

Cutaneous-Only Disease?

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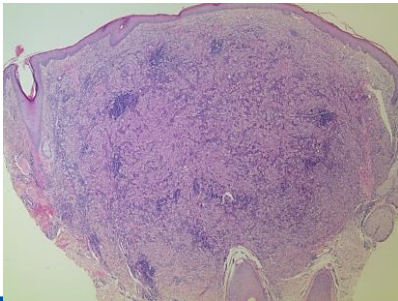
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2004

- 44 year old male with a "cyst" on his right cheek

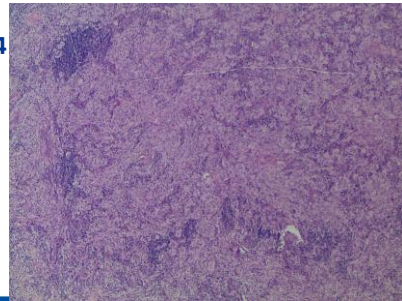
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2004



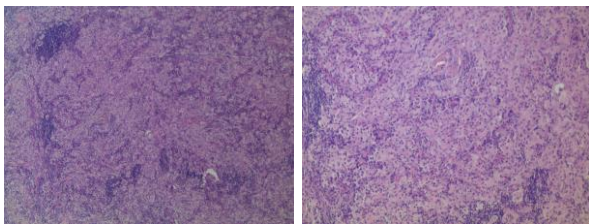
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2004



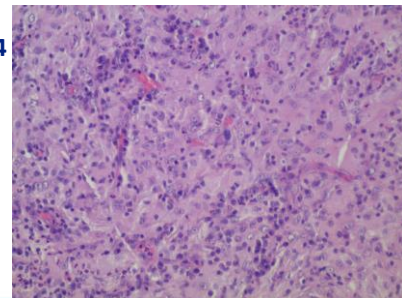
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2004



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2004



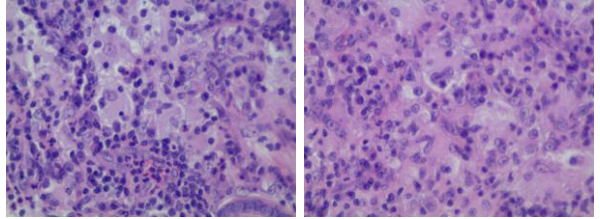
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Differential Diagnosis

- Folliculitis
- Rosacea
- Xanthoma
- Histiocytosis
- Histiocytoma
- Xanthogranuloma
- Infection

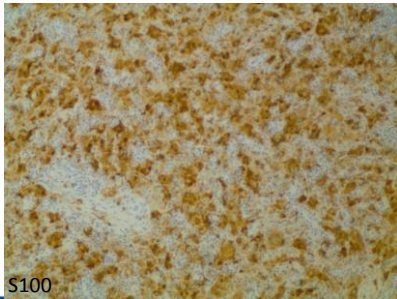
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2004



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2004



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Diagnosis - 2004

- Rosai-Dorfman Disease (RDD)

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Question 1

- I have seen/treated Rosai-Dorfman Disease
 - A. Yes
 - B. No
 - C. Maybe (Suspected)
 - D. Don't know

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Rosai-Dorfman Disease

- "Sinus histiocytosis with massive lymphadenopathy"
- Non-neoplastic or reactive proliferative process, composed of histiocytes with features activated macrophages
- Investigated etiologies:
 - Human herpesvirus-6
 - Other viral (EBV, parvovirus B19)
 - IgG4

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Rosai-Dorfman Disease - Clinical

- Usually presents in late childhood/early adulthood, male predominance
- Lymphadenopathy
 - Usually cervical (87%), massive and bilateral, painless
 - Inguinal (26%), axillary (24%), mediastinal (15%)
- Extranodal disease
 - Skin, soft tissue, nasal cavity, sinuses, CNS, bone

