A low grade fibromyxoid sarcoma originating from the masseter muscle: a case report

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Abstract

Introduction: Low grade fibromyxoid sarcoma is a distinctive variant of fibrosarcoma. We recently experienced a rare case of low grade fibromyxoid sarcoma arising in the masseter muscle.

Case presentation: A 63-year-old Korean woman with a large growing mass in her right parotid gland area for 1 year visited our clinic. Complete removal of the tumor was achieved by parotidectomy with facial nerve preservation. The tumor measured over 4cm with pathologic findings compatible with low grade fibromyxoid sarcoma.

Conclusions: Low grade fibromyxoid sarcoma is an extremely rare tumor, and report of the present case is noteworthy since it represents a rare localization of low grade fibromyxoid sarcoma in the head and neck. Close follow up on a long-term basis is considered necessary because of its high potential to metastasize.

Keywords: Fibromyxoid sarcoma, Head and neck, Masseter

Introduction

Low grade fibromyxoid sarcoma is a cytologically bland malignant neoplasm with alternating fibrous and myxoid stroma of low grade and malignant potential [1]. Low grade fibromyxoid sarcoma is considered a rare soft tissue tumor with a high metastasizing potential, despite its benign histologic appearance. Low grade fibromyxoid sarcomas represent approximately 10% of soft tissue sarcomas and are rarely found in the head and neck region. The most common locations are the extremities, trunk, chest and soft tissues.

There have been only five reported cases of low grade fibromyxoid sarcoma in the head and neck [2]. Low grade fibromyxoid sarcoma originating from the masseter muscle has never been reported before. We recently experienced a rare case of low grade fibromyxoid sarcoma in the masseter muscle. Due to the relative rarity of low grade fibromyxoid sarcomas, there is no consensus regarding treatment and postoperative follow-up recommendations. Nevertheless, the low grade fibromyxoid sarcoma has potential for late local recurrence and distant metastasis [3]. In order to detect possible metastasis in advance it is important to inform the patients about the longstanding metastatic potential of the disease and recommend prolonged follow-up.

Case presentation

A 63-year-old Korean woman who had no medical history presented with swelling at her right parotid gland area that had persisted for 1 year. No abnormal findings were found elsewhere, and the other laboratory tests showed normal. A physical examination revealed a fixed non-tender round-shaped mass at her right parotid gland area. Images of neck computed tomography (CT) revealed a 3.2cm-sized heterogeneously enhancing mass which was located lateral to the mandibular rami and next to her right masseter muscle (Fig. 1a, b). Neck magnetic resonance imaging (MRI) showed a well-demarcated, 2.5×3.0cm-sized, gadolinium-enhanced, and fungating mass with internal hemorrhage and increased vascularity (Fig. 1c, d). Ultrasonography showed a heterogeneous hypoechoic mass in her right parotid gland suspicious for parotid gland tumor, and a fine-needle aspiration biopsy showed a few fragments of relatively short spindled cells in myxoid and bloody background.

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Cytologists also suggested that we rule out pleomorphic adenoma.

Superficial parotidectomy was done with modified facelift incision. The main mass adjacent to her masseter muscle was located at the anterior aspect of her parotid gland (Fig. 2a). Pes anserinus and all branches of her facial nerve were identified and preserved (Fig. 2b, c). The retrieved specimen revealed a 4 cm-sized mass (Fig. 2d).

On macroscopic examination, the tumor was well-demarcated and had a yellow-white appearance with focal glistening areas on a cut section (Fig. 3a). On microscopic examination, the tumor had alternating fibrous (cellular) and myxoid (hypocellular) areas (Fig. 3b). The tumor cells were spindle and bland-looking with rare mitosis (Fig. 3c, d). The histopathologic findings of this tumor were compatible with low grade fibromyxoid sarcoma. The diagnosis of low grade fibromyxoid sarcoma was confirmed by the characteristic histological appearance with immunohistochemical staining [6, 7]. On histological examination, low grade fibromyxoid sarcoma shows alternating fibrous and myxoid areas. Also, the surrounding cells are more crowded, round or ovoid, and blend imperceptibly with the surrounding spindle-shaped cells. On immunohistochemical examination, it usually shows intense immunostaining for vimentin, P53, bcl-2 and smooth muscle actin (SMA) and negative for cytokeratin (CK)-19, galectin-3, and calcitonin; this finding may help distinguish low grade fibromyxoid sarcoma from other sarcomas [7, 8]. In the present case,
Fig. 2 Intraoperative findings. Communicating branch (white arrow) between marginal branch and buccal branch was noted and sacrificed to remove complete mass excision (a). The main mass was located adjacent to masseter muscle in the deep lobe of parotid gland (b). All facial nerve branches were preserved: temporal (T), zygomatic (Z), buccal (B), marginal mandibular (M), and cervical branches (C) (c). Gross finding of main mass showed well-demarcated margin (d). margin (Supf. = superficial lobe of parotid gland, Main (Deep) = deep lobe of parotid gland)

Fig. 3 Gross finding and histopathological view. The main mass showed well-margined shape (a). Cut section showed well-demarcated, yellow-whitish mass with focal glistening area (b). Low power view showed tumor with alternating fibrous and myxoid areas on hematoxylin and eosin ×40 (c). High power view showed spindle tumor cells with minimal atypia on hematoxylin and eosin ×400 (d)
Gene rearrangement or FUS-CREB3L1 contributions that results in the in situ FUS gene arrangement will also be used by hybridization; in situ et al. Journal of Medical Case Reports. reported the 5-year overall survival rate following surgery for low grade fibromyxoid sarcoma is over 90%, with a more favorable prognosis associated with smaller tumors [12]. Local recurrences of low grade fibromyxoid sarcoma have been reported from a few months to 15 years after initial treatment, while distant spread has occurred from 0 to 45 years after surgery. Radiotherapy or chemotherapy may be necessary for patients whose tumors recur locally or spread to distant sites.

Low grade fibromyxoid sarcoma is a slow-growing tumor, but this cancer has a tendency to recur. It has been known to spread to distant organs many years after surgery; therefore, close serial follow up on a long-term basis would be required to prevent local or distant metastasis due to the tendency to recur after initial treatment.

Conclusions
This is the first report in the medical literature of a low grade fibromyxoid sarcoma arising from the masseter muscle. Although low grade fibromyxoid sarcoma has the possibility of distant metastasis, it is likely that this patient may have a favorable prognosis with long-term close follow up because the tumor was solitary, well capsulated, confined to the localized area, and did not show severe mitosis in histology.

Consent
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations
FISH: Fluorescent in situ hybridization; FUS: Fusion in sarcoma; MRI: Magnetic resonance imaging; MUC4: Mucin 4.

Competing interests
The authors declare that they have no competing interests.

Authors' contributions
HJH managed the patient and acquired data. EJL participated in the operation, acquired and interpreted the data, and wrote the manuscript. HKB participated in the operation and reviewed the manuscript. HSP reviewed the pathology slide and commented about pathologic diagnosis. H-SC was a major contributor in writing the manuscript and interpreted the patient data. All authors read and approved the final manuscript.

Acknowledgements
No funding was obtained for this report. We would like to thank the Department of Pathology for their help with the photographs and to other collaborators in the Department of Otorhinolaryngology for their help with caring for the patient.
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