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Case report

Low-grade myofibroblastic sarcoma. Two rare tumors in two rare locations

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ABSTRACT

Low-grade myofibroblastic sarcoma (LGMS) represents an atypical tumor composed of myofibroblasts with a predilection for the head and neck, especially in the tongue and oral cavity, with a high tendency to local recurrences and metastases, even after a long period. LGMS arising in the maxillary sinus and in the neck are extremely uncommon. To the best of our knowledge, only 50 cases of low-grade myofibroblastic sarcoma have been reported. We report two cases of LGMS of the maxillary sinus and neck, discussing clinical, histological, inmunohistochemical and therapeutic features.

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Sarcoma miofibroblástico de bajo grado. Dos tumores infrecuentes en dos localizaciones poco habituales

Palabras clave:
Miofibrosarcoma de bajo grado
Inmunohistoquímica
Actina
Recidivas.

RESUMEN

El sarcoma miofibroblástico de bajo grado (SMFBG) representa un tumor atípico, formado por miofibroblastos, que tiene predilección por cabeza y cuello, en especial la lengua y la cavidad oral, y se caracteriza por una elevada tendencia a las recidivas locales y a las metástasis, incluso después de transcurrido un período prolongado. Los SMFBG que se originan en el seno maxilar y en el cuello son excepcionales. Hasta lo que conocen los autores, solo se han publicado 50 casos de sarcoma miofibroblástico de bajo grado. Describimos 2 casos en los que se identificaron estos tumores, uno en el seno maxilar y el otro en el cuello, y abordamos sus características clínicas, histológicas, inmunohistoquímicas y terapéuticas.

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Introduction

Low-grade myofibroblastic sarcoma (LGMS) represents an atypical tumor composed of myofibroblasts with a predilection for the head and neck, especially in the tongue and oral cavity.[1,2] Myofibroblasts are mesenchymal spindle-shaped cells present in almost every soft tissue. LGMS usually occurs in adult patients with a slight male predominance. Children are rarely affected.[2] Although LGMS has been studied extensively, determining and defining its exact role in the spectrum of proliferative and neoplastic spindle cell lesions remains problematic.[2] Clinically, patients complain of a painless swelling or an enlarging mass.[2] It is a slow growing neoplasm with a tendency to local recurrences and metastases, even after a long period.[3] The differential diagnosis includes both benign and malignant lesions such as nodular fasciitis, fibromatoses, fibrosarcoma or leiomyosarcoma.[2,4] To the best of our knowledge, only 50 cases of low-grade myofibroblastic sarcoma have been reported. Now, we report 2 patients with LGMS of the maxillary sinus and neck. Clinical, histological, inmunohistochemical and therapeutic features are described.

Case report

Case 1 (Fig. 1)

A 75-year-old female Caucasian was referred to our Oral and Maxillofacial Department from the Otolaryngology Department, for evaluation of a painful mass in the left maxillary sinus. The mass had appeared one month previously. Her personal and familial medical histories were unremarkable and she had never smoked or abused alcohol.

Oral examination revealed a minimum but very painful swelling of the left vestibular sulcus of the maxilla. The overlying mucosa appeared normal. No clinical evidence of lymphadenopathy was observed.

CT-scan revealed a well-defined mass in the left maxillary sinus with bone destruction of the three walls and partial infiltration of the orbital floor.

An incisional biopsy of the lesion was performed. Microscopic examination revealed an expansive mesenchymal tumor characterized by a diffuse proliferation of spindle-cells with rounded nuclei, small nucleolus surrounded by an intense lymphocytic infiltrate. On the basis of these clinical, radiological and histological findings, the patient was diagnosed with fibromyxoma versus low grade sarcoma.

The patient underwent, under general anesthesia, a total maxillectomy including pterygoid region, orbital floor and zygomatic body, preserving the ocular globe, by means of a Weber-Fergusson incision. The defect was then reconstructed with two titanium meshes covered with a myofascial temporal flap.

Definitive histological examination showed a firm mass of white myxoid tissue with a central cystic area, characterized by a mesenchymal proliferation of spindle-cells, figures of mitosis, areas with myxoid and fibrosclerotic changes as well as areas of necrosis. There was extensive infiltration of the fat tissue and bone, extending into the zygomatic and palatine tissue.

Immunohistochemistry revealed that most spindle-cells were stained diffusely for vimentin, smooth muscle actin, CD10 and cytokeratin and focally for caldesmon. Staining for other markers, such as desmin, CD34, ALK, EMA and S-100 protein was not observed. Proliferative index (Ki-67) was moderate.

Given the age of the patient and the presence of tumor in the surgical margins, postoperative radiotherapy was performed. After 1 year of follow-up, the patient had a distant metastasis into the left humerus bone that was resected by the Traumatology Department. On the other hand, the patient has no facial or cervical pain and swelling.

Case 2 (Fig. 2)

A 74-year-old man was referred to our Department for evaluation of an oropharyngeal squamous cell carcinoma relapse after three previous surgical excisions in another Department. Physical examination showed a total absence of the tongue and the presence of an intraoral pectoral flap with no evidence of oropharyngeal lesions. CT-scan showed a high density heterogeneous mass in the tongue base measuring 4.1 cm × 2.5 cm that obliterated the left vallecula. The patient was diagnosed with tumor relapse and underwent a total laryngectomy.

Histological examination demonstrated a diffuse mesenchymal proliferation composed of spindle cells arranged in fascicles of varying length that infiltrates between individual muscle fibers. The nuclei were mostly irregular. Most tumor cells showed moderate atypia and there were many mitotic figures (more than 20 in 10 high power fields) and focal areas of necrosis. Proliferative index was high (Ki-67 80%). Immunohistochemical examination showed diffuse staining for smooth muscle actin with focal staining for caldesmon. No other stains were taken up, including desmin, cytokeratin, EMA and S-100 protein.

6 months after surgery, the patient presented another cervical relapse treated by means of another surgical intervention and died in the early postoperative period because of a carotid artery breakage.

Discussion

Low-grade myofibroblastic sarcoma (LGMS) has a predilection for the head and neck, especially in the tongue and oral cavity.[1,2] Rare cases involving the salivary glands, the paranasal sinuses and even the mandible have also been reported.[4–6]

Despite isolated case reports, myofibrosarcoma was defined as a distinct entity only recently.[7,8] It usually occurs in adult patients with a slight prevalence in males. Children are rarely affected.[2]

The tumor is composed of myofibroblasts. First described by Gabbiani et al.[9] in 1971, they are mesenchymal spindle-shaped cells that share ultrastructural features with both fibroblasts and smooth muscle cells.[2,10] They are present in connective tissue and contribute to reparative and reactive

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