Follicular Dendritic Cell Sarcoma Presenting in the Submandibular Region in an 11 Year-Old
Amanda L. Silver, MD,1 William C. Faquin, MD, PhD,2 Paul A. Caruso, MD,3 and Daniel G. Deschler, MD1
1 Dept. of Otolaryngology, Division of Head and Neck Surgery, Massachusetts Eye & Ear Infirmary, Boston, MA
2 Dept. of Pathology, Massachusetts General Hospital, Boston, MA 3 Dept. of Radiology, Massachusetts Eye & Ear Infirmary, Boston, MA

OBJECTIVES

• To review the differential diagnosis of follicular dendritic cell sarcoma (FDCS) and to highlight key features differentiating FDCS from other poorly differentiated neoplasms in order to enable prompt diagnosis and appropriate treatment.
• To develop a management approach to the uncommon extranodal follicular dendritic cell sarcoma of the head and neck.

INTRODUCTION

Follicular Dendritic Sarcomas (FDCS), formerly known as follicular dendritic cell tumors (FDCT), are rare and likely under-diagnosed malignancies arising in the dendritic reticulum cells of lymph nodes. Extranodal cases are rare, with fewer than 70 cases reported in the head and neck.1 The tonsils are the most common extranodal site in the head and neck,2 other sites include the nasopharynx, parapharyngeal space, maxillary alveolar ridge, hard and soft palate.1 FDCS typically affects young or middle aged patients and there may be a slight female predominance.3,4

We present the case of an 11 year-old patient with FDCS involving the right submandibular region. To our knowledge, this is the third reported case of FDCS in the head and neck in a patient under the age of 16.

CASE REPORT

An 11 year-old boy presented for evaluation following resection of a submandibular mass thought to be a salivary gland cyst on pre-operative imaging. Frozen section was thought to be an inflammatory mass. Final pathology however demonstrated a follicular cell dendritic sarcoma. He was closely followed for a year until he developed fullness of the right submandibular space and underwent exenteration of the right submandibular space with removal of a 3 cm mass and associated facial lymph nodes. The marginal mandibular, hypoglossal, and lingual nerves were preserved. Pathology again was consistent with follicular cell dendritic sarcoma. He was treated post-operatively with proton beam radiotherapy because of positive microscopic margins and his history of recurrence. Over 29 months following salvage treatment, he continues to have no evidence of recurrence.

DISCUSSION

Follicular dendritic cells (FDCs), also known as dendritic reticulum cells, are nonlymphoid elements found in the germinal centers of lymph nodes and other areas of the reticuloendothelial system.5,6 Although their exact mechanism is not known,3 FDCs are a major component in the humoral immune response via antigen presentation and antigen-dependent maturation of the B-cell immune response.1,2

Together with interdigitating dendritic cells and Langerhans cells, FDCs are the nonlymphoid and nonphagocytic accessory cells of the lymphoid system.3

Although the existence of FDCS was hypothesized in 1978 by Lennert,7 the first reported case of malignant transformation from FDCs was not until 1986, when Monda et al described four cases, each initially misdiagnosed.4,5 FDCS are termed sarcomas to emphasize their distinction from lymphomas.2,3,5 An estimated one-third of FDCS are initially misdiagnosed.4

REFERENCES