대한두경부종양학회지
제 29 권 제 2 호 2013

Primary Leiomyosarcoma of the Submandibular Gland*

Dong Hoon Lee, MD[1], Young Kim, MD[2], John Jae Woon Lee, MD[1], Joon Kyoo Lee, MD, PhD[1]

Departments of Otolaryngology-Head and Neck Surgery[1] and Pathology,[2] Chonnam National University Medical School and Chonnam National University Hwasun Hospital, Hwasun, Korea

악하선에 발생한 평활근육종*

전남대학교 의과대학 화순전남대학교병원 이비인후과학교실,[1] 병리과학교실[2]

이동훈[1] · 김 영[2] · 이재운[1] · 이준규[1]

= 국문초록 =

평활근육종은 주로 사지와 몸통에 주로 발생하며, 악하선의 원발성 평활근육종은 매우 드물다. 악하선의 원발성 평활근육종은 증상과 방사선 검사 모두에서 비특이적이며, 치료는 광범위 절제술이다. 최근 저자들은 악하선에 발생한 원발성 평활근육종을 광범위 적출술을 시행한 후에 술후 방사선 치료를 시행하여 치험하였기에 문헌 고찰과 함께 보고하는 바이다. 악하선의 평활근육종은 드물지만, 방사선 검사에서 경계가 분명하고 조영 증강이 잘되며, 세침흡입검사에서 방추세포가 발견될 경우에 감별 진단으로 고려해야 한다.

중심 단어 : 평활근육종 · 악하선 · 적출 · 방사선.

Introduction

Leiomyosarcoma(LMS) is a malignant smooth-muscle tumor.[1-5] The tumor usually occurs in the uterus, gastrointestinal tract and retroperitoneum. Occurrence in the head and neck is extremely rare.[5] The oral cavity and jaw bones are the most prevalent location of LMS in the head and neck region, while the occurrence of LMS in the major salivary gland is exceptional, especially in the submandibular gland.[1,5] Herein, we present a case of LMS of the submandibular gland treated by radical excision of the tumor and postoperative radiotherapy.

Case Report

A 58-year-old male presented with right submandibular swelling. A lesion had appeared 3 months previously. The lesion was firm, mobile, painless and covered by normal skin. Clinical history revealed the diagnosis of diabetes mellitus 2 years prior to the present admission. Nineteen years previously, the patient underwent right submandibular mass removal at local hospital. However, the patient did not recognize the biopsy result of the right submandibular mass.

Computed tomography(CT) scan with contrast enhancement revealed an approximately 3×3 cm-sized heterogeneous lesion of the right submandibular area without regional pathologic lymph nodes(Fig. 1). Ultrasound-guided fine needle aspiration cytology was performed, which revealed fascicles of spindle cells. Fine needle aspiration cytology was repeated, but the result showed a virtually acellular smear.

The patient underwent radical excision of the right submandibular gland extending to the surrounding tissues via a submandibular approach while under general anesthesia. We did not perform frozen-section during surgery. In addition, we did not perform radical neck dissection, because we could not recognize the possibility of malignancy until surgery.

The surgical specimen measured 4 cm in diameter and ap-

Received : June 10, 2013 / **Revised** : June 30, 2013
Accepted : July 5, 2013
교신저자 : 이준규, 519-809 전남 화순시 일심리 160
전남대학교 의과대학 화순전남대학교병원 이비인후과학교실
전화 : (061) 379-8190 · 전송 : (061) 379-7761
E-mail : joonkyoo@chonnam.ac.kr

peared as a fleshy white-grey mass with ill-defined margins and foci of necrosis(Fig. 2). Mitosis averaged 5 per 10 high-power fields, and the stroma was characterized by necrotic foci. Histopathologic evaluation showed spindle cells arranged in fascicular growth pattern(Fig. 3). Immunohistochemistry demonstrated positive staining of the tumor cells for desmin and smooth muscle actin and negative staining for S-100 protein antigen(Fig. 4). The final diagnosis of low-grade leiomyosarcoma of the submandibular gland was established. After surgery, the distant metastasis workup(including positron-emission tomography CT) was performed. The result was negative. The postoperative course was uneventful, and the patient underwent radiotherapy with a total dose of 5,600 cGy. The patient remains well with no signs of tumor recurrence at the follow-up examination.

