Cutaneous-Only Disease?

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2004

- 44 year old male with a “cyst” on his right cheek
Differential Diagnosis

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

2004

Diagnosis - 2004

- Rosai-Dorfman Disease (RDD)

Question 1

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

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
**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%)
- Extraneural 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

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

- Rosai-Dorfman Disease
  - Compatible with Cutaneous RDD
Question 3

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

More Patient History - 2016

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

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
**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

**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

**Question 4 – Which is RDD?**

A  
B  
C  
D  
E  
F

**References**