Myeloid Sarcoma of the Temporal Bone: Therapeutic Insights
Daniel R. Cox, M.D. and Richard K. Gurgel, M.D.
Division of Otolaryngology – Head and Neck Surgery, University of Utah, UT, USA

INTRODUCTION

Myeloid sarcoma is a rare extra-hematogenous manifestation of acute myeloid leukemia (AML) in which myeloblast cells form soft tissue masses, destroying the normal tissue architecture at the site of involvement. Myeloid sarcomas have been reported in a wide variety of anatomic locations including skin, soft tissues, lymph nodes and the GI tract. The torso is the most frequently involved region, however involvement of the head and neck has also been reported. The history and physical exam findings are non-specific and location dependent. Biopsy is required to make the diagnosis. Histopathological analysis of the biopsy specimen reveals proliferations of immature myelocytes (blast cells), which stain positive for a number of cellular markers including CD43, lysozyme, myeloperoxidase, CD68, CD33 and CD117. Because of the rare nature of this disease, there are no established treatment guidelines, however treatment options usually include radiation, chemotherapy and immunotherapy with donor lymphocyte infusions. As with other hematologic malignancies, surgical resection is indicated for diagnostic purposes only.

We report a case of myeloid sarcoma presenting as a temporal bone mass and discuss management considerations when confronted with possible myeloid sarcoma lesions that involve important structures.

CASE DESCRIPTION

The patient is a 31-year-old male with a history of AML, first diagnosed in May of 2008. He underwent induction chemotherapy at that time consisting of ATRA, Ara-C, and Daunorubicin followed by consolidation chemotherapy. He responded well to treatment and had been in remission since September 2009. In June 2014, he presented with left otalgia of 3 months’ duration and acute left facial paralysis. He had received 2 courses of antibiotic therapy from his primary care provider for presumed otitis media. Physical exam revealed a skin-covered soft tissue mass in the left EAC. Audiogram was consistent with a left conductive hearing loss with an average ABG of 20-30 dB. A temporal bone CT (fig. 1) and MRI (fig. 2) were obtained. The MRI revealed a diffusely enhancing mass of the left EAC, middle ear and mastoid, with enhancement of the entire course of the facial nerve. A biopsy was also obtained which showed histiocytosis, but no definitive diagnosis was able to be made. Differential considerations included benign entities such as adenoma or schwannoma vs a malignant neoplasm such as squamous cell carcinoma.

The decision was made to proceed with tympanomastoidectomy with removal of the lesion. Intraoperatively, a fleshy subcutaneous mass was identified in the EAC, invading the posterior tympanic membrane and insinuating within the mastoid air cells without frank bony destruction. Frozen sections showed non-specific small blue cells. Given the uncertainty of the diagnosis based on preliminary histopathology, we elected to proceed conservatively and care was taken to preserve the ossicular chain until a definitive diagnosis could be made. Final pathology revealed myeloid precursors positive for CD117, CD68, MPO, and lysozyme (fig. 4) and negative for CD34, consistent with myeloid sarcoma. The patient was subsequently referred to medical and radiation oncology for further workup and treatment.

DISCUSSION

Myeloid sarcoma is an uncommon extra-medullary manifestation of AML, with an incidence of 2-5% in adult AML patients. There have been a few cases of myeloid sarcoma of the temporal bone reported in the literature. Patients commonly present with aural fullness, otalgia, and retroauricular swelling. Facial paralysis has also been described. Diagnosis of temporal bone myeloid sarcoma can be challenging because symptoms are non-specific and can mimic those of otitis media or mastoiditis. Additionally, although myeloid sarcoma commonly presents as evidence of disease recurrence in a patient with a previous diagnosis of AML, Pileri et al demonstrated that a significant subset of myeloid sarcoma presents as de novo disease. In that case, a high index of suspicion must be maintained in order to make the diagnosis. Imaging studies such as CT and MRI can be useful and typically show an ovoid, homogeneously enhancing mass with adjacent bony invasion. Biopsy is the preferred diagnostic method, but this may not be feasible depending on the location of the lesion. Because myeloid sarcoma is rare, there have been no large-scale trials comparing treatment modalities and there are no established treatment guidelines, however treatment options usually include radiation, chemotherapy and immunotherapy with donor lymphocyte infusions. As with other hematologic malignancies, there is no role for complete surgical resection if the diagnosis of myeloid sarcoma is being entertained, care should be taken in surgery to preserve normal ear architecture until a definitive diagnosis can be made since these lesions are treated medically, and more extensive resection may compromise hearing unnecessarily.

REFERENCES


Figure 1. Axial CT of the left temporal bone without contrast. White arrow: Soft tissue density in the left EAC with mild thinning of the anterior wall of the EAC. Note the mastoid air cell opacification

Figure 2. Axial T1 post-gadolinium MRI of the temporal bones. White arrow: Ovoid, enhancing mass centered in the left EAC extending to the middle ear space.

Figure 3. H&E staining of surgical specimen. Inset: Appearance of specimen under 40X magnification

Figure 4. Immunohistochemical staining of the surgical specimen. Top left: CD68; Top right: Myeloperoxidase; Bottom left: Lysozyme; Bottom right: CD117