ABSTRACT

- Objectives: 1) To describe the diagnosis and treatment of granulocytic sarcoma presenting in a submandibular gland; and 2) to identify granulocytic sarcoma as a rare but important diagnosis to be considered in patients presenting with a submandibular gland mass.

- Study Design: The study is a case report and review of the literature.

- Methods: The patient data relevant to the case were reviewed. An extensive literature search was carried out using the PubMed NIH database.

- Results: A young woman, with medical history significant only for radiation exposure near Chernobyl at age 6 presented for evaluation of a right neck mass. She first noticed the rapidly enlarging mass 3-4 weeks prior to presentation. On exam, there was a firm, mobile, 1.5 cm mass in the right submandibular triangle contiguous with the right submandibular gland, with no associated cranial nerve defects. A CT scan confirmed the presence of a right submandibular gland mass; however, fine needle aspiration biopsy was non-diagnostic.

- Conclusions: Early diagnosis of GS is important for both staging and treatment. It may therefore be important to entertain the relatively rare diagnosis of GS within the broader differential diagnosis of salivary gland tumors, especially in patients with previous radiation exposure. Understanding the mechanisms by which GS can present as a salivary gland mass may eventually shed light on general principles determining tumorigenesis within salivary glands.

INTRODUCTION

- Granulocytic sarcoma (GS) is an extramedullary leukemic tumor comprised of cells derived from myeloid precursor cells.

- GS is most often diagnosed in patients with known acute myelogenous leukemia (AML), although it may also present in patients without a known history of AML and, in such 'non-leukemic' cases, is usually a harbinger for the development of AML. GS may also be a feature of blast crisis in chronic myelogenous leukemia or of leukemic transformation in myelodysplastic syndromes (Meis et al., 1996).

- A recent review summarizing published cases of non-leukemic GS noted that, with respect to the head and neck, GS has been reported in lymph nodes, tonsil, and the oral cavity (Yamauchi and Yasuda, 2002). To our knowledge, no previous studies have reported non-leukemic GS in a salivary gland. Here, we describe a case of non-leukemic GS that initially presented as a submandibular gland mass.

RESULTS

- A 25 yo female with a previous medical history significant only for radiation exposure near Chernobyl at age 6 presented for evaluation of a right neck mass. She first noticed the rapidly enlarging mass 3-4 weeks prior to presentation. On exam, there was a firm, mobile, 1.5 cm mass in the right submandibular triangle contiguous with the right submandibular gland, with no associated cranial nerve defects. A CT scan confirmed the presence of a right submandibular gland mass; however, fine needle aspiration biopsy was non-diagnostic.

- A subsequent excision of the right submandibular gland and associated lymph nodes revealed a large gland with focal firmness and significant inflammation along the anterior border of the sternocleidomastoid muscle (SCM) and the submandibular floor, along with some minor submandibular lymphadenopathy. Microscopic examination of the resected specimen demonstrated a diffuse infiltrate of medium to large cells with lobulated, vesicular nuclei, admixed with numerous eosinophils within the salivary gland parenchyma.

- Flow cytometry analysis of the tumor cells showed a population of blasts that expressed CD34 and the myeloid markers CD33, CD13, CD117, and CD33. Fluorescent in situ hybridization on paraffin-embedded tissue revealed translocation of the CBPB gene at chromosome 16q22, consistent with inversion of chromosome 16. The final diagnosis according to the WHO Classification was AML with inv(16) (FAB Classification AML- M4Eo, acute myelomonocytic leukemia with abnormal eosinophils), a type of AML associated with a relatively favorable prognosis. The patient remains disease-free more than 2 years later.

REFERENCES

Jaffe ES et al., ed, WHO Classification of Tumours: Tumours of Haematopoietic and Lymphoid Tissues, IARC Press, Lyon 2001
Meis et al. 1986, Cancer 58: 2697.