Rosai-Dorfman in the Submandibular Salivary Glands of a Pediatric Patient

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INTRODUCTION
- Sinus histiocytosis with massive lymphadenopathy (SHML or Rosai-Dorfman disease) is a painless enlargement usually of the cervical lymph nodes, but has rarely been reported as presenting in the salivary glands. We describe a case of a pediatric patient with submandibular disease and present an update on management and surgical indication.

CASE
HPI
- A previously healthy 11 year old African-American male presented with a 1 year history of painless bilateral neck swelling.

MEDICAL HISTORY:
- No past medical, surgical, contributory family history, medications or allergies reported. The patient is a sixth grade student.

REVIEW OF SYSTEMS:
- Negative for constitutional B symptoms, pain or compressive symptoms.

PHYSICAL EXAMINATION
- Vital Signs: afebrile and stable
- General: Well developed, well nourished child, NAD
- Neuro: CN II- XII grossly equal and intact
- Head: Normocephalic
- Eyes: PERRLA and EOMI
- Ears: No effusions, normal pinnae and ext. auditory canal
- Nose: Marked septal deviation, nasal congestion and turbinate hypertrophy
- Oropharynx: 2+ tonsils
- Neck: Extremely firm, enlarged, bilateral submandibular glands at neck level 2 without erythema or tenderness
- Lymph: no generalized lymphadenopathy
- Heart, Lung and Abdomen: normal

CT FACE AND NECK
- Moderate to marked enlargement of the submandibular glands bilaterally with evidence of sialadenitis vs. sialodenosis.
- Enlargement of the bilateral parotid glands.
- Mildly enlarged lymph nodes in the jugulodigastric chains and the posterior triangular spaces

LABORATORY
- WBC 6.25 cells/µL (normal differential, lymphocyte subpopulations not consistent with ALPS)
- Hemoglobin 13.2 g/dL
- Platelet 314,000
- ESR 7
- CRP <0.05

DISCUSSION
- Rosai-Dorfman disease is a rare clinical entity characterized by benign pseudolymphomatous proliferation with significant histiocytic infiltration
- The most common presentation is painless bilateral cervical adenopathy
- Extra nodal involvement can occur in up to half of patients
- Three sites: Eyes and eyelids
- Oral cavity
- Salivary glands
- Panikar et al described a case of a 45-year old woman with unilateral submandibular gland enlargement with ipsilateral adenopathy
- Fine needle aspiration demonstrated moderated cellularity, with large histiocytes dispersed in background of intense lymphoplasmacytic infiltrate; diagnosis of SHML was made
- No further treatment was pursued
- There is no consensus on treatment guidelines for this benign disease

PROCEDURE #1:
RIGHT SUBMANDIBULAR GLAND AND MASS + LYMPH NODE EXCISION

Pathology:
- Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease)
- Both sites show infiltration with large histiocytes and a mixed inflammatory background consisting of lymphocytes and plasma cells. This process replaces the salivary gland lobules and the lymph node sinuses and interfollicular zones.

PROCEDURE #2:
LEFT SUBMANDIBULAR GLAND AND MASS EXCISION

Pathology:
- Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease)
- Post-operatively the patient developed further enlargement of the left submandibular gland with potential airway impingement, feeding difficulty and tenderness.

HISTOLOGY

CONCLUSIONS
- This case is a rare presentation of Rosai-Dorfman disease in the submandibular salivary glands of a pediatric patient
- Management of this histologically benign disease should focus on compressive symptoms, which can potentially be lethal