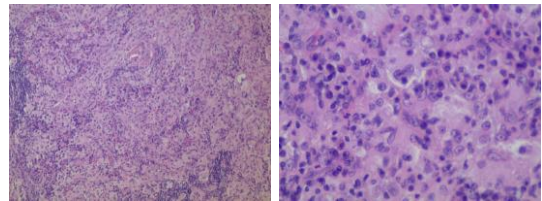
Rosai-Dorfman Disease - Clinical

- Signs and Symptoms
 - Fever, night sweats, weight loss, maculopapular eruptions
 - Anemia, polyclonal hypergammaglobulinemia, elevated ESR
- Rare associated disease
 - Immune-mediated (glomerulonephritis, autoantibodies, recurrent infection)
 - H Syndrome, Lymphoma, Langerhans Cell Histiocytosis

Question 2

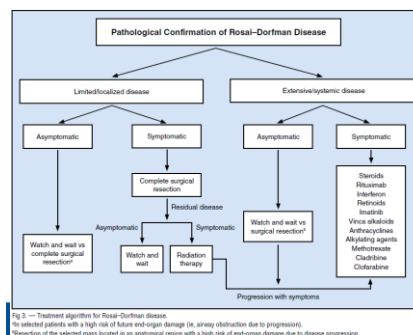
- What percentage of patients with RDD have extranodal involvement?
 - A. < 13%
 - B. 23%
 - C. 43%
 - D. 73%

Rosai-Dorfman Disease - Histopathology



Rosai-Dorfman Disease – Work-up and Treatment

- Clinical work-up varies depending on sites of involvement
- Most cases require no treatment
 - 20-40% have spontaneous resolution
 - Remainder indolent, stable but persistent
- Less than 10% of patients have progressive or “aggressive” disease
- If treatment necessary:
 - Excision
 - Radiation
 - Steroids
 - Chemotherapy



Datta S, Sagarika C, Sankar L, et al. Rosai-Dorfman disease: basic biology, clinical features, pathogenesis, and treatment. *Cancer Control* 2014;21:322-327.

Our Patient

- Differential:
 - Cutaneous RDD
 - Conventional RDD: Nodal disease with extranodal lesions
 - Disseminated

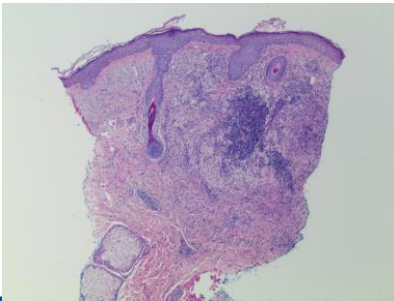
More Patient History

- Patient seen by Dermatology. No known or documented lymphadenopathy.
- 2012: Left Cheek lesion – R/O BCC
- 2013 and 2015: Right Cheek lesions – RDD vs. BCC vs. Rosacea

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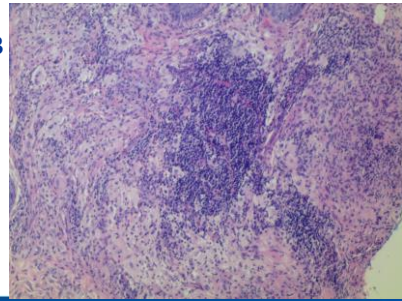
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2013



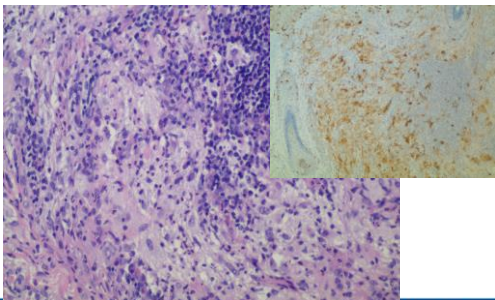
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2013



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2013



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2013

- Rosai-Dorfman Disease
 - Compatible with Cutaneous RDD

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Question 3

- What percentage of patients have ONLY extranodal disease (with no lymph node involvement)?
 - A. <15%
 - B. 25%
 - C. 35%
 - D. 45%

