Myofibroblastic Sarcoma of the Base of Tongue
Case Report and Review of the Literature

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Background: Mesenchymal malignancies with myofibroblastic differentiation exhibit a spectrum from low-grade myofibroblastic sarcoma mimicking fibromatosis to pleomorphic high-grade sarcoma. Low-grade myofibroblastic sarcoma shows a wide anatomic distribution with a predilection for the head-and-neck region; however, intermediate- and high-grade myofibroblastic sarcomas in this localization are exceptional.

Case Report: A 56-year-old woman with intermediate-grade myofibroblastic sarcoma of the base of tongue is presented. She was treated with surgical excision, but computed tomography proved local residual tumor. Reexcision and chemotherapy were refused by the patient. Irradiation was given to a total dose of 66 Gy.

Result: 50 months after completion of radiotherapy, the patient is in good health without any evidence of disease. According to the review of the literature, base of tongue as the primary site of myofibroblastic sarcoma has not been published so far.

Conclusion: Similarly to the low-grade form, intermediate- and high-grade myofibroblastic sarcomas may also occur in the head-and-neck region. In case of incomplete excision, radiotherapy may be an effective treatment.

Key Words: Myofibroblastic sarcoma · Base of tongue · Radiotherapy

Myofibroblastisches Sarkom des Zungengrundes. Fallbericht und Literaturübersicht


Schlüsselwörter: Myofibroblastisches Sarkom · Zungengrund · Strahlentherapie

Introduction
More than 80% of malignant tumors of the head-and-neck region originate from squamous epithelium. The incidence of mesenchymal tumors is low, and among these, myofibroblastic sarcomas are extremely rare [14, 16, 22].

Myofibroblasts are mesenchymal spindle cells that share ultrastructural features of both fibroblasts and smooth muscle cells [22]. The presence of myofilaments in the cytoplasm of myofibroblasts bestows them with contractile properties. Gabiani et al. were the first to describe these cells in 1971 [7].

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Histologically, myofibroblasts are spindle-shaped cells with ill-defined eosinophilic cytoplasm. The nucleus is fusiform containing a small central eosinophilic nucleolus [16]. Myofibroblastic cells or myofibroblastic differentiation are present in wound healing and in different reactive and neoplastic conditions (reactive stromal component in numerous neoplasms). Pseudosarcomatous proliferations, fasciitis, hypertrophic scar, superficial and deep fibromatoses generally also show myofibroblastic differentiation [16].

Tumors with myofibroblastic differentiation present with variable morphological and immunohistochemical characteristics due to the plasticity of the myofibroblasts [14]. Myofibroblastic sarcomas occur in almost every organ, more commonly in the superficial soft tissues, particularly in the head-and-neck region; however, until now base of tongue as the primary site of myofibroblastic sarcoma has not been published in the literature.

Here, we describe the case of a 56-year-old woman with an intermediate-grade myofibroblastic sarcoma of the base of tongue, who was treated with tumor excision and – because of the presence of residual tumor – postoperative radiotherapy.

**Case Report**

In December 2003, a 56-year-old woman presented with progressive swallowing difficulties and suffocation at the Department of Otorhinolaryngology, Szent Imre Hospital, Budapest, Hungary. The clinical history was unremarkable. She was smoking two packets of cigarettes per day.

During the examination, a round, pedunculated, livid mass measuring 4 cm in diameter and covered with fibrinoid fur was detected on the right side of the base of tongue protruding to the pharyngeal space, which fell on the larynx. Nevertheless, the structure of the larynx was normal and the glottis was duly wide. There was no clinical involvement of cervical lymph nodes.

The exophytic tumor was resected without delay. A computed tomography (CT) scan was requested after the surgical procedure. This showed the presence of a residual tumor mass at the base of the tongue (Figure 1).

Microscopic examination of the resected specimen showed an ulcerated tumor composed of spindle-shaped, elongated and stellate cells embedded in myxoid matrix (Figure 2a). Focally, the spindle-shaped cells formed short fascicles. Centrally, small hyalinized areas were also present. The neoplastic cells showed mild to moderate nuclear pleomorphism and high mitotic activity (1–5/1 high-power field [HPF]) with numerous atypical mitoses (Figure 2b). The amount of cytoplasm was variable showing pale eosinophilic to pale basophilic staining characteristics. Scattered multinucleated neoplastic giant cells also appeared. The spindle-shaped neoplastic cells were vimentin-positive and showed negative reactions with anti-h-caldesmon, anti-desmin, anti-S-100 and anti-AE/AE3 cytokeratin antibodies. Approximately 15% of the cells reacted with smooth muscle α-actin. The histopathologic diagnosis was intermediate-grade myofibroblastic sarcoma.

The patient refused an extended reoperation, or chemotheraphy. So irradiation was carried out with a dose of 66 Gy (2 Gy/day; five fractions/week) to the primary site and the upper neck using opposed lateral 6-MV photon beams. Dose was prescribed to the midline. The lower neck was also irradiated up to 50 Gy using an anteroposterior field. Dose prescription was at 3 cm depth. The spinal cord was shielded after 40 Gy. Electron beams (9 MeV) were used to supplement the posterior cervical lymph nodes up to 50 Gy. 2 months after irradiation, the control CT showed complete tumor regression. Since then, she has been followed with CT or magnetic resonance imaging (MRI) performed once a year (Figure 3). After a follow-up period of 50 months the patient is alive without any evidence of disease.

**Discussion**

Myofibroblasts are altered fibroblasts, which occur in the stroma of normal organs and in reactive processes, such as granulation tissue. A variety of benign and intermediate soft-tissue tumors show myofibroblastic differentiation. Malignant mesenchymal neoplasms showing evident and dominant myofibroblastic differentiation are described as myofibroblastic sarcoma, myofibrosarcoma, low-grade myofibroblastic sarcoma, low-grade spindle cell sarcoma consisting of myofibroblasts,