Metastatic Sarcoma of the Tongue: Pleomorphic Malignant Fibrous Histiocytoma and Literature Review

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Abstract: Metastatic tumors of the tongue are rare, most commonly arising from carcinomas of the lung, kidney, breast, and melanomas. They usually appear in end-stage disease, especially in the tongue base, probably because of the rich blood supply. Metastatic sarcomas of the tongue are even rarer. We report the seventh case of metastatic tongue sarcoma and the first one in the literature, arising from a pleomorphic malignant fibrous histiocytoma (MFH) of the lower extremity. ENT surgeons, Maxillofacial surgeons, and Oncologists should be aware of the possibility of metastatic sarcomas of the tongue, even though these lesions are extremely rare. Furthermore, metastatic sarcomas of the tongue represent a late manifestation of a previously identified primary tumor, although if the primary site has not been identified, a thorough investigation to locate an occult tumor should be conducted.

Keywords: Sarcoma, metastatic, tongue, pleomorphic malignant fibrous histiocytoma, lingual, cancer.

INTRODUCTION

Sarcomas are rare malignant tumors from mesenchymal cells, accounting for less than 1% of all malignancies [1]. Five to 15% of all sarcomas in adults occur primarily in the head and neck region, while in pediatric patients the respective percentage reaches 35% [2].

Metastatic tumors of the tongue are rare, with a prevalence of 0.1% to 0.2% in large series, usually arising from carcinomas of the lung, kidney, breast, and melanomas. They usually appear in end-stage disease, especially at the base of the tongue, probably because of the rich blood supply [3].

Metastatic sarcomas of the tongue are even rarer. Only six cases of metastatic sarcoma of the tongue have been identified in a literature review; three of them were leiomyosarcomas, two epithelioid sarcomas, and one alveolar soft part sarcoma.

The present paper reports the seventh case of metastatic sarcoma of the tongue, and the first one arising from a pleomorphic malignant fibrous histiocytoma (MFH) of the lower extremity.

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CASE REPORT

A 43-year-old man presented with a 15 day history of tongue swelling and difficulty in swallowing. The patient did not have any respiratory distress, and was otherwise fit. Clinical examination revealed a hard mobile pedunculated chestnut-colored mass on the right side of the tongue base, and a smaller one on the left side of the soft palate. The mobility of the tongue was impaired, but no palpable cervical lymph nodes were found. A neck MRI confirmed the clinically observed lesions, but was not suggestive of nodal involvement.

The patient had been treated with surgical intervention and irradiation for a pleomorphic MFH of the left lower extremity three years earlier. One year post-intervention he had sustained pulmonary metastases, which were treated with bilateral pneumonectomies and chemotherapy.

The treatment plan regarding both the base of tongue and the palatal tumor involved surgical excision, with postoperative irradiation of the tumor sites, as well as radiotherapy in levels II – IV bilaterally. The histological specimens included a 5 cm in diameter round chestnut-colored mass from the base of the tongue, and a 2 cm in diameter lesion from the soft palate. Both lesions were malignant spindle cell neoplasms, with overlying ulceration, and infiltration of

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the underlying muscles (Figures **1**, **2**, and **3**). The differential diagnosis included metastatic sarcoma and sarcomatoid carcinoma. The tumors were keratin positive. However, the patient's past medical history, the histological/immunohistochemical report from the resected tumor of the lower extremity, and other immunohistochemical markers from the tongue/palatal tumors contributed to the diagnosis of a metastatic sarcoma of tongue (cells positive for vimentin (Figure **4**), SMA, MSA, AE1/AE3 and negative for EMA, HMB-45, CD-34 and CK7).