Discussion

LMS typically arises in the soft tissues of the extremities and trunk.[3,4] Head and neck region involvement has been reported in only 3% of cases, with the majority occurring in the oral cavity.[1-5] Primary LMSs in the salivary gland are extremely rare, with only two cases of the submandibular gland reported in the literature.[1,5]

LMS occurs over a wide age range, with its peak incidence in the sixth decade of life.[4] The etiology of most sarcomas remains unknown.[5] LMS of the head and neck region are believed to develop from the tunica media of the blood vessels or pluripotent mesenchymal cells, which undergo differentiation during the course of neoplastic transformation.[2,5]

Signs and symptoms of LMS involving the head and neck region depend on the site and the size of the tumor.[2,3] Symptoms of LMSs in the submandibular gland are usually nonspecific, with a painless and progressive mass being the most significant finding, as in this case.[1,3,5]

The reported CT and magnetic resonance imaging features of LMS have been nonspecific.[3,5] Although the imaging studies are very helpful for surgical planning by adequate demonstration of the LMS extent, the specific radiologic diagnosis

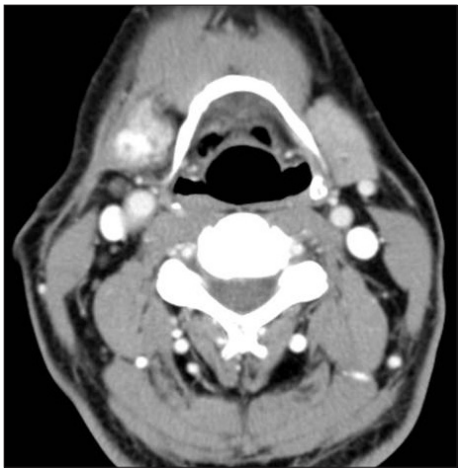

Fig. 1. Computed tomography with contrast enhancement shows an approximately 3×3 cm sized heterogeneously lesion of the right submandibular area without regional pathologic lymph nodes.

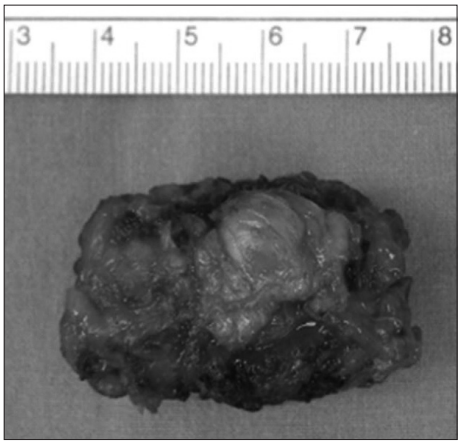

Fig. 2. Surgical specimen.

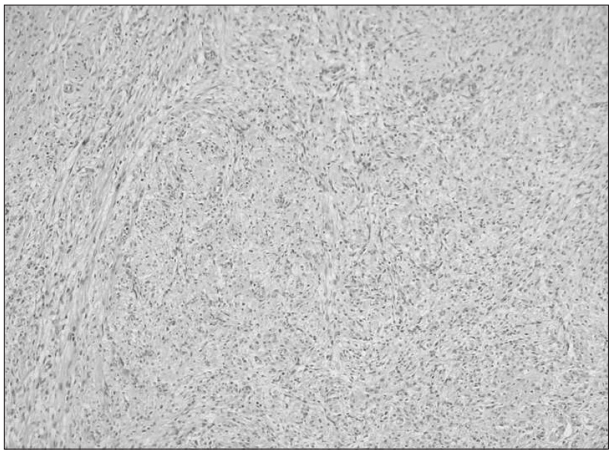

Fig. 3. Histopathologic section demonstrates spindle cells arranged in fascicular growth pattern(H-E stain, ×100).

