Extramedullary Plasmacytoma of the Lacrimal Sac and Duct Invading the Inferior Meatus: An Extremely Rare Occurrence

Joon Lim, MD†, Sang Woo Yoo, MD†, Keum Ha Choi, MD‡ and Jae Hoon Lee, MD†

†Department of Otolaryngology; ‡Pathology, Institute of Wonkwang Medical Science, Wonkwang University School of Medicine, Iksan, Chonbuk, Korea

ABSTRACT
Extramedullary plasmacytoma (EMP) is a rare malignant plasma cell tumor. EMPs originate outside the bone marrow, particularly in the head and neck region, and can be associated with multiple myelomas. Radiotherapy is considered the treatment of choice, since surgery is limited to biopsy and excision of the residual tumor tissue. The authors report a case of a patient presenting with a history of chronic epiphora and a palpable mass in the medial canthal area, who was found to have an EMP of the lacrimal sac and duct invading the inferior meatus.

KEY WORDS: Extramedullary · Plasmacytoma · Lacrimal Sac · Lacrimal Duct · Inferior Meatus.

INTRODUCTION
Plasmacytomas can present as either medullary or extramedullary neoplasms. Extramedullary plasmacytomas (EMPs) are rare tumors, accounting for approximately 1% of all head and neck malignancies. EMPs originate from plasma cells that express a single class of heavy and light chains in a monoclonal proliferation of B cells. EMPs primarily originate in submucosal areas with an abundance of plasma cells and mainly occur in the upper aerodigestive tract. EMPs have been reported in the submucosal tissues of the nasal cavity, paranasal sinus, and the nasopharynx. EMPs most commonly present as a solitary lesion, and in approximately 10% of cases, multiple sites are involved. EMPs are more common in males, with a male-to-female ratio of 4:1. These tumors usually occur in the fifth or sixth decade of life. To our knowledge, EMP originating from the lacrimal sac and duct has not been previously reported. Here, we report a case of EMP of the lacrimal sac and duct invading the inferior meatus.

CASE
A 55-year-old woman with right epiphora for 1 year was referred to our otolaryngology department. A palpable mass had been noted in the right medial canthal area 1 month ago. The results of ophthalmologic examinations of both eyes for visual acuity and ocular mobility were unremarkable. Nasal endoscopy revealed a red polypoid mass at the right inferior meatus. The left nasal cavity was normal. On palpation, no cervical adenopathy was detected.

Computed tomography showed a mass with soft-tissue density filling the entire length of the right nasolacrimal system and extending to the inferior meatus with mild lacrimal bony erosion (Fig. 1A, B). Magnetic resonance imaging showed a homogenous mass lesion filling the right lacrimal sac and extending to the inferior meatus (Fig. 2A, B).

The biopsy was performed endoscopically at the lesion of the inferior meatus under local anesthesia. The histological finding suggested EMP. Monotonous cells that have eccentrically located nuclei is a rather characteristic finding of plasmacytomas (Fig. 3A). The tumor was positive for the expression of the cytoplasmatic κ-light chain (Fig. 3B) but was negative that of the λ-light chain (Fig. 3C), CD20, CD3, leukocyte common antigen, and pan-cytokeratin.

Positron emission tomography showed no evidence of locoregional lymph node involvement or distant metas-
The patient underwent bone marrow biopsy, urine and serum electrophoresis, and serum biochemistry and all results were normal.

Because there was no evidence of systemic involvement, radiotherapy (total dose, 4500 cGy in 25 fractions) was administered. After radiotherapy, the patient was free of disease. During follow-up period, she remained healthy without recurrence.

**DISCUSSION**

Most malignant tumors of the lacrimal drainage system are primary tumors originating from the epithelium or the mesenchymal components of the lacrimal sac. Primary neoplasms originating from the lacrimal sac are epithelial (75%) or non-epithelial (25%) tumors, such as mesenchymal tumors (12%), melanomas (5%), and malignant lymphomas (6%). Tumors confined to the lacrimal sac are extremely rare, and most of them affect adjacent structures, such as the eyelid, nose, sinuses, and the orbit.

Lacrimal sac tumors in patients with epiphora usually present as palpable masses over the lacrimal sac region, particularly extending above the medial canthal ligament. In cases of masses that exhibit rapid growth or bone destruction over the lacrimal sac region, a biopsy should be performed instead of an unwarranted surgery.
In a histological examination, the appearance of an EMP may be similar to that of other small-cell tumors, such as melanomas, lymphomas, undifferentiated carcinomas, and other benign inflammatory plasma cell tumors. EMP is composed of plasma cells that express cytoplasmic κ- or λ-light chains; therefore, immunohistochemical analyses can be used to differentiate EMP from other entities. The detection of negative immunostaining for CD20 and positive immunostaining for CD79 also support this diagnosis.

EMP can be treated by surgical resection, radiation, or a combination of both. Since localized EMP is highly radiosensitive, the recommended primary treatment is radical radiotherapy. Surgery is the recommended mode of treatment for tumors that are well localized and can be excised completely. With larger tumors, a combination of radiotherapy and surgery is required for a greater chance of survival.

A long-term follow-up study of EMP showed that local recurrence rates were 21% for radiotherapy alone; 20% for surgery alone; and 46% for combined treatment. The higher recurrence rate following combined therapy may be explained by the fact that this mode of therapy was used in cases where the disease was more extensive. Chemotherapy is indicated only when there are multiple lesions occurring in sites other than the primary site.

The prognosis is more favorable when a lesion occurs in a solitary mass rather than in multiple areas. The overall prognosis is good when multiple myeloma has been ruled out. The 5-year survival rate is approximately 50%.

Orbital involvement of EMP is extremely rare, and thus far, only 6 cases have been reported. In these cases, the EMP had originated from the lacrimal gland. All cases described a painless enlarging mass with no bony involvement. These tumors often present with non-specific symptoms that slowly progress over a number of months. Pain is uncommon and most symptoms are secondary to the mass effect. To our knowledge, this is the first case report of EMP of the lacrimal sac and duct invading the inferior meatus. Rhinologists should consider that the mass in the inferior meatus can be originated from the lacrimal apparatus.

Acknowledgments

REFERENCES