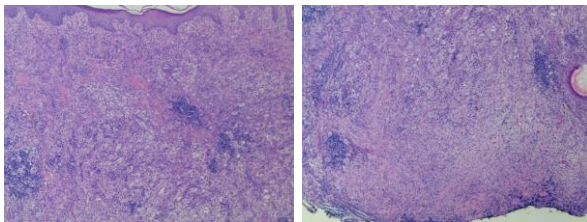
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More Patient History - 2016

- Perianal/Perineal lesion, intermittently draining
- No lymphadenopathy
- Inflamed, possibly abscess, clinical differential of hidradenitis suppurativa

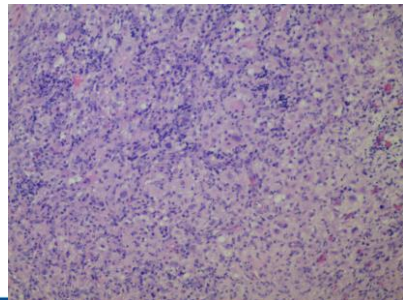
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2016



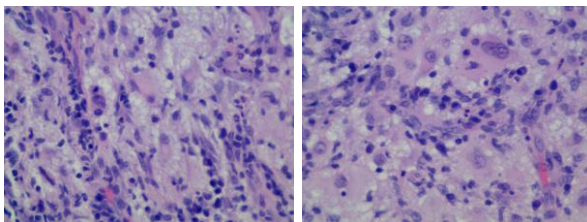
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2016



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2016



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Cutaneous Rosai-Dorfman Disease

- Considered a distinct clinical entity
- Slightly older patient population (5th decade), slight female predominance
- Papules and nodules
 - Purple or brown, erythematous
 - Clustered or multiple foci
- No lymphadenopathy or visceral involvement

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Cutaneous Rosai-Dorfman Disease

- Distinction between cutaneous RDD and RDD with extranodal skin involvement is not possible on morphologic grounds
- Clues to cutaneous RDD:
 - Lesional histiocytes in vascular spaces
 - Storiform growth pattern

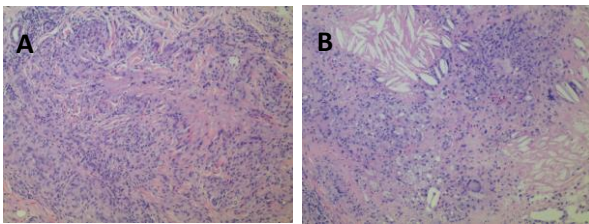
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Summary

- Rosai-Dorfman Disease benign, idiopathic histiocytic proliferation
- Broad initial clinical and histologic differential diagnosis
- Classification of disease requires clinicopathologic correlation and follow-up
 - Classic RDD with lymphadenopathy +/- extranodal disease
 - Extranodal RDD
 - Cutaneous RDD
 - RDD with latent lymph node involvement

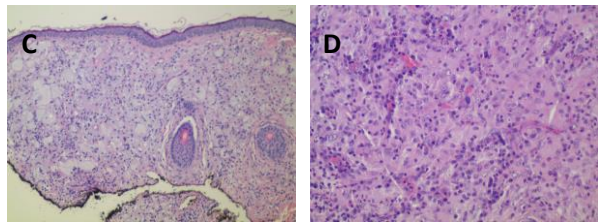
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Question 4 – Which is RDD?



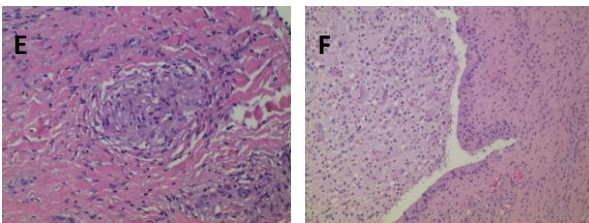
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Question 4 – Which is RDD?



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Question 4 – Which is RDD?



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