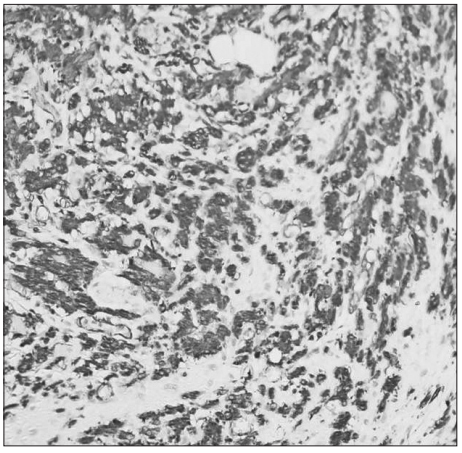

Fig. 4. Immunohistochemistry demonstrates positive staining of the tumor cells for smooth muscle actin(×200).

can be difficult.[4,5] In addition, diagnosis of LMS is difficult with small biopsy specimens by aspiration cytology.[3] A preoperative radiologic and pathologic diagnosis was not achieved in our case. The differential diagnosis of LMS may be challenging because of similar findings may occur in a wide number of different neoplasms, such as leiomyoma, spindle cell carcinoma, fibrosarcoma, myofibrosarcoma, rhabdomyosarcoma, malignant melanoma and inflammatory myofibroblastic tumor.[3,5] Therefore, immunohistochemistry is important for correct diagnosis.[1-5] The LMS is positive for smooth muscle actin and vimentin and negative for S-100 protein.[1-5] The reaction with desmin may be either positive, as in our case, or negative.[5]

The treatment of choice for LMS of the head and neck is the radical excision of the lesion.[1-6] Adjuvant radiotherapy is generally recommended for high-grade sarcomas, large tumors and close or positive surgical margins.[1,2,5] However, for LMS of the head and neck, the lesion is almost impossible to resect with a wide margin of safety because of the proximity of adjacent neurovascular structures or vertebral column.[2] Therefore, we recommended adjuvant radiotherapy for the most of LMS cases to reduce the risk of the recurrence, although it was controversial. In our case, we performed postoperative radiotherapy, because LMS was a close surgical margin and the possibility of delayed recurrent lesion 19 years after previous surgery.

The 5-year disease free survival accounts for 23%.[1] The recurrence rate of LMS has been reported as 40−60%.[3,5] LMSs are aggressive and tend to recur and undergo local metastasis in regional lymph nodes and at a distance in the lung.[4] The prognosis seems to related to the grade and size of tumor, location of lesion and type of treatment modality used.[5] Primary sarcomas of the major salivary glands usually present a worse prognosis than those arising from the surrounding tissues and secondary invading the gland.[1]

In conclusion, LMS of the submandibular gland is extremely rare. However LMS of the submandibular gland should be included in differential diagnosis, even when a well demarcated strongly enhancing tumor arising in the submandibular gland is demonstrated in radiologic examinations, and fine needle aspiration cytology revealed mesenchymal cells or spindle cells.[1,5]

References

1) Bucci T, Longo F, Mangone GM, Errico ME, Califano L. *Leiomyosarcoma of the submandibular gland. Report of a case and review of the literature. Int J Oral Maxillofac Surg. 2005;34(6): 690-692.*
2) Akcam T, Oysul K, Birkent H, Gerek M, Yetiser S. *Leiomyosarcoma of the head and neck: report of two cases and review of the literature. Auris Nasus Larynx. 2005;32(2):209-212.*
3) Haberal I, Alagöz S, Oksüzoğlu B, Tosun O, Ustün H, Samin EE. *A rare diagnosis in the head and neck region: Leiomyosarcoma. J Craniofac Surg. 2010;21(3):823-825.*
4) Rege IC, Costa NL, Batista AC, da Silva CM, Meneghini AJ, Mendonça EF. *High-grade primary leiomyosarcoma in the mandible: diagnosis and treatment. Head Neck. 2013;35(2):E44-E48.*
5) Boffano P, Roccia F, Zavattero E, Gallesio C, Cassarino E, Campisi P, et al. *The surgical management of a leiomyosarcoma of the submandibular gland in a 95-year-old patient. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2011;112(6):e34-e38.*
6) d'Adesky C, Duterme JP, Lejeune D, Mehta R, Chaikh A, Castadot P, et al. *Leiomyosarcoma of the inferior nasal concha: A report and literature review. B-ENT. 2012;8(3):213-217.*