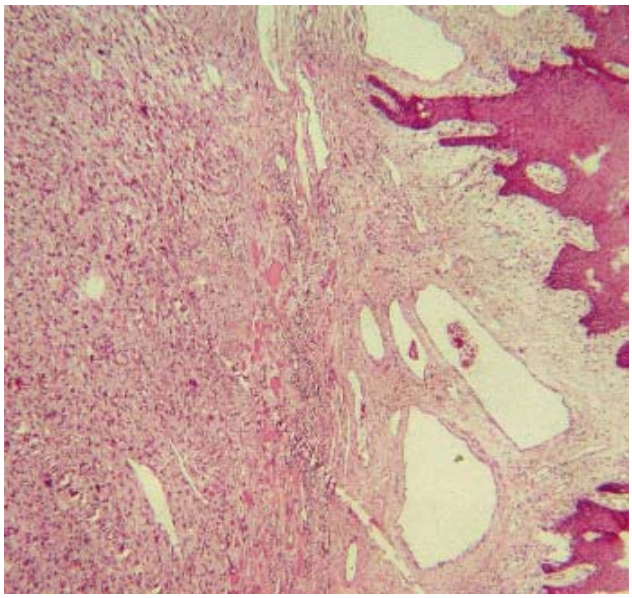

Figure 1: Histologic appearance of metastatic sarcoma of the tongue arising from pleomorphic MFH of the lower extremity (Oral cavity mucosa and tumor cells).

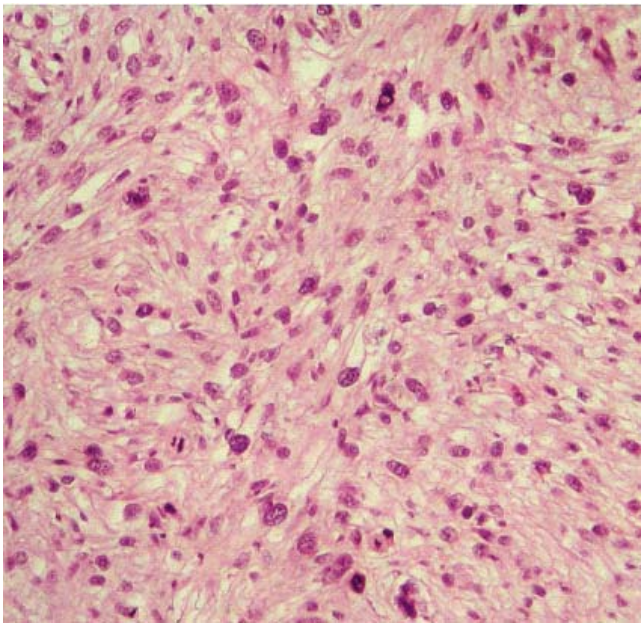

Figure 2: Metastatic sarcoma of the tongue, tumor cells.

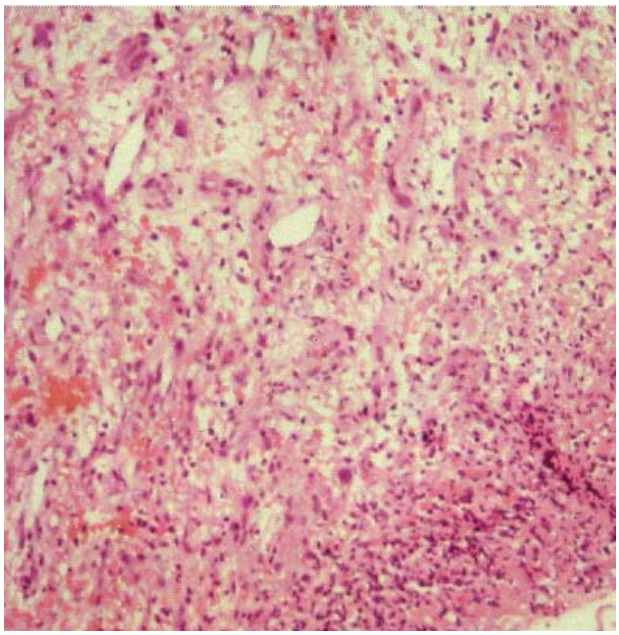

Figure 3: Histopathologic specimen showing ulceration in metastatic pleomorphic MFH of the tongue.

