Southern Medical Association

A Case of Rosai Dorfman Disease with Intracranial Extra-nodal Involvement

ONC-3

The following abstract will be presented at the Southern Medical Association Annual Scientific Assembly, October 30-November 1, 2014 in Destin, Florida.

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Objectives

Upon completion of the lecture, attendees should be better prepared to:

1) Identify and understand benign causes of intracranial tumors that may not require surgical treatment.
2) Identify RDD with further emphasis of prevalence of Extranodal form.
3) Steroid-responsiveness of Extra-nodal RDD.

Abstract

Introduction: Rosai Dorfman Diseasei is a Sinus Histiocytosis that commonly presents w/ massive painless lymphadenopathy. Extra-nodal involvement is noted in 40% of cases such as skin, GI, bone, soft tissue, kidneys and more. Less than 5% of reported cases display central nervous lesions. Of all the CNS cases, the lesions involving the Dura matter are more common thereby implicating the diagnosis as it mimics Meningiomas radiologically.

Case Presentation: 28 y/o AAF with no PMHx presented with headaches, episodic blurred vision and tinnitus. Physical examination was unremarkable. Imaging studies revealed a extracranial and intracranial mass measuring 3.0 centimeter with the extent along the dural surface measuring approximately 4 centimeter w/ associated lymphadenopathy. At this time, the patient denied any intervention. She re-presented, a year later, with worsening headaches and an enlarging scalp lesion. Physical examination was remarkable for a right parietal lesion that was described as soft, nontender, semi-fluctuant mass with no other masses or lymphadenopathy. She underwent a Craniotomy w/ excisional biopsy. Final pathology was consistent with Extra-nodal RDD of Soft tissue.

Final Diagnosis: Immunohistochemical studies showed lesions displaying numerous large, foamy histocytes with centrally placed nuclei, and intracytoplasmic lymphocytes that are uniformly positive with S-100 protein immunostain consistent with Extranodal RDD.

Management/Outcome/F/U: 50% of RDD cases have spontaneous regression, but this has not been reported in intracranial cases. She was started on short-term steroids w/ near-resolution of the mass (radiological) and symptoms.

Disclosure

Thankam Nair, MD – No Relevant Financial Relationships to Disclose
Charmi D. Patel, MD – No Relevant Financial Relationships to Disclose
Siva Yadlapati, MD – No Relevant Financial Relationships to Disclose

Unlabeled Use Disclosure: Unlabeled use of steroids as a treatment modality based off previous case reports.