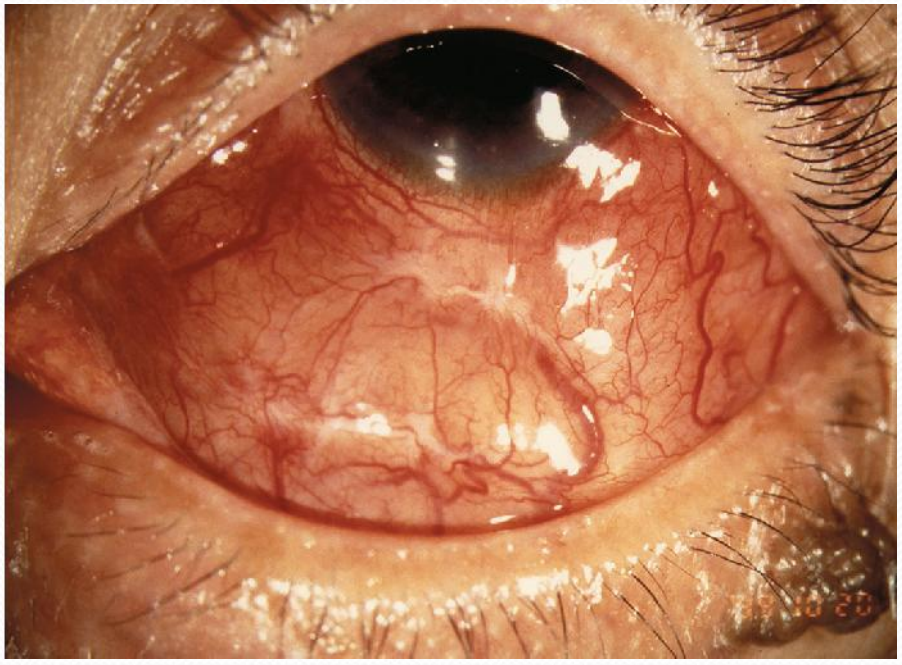
Salmon-colored lesion is present beneath the conjunctiva of his left eye



Review Article- RDD

- It was recognized as a distinct clinicopathologic entity by Rosai and Dorfman in 1969.(4 cases)
- 1972 : 30 cases
- as awhole : 46 case (2011)

- It is characterised by expansion of sinuses of the lymph nodes and the lymphatics of extranodal sites by proliferation of histiocytes with abundant pale eosinophilic cytoplasm containing engulfed lymphocytes.

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RDD–Presentation

- First & second decades of life (81%),
- 2:1 male/ female ratio
- Most common presentations :painless bilateral cervical lymphadenopathy (90% of the patients),
- Axillary (23.7%), inguinal (25.7%), and mediastinal (14.5%)
- 25 to 40 % (43%) of the cases : at least one site of extra nodal involvement.
- skin and soft tissue, upper respiratory system, GU tract, eye, orbit, kidney, thyroid, breast
- **skin (27.4%), nasal and paranasal cavities (26.8%), subcutaneous tissue (22.2%), orbit and eyelids (20.1%) and bone (18.4%).**

Extranodal involvement- RDD

- In general, extranodal involvement does not determine a more aggressive character or poor outcome

However:

- Generalised lymphadenopathy,
- Extranodal involvement of multiple organs (kidney, lungs and liver)
- Immunologic alterations

lead to a poor prognosis.

CNS & Ocular involvement- RDD

- Central nervous system can be affected.
- Suprasellar involvement-mimicking meningioma has been reportedent,
- Ocular involvement is rare.
- In the largest series of 243 cases reported by

Foucar in 1990, the eye was involved in 36 cases (8.5%) and was highly associated with nasal sinuses involvement

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Chang Gung Med J Vol. 25 No. 9
Rosai-Dorfman Disease Manifesting as Relapsing Uveitis and Subconjunctival Masses. Taiwan. 2002.

Soft tissue of the orbit and the eyelids infiltrations were the most common manifestations of ocular involvement.

In addition, infiltration of the lacrimal system, conjunctiva/subconjunctiva, cornea, uveal tract, and optic nerve, have all been reported.

Uveitis was present in six cases as found in a literature reviewed

Etiology-RDD

- Although the aetiology remains unknown, the disease is thought to be a disorder of immune regulation or response to a presumed infectious agent (**HHV-6 / EBV**) with its major manifestation in lymph nodes with resultant proliferation of sinusoidal histiocytes.

RDD-Findings

- **Findings:leucocytosis, elevated ESR& anemia,hypergammaglobulinemia**
- **Association with autoimmunedisease and malignancy**

- The characteristic cytomorphology of this entity is the presence of **large histiocytes with abundant cytoplasm having variable number of intact lymphocytes within it**; a phenomenon referred to as lymphophagocytosis or **emperipolesis.**

IHC- RDD

- IHC:immunostaining show positivity for **S100 protein, CD14, CD33 & CD68 , Cd11 C** in cytological smears. **CD1 a is negative**

- Extranodal involvement is seen in up to 40% of cases which show similar morphological features to its nodal counterpart although **fewer histiocytes with emperipolesis , and more fibrosis are encountered.**

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Diff Diagnosis- RDD

- **The main differential diagnosis on FNAC of the lymph nodes include:**

- **Reactive lymphoid hyperplasia with sinus histiocytosis,**
- **Malignant histiocytosis,**
- **lymphoma**
- **Tuberculosis**
- **LCH**

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RRD-Treatment

- Nonspecific and include:
- corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low dose interferon, antibiotics therapy, radiation therapy and surgical treatment with partial or total resection.

- Surgical option may be reserved for compressive symptoms, like airway obstruction, neurologic or ocular compressions, or severe deformation.

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Conclusion

Massive cervical lymphadenopathy is the hallmark of Rosai Dorfman syndrome and head Neck region is the preferred site of the extranodal form of disease.

The diagnosis of RDS is made on the basis of clinical suspicion and confirmed by histopathology.

Clinicians and pathologists should always be aware of RDS in making a differential diagnosis of lymphadenopathy