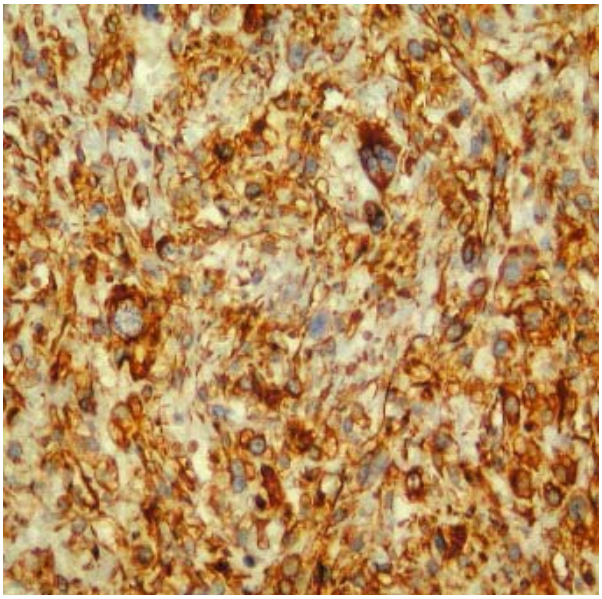

Figure 4: Tumor cells positive for vimentin.

The final diagnosis was metastatic pleomorphic MFH, arising from the lower extremity. Due to the low possibility of occult nodal involvement in the neck, postoperative neck radiotherapy was not performed. One month later the patient sustained local recurrence in the left lower extremity and cerebral metastases. One week later he passed away.

DISCUSSION

Sarcomas are relatively rare malignant tumors, arising from the connective tissue. They include tumors

from bony elements (osteosarcoma and chondrosarcoma), and from soft tissue elements (soft tissue sarcomas).

Approximately, 80% of all sarcomas in the head and neck are soft tissue sarcomas, and only 20% include osteosarcomas or chondrosarcomas [2]. Soft tissue sarcomas are named according to their tissue of origin; rhabdomyosarcoma, fibromyosarcoma, synovial sarcoma, liposarcoma, malignant fibrous histiocytoma (MFH), alveolar soft part sarcoma, hemangiopericytoma, neurogenic sarcoma, etc.

The classification of soft tissue sarcomas is still evolving [4]. Sturgis *et al*. [2] classified sarcomas according to the tissue of origin, the histological grade, or the respective anatomical site in the head and neck region (Tables **1** and **2**).

Table 1: The Sturgis-Potter Classification of Sarcomas According to the Histological Grade [2]

Histological grade
1. Low grade sarcomas (dermatofibrosarcoma protuberance, desmoid tumors and atypical lipomatous tumors)
2. High grade sarcomas (alveolar soft part sarcomas, MFH, synovial sarcomas, Ewing sarcomas, rhabdomyosarcoma, and angiosarcoma).
3. Sarcomas requiring individualized grading (fibrosarcoma, leiomyosarcoma, liposarcoma, neurogenic sarcoma and hemangiopericytoma).

Table 2: The Sturgis-Potter Classification of Soft Tissue Sarcomas According to the Anatomical Location in the Head and Neck [2]

Head and neck area
1. Scalp and face sarcomas
2. Sinonasal tract, anterior skull base sarcomas
3. Ear, lateral skull base sarcomas
4. Upper aerodigestive tract sarcomas
5. Parotid gland and neck sarcomas

With regard to the histological classification, soft tissue sarcomas, when poorly differentiated, may be categorized as pleomorphic sarcomas, spindle cell sarcomas, or malignant fibrous histiocytomas (MFH) [4]. However, the differential diagnosis in soft tissue sarcomas may be difficult, and the terminology is not always clear, or accepted by all centers. Therefore, misdiagnosis is always possible.

Recent advances in immunohistochemistry and molecular biology have confirmed these difficulties, and the associated problems in diagnosis. In a review of 281 sarcomas Duagard *et al.* reclassified their cases using immunohistochemistry, and changed their original diagnosis in 57% of the cases [5]. Moreover, 7% of these cases were not found to be sarcomas. However, we should take into account that up to 10% of soft tissue sarcomas remain unclassifiable [5].

Condre 2003 reported a new classification of sarcomas, based on the role of immunohistochemistry in the diagnosis of the disease [6]. In the respective classification, the sarcomas are classified as:

- Sarcomas for which immunohistochemistry is definitive for diagnosis. These include rhabdomyosarcomas, epithelioid sarcoma, clear cell sarcoma, desmoplastic round cell tumor, and gastrointestinal stromal tumors.

- Sarcomas for which immunohistochemistry may be useful, but the immunohistochemical profile is neither specific, nor constant, and therefore should be interpreted with caution. These include Ewing sarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, dermatofibrosarcoma protuberans and giant cell fibroblastoma, extraskeletal myxoid chondrosarcoma, liposarcomas, and alveolar soft part sarcomas.

- Sarcomas with non-specific immunohistochemical profile, not displaying specific markers, in which, however, immunohistochemistry may be useful to exclude non-mesenchymal malignant tumors (or sarcomas with a specific line of differentiation). These include fibroblastic and poorly differentiated sarcomas, such as fibrosarcoma, myxofibrosarcoma, and pleomorphic malignant fibrous histiocytoma.

Soft tissue sarcomas are tumors with extensive local invasion, resulting in a high rate of local recurrence. This depends on the histological type of the tumor. Liposarcoma, well-differentiated sarcoma, and MFH have an increased tendency in local recurrence, while fibrosarcoma, synovial sarcoma, and alveolar soft part sarcoma have an increased tendency in long delayed metastases. The occurrence of nodal metastases in soft tissue sarcomas is usually less than 5%, while the lung is the initial site of distal metastasis, in approximately 90% of patients [7].

The histological grade and the prognosis of the disease are closely related, even though this

relationship is not always clear, primarily because many of these tumors are too rare to establish prognostic factors, especially in single centers. Deyrup *et al.* suggested that the presence of even a single myoid marker (desmin, muscle-specific actin, α-smooth muscle actin, and myoglobin) may have a significant negative impact on survival in these patients [4].

MFH is the most common soft tissue sarcoma, even though more than half of these cases could be reclassified in other categories, when immunohisto-chemistry is used [4]. Only 3%-10% of MFHs occur in the head and neck.

MFH is composed of spindle cells, arranged in a storiform pattern with frequent neoplastic histiocyte cells which may be multinucleated. It is rarely multifocal, and it seldom presents with local or distal metastases [2].

Metastatic pleomorphic MFH of the tongue is extremely rare. In a review of the English literature only six cases of metastatic sarcoma of the tongue were found; three of them arose from uterine leiomyo-sarcoma [8, 9, 1], two from epithelioid sarcomas of the extremities [10, 11] and one was an alveolar soft part sarcoma from the right brachialis muscle [12]. The present case is the seventh metastatic sarcoma of the tongue ever reported, and the first one arising from a pleomorphic MFH of the lower extremity.

CONCLUSION

ENT surgeons, Maxillofacial surgeons, and Oncologists should be aware of the possibility of metastatic sarcomas of the tongue, even though these lesions are extremely rare. Furthermore, metastatic sarcomas of the tongue represent a late manifestation of a previously identified primary tumor, although if the primary site has not been identified, a thorough investigation to locate an occult tumor should be conducted. The reported case represents the first metastatic sarcoma of the tongue arising from a pleomorphic MFH of the lower extremity.

CONFLICTS OF INTEREST

None declared. The authors have no financial interests, and have not received any financial support for this article.

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