ABSTRACT

A laryngeal leiomyosarcoma is a very rare tumor that originates from heterotopic mesenchymal tissues or smooth muscle cells. It is diagnosed by immunohistochemical staining.

Keywords: Glottis, larynx, leiomyosarcoma

Introduction

Squamous cell carcinomas constitute approximately 95%-98% of laryngeal tumors (1). Sarcomas are rarely encountered in the larynx, and their symptoms show similarity (2). Leiomyosarcomas originate from smooth muscles and constitute 5%-6% of all soft tissue sarcomas (1). On the other hand, the occurrence of laryngeal leiomyosarcoma (LLMS) is very rare (2). Its diagnosis is established based on only immunohistochemical staining techniques in histopathological examinations. In this case report, clinical and histopathological features of LLMS were examined.

Case presentation

A 65-year-old male patient was admitted due to complaints of hoarseness since 1 month and increasing dyspnea. He had no history of any disease but had a history of 45 year/pack smoking and social alcohol consumption. On indirect laryngoscopic examination, a pedunculated polypoid mass located in the anterior 2/3 of the left vocal cord was observed. Considering the presence of benign pedunculated polyp in the patient, no preoperative imaging was performed. The patient was planned to undergo mass excision through direct laryngoscopy. Neck examination of the patient revealed no pathological lymph node. The procedure was explained to the patient in detail, and his written informed consent was received.

On direct laryngoscopy performed under general anesthesia, a smooth polypoid mass holding on the anterior commissure and the left cord with a wide pedicle was observed (Figure 1). After the mass was cut from the junction point, the adhesion site in the anterior commissure and left vocal cord was removed with laser. Using frozen section analysis, malignant tumor cells were found; laser cordectomy was performed on the left vocal cord, and biopsy samples were obtained from the margins.

On pathological examination, 1.4×1-cm polypoid lesion covered with mucosa was observed macroscopically. Microscopic examination revealed a tumor consisting of fusiform cells including mitosis, ulceration, focal necrosis, and pleomorphism on the surface. On immunohistochemical examination, positive staining with vimentin, smooth-muscle actin (SMA), and caldesmon and negative staining with pancytokeratin, S-100, and CD31 were observed in the tumor (Figure 2-5). Based on immunohistochemical and histopathological findings, the case was diagnosed as pleomorphic leiomyosarcoma. Tumor tissue was not found both in the sample obtained using laser and in the second margin examination. Only the first piece
resected with its pedicle had tumor tissue. The patient was simultaneously administered postoperative chemoradiotherapy (cisplatin and curative RT-35 sessions-6000 rad).

This case was presented because of its rarity, the lack of a consensus on its treatment, and application of treatment with limited surgery.

Discussion

Laryngeal cancer is the most commonly seen cancer in the head and neck region. Most of these tumors are squamous cell cancers. Sarcomas constitute less than 1% of head and neck tumors (3). Leiomyosarcoma (LMS) often originates from the gastrointestinal system, uterus, and retroperitoneum, where smooth muscles are more commonly found. Head and neck LMSs constitute 3% of all LMSs (2). In the head and neck region, LMSs are encountered in the oral cavity, sinonasal region, scalp, neck, orbita, and cervical esophagus (1, 2). On the other hand, laryngeal sarcomas constitute a small proportion of all laryngeal tumors (2). Fibrosarcomas, chondrosarcomas, rhabdomyosarcomas, osteosarcomas, LMSs, and hemangiosarcomas develop in the larynx (4). The most commonly encountered sarcomas in the larynx are chondrosarcomas (1).

Following the first report of a LLMS case in 1939, approximately 50 cases have been reported (2, 5). LMS is encountered at a 4-time higher rate in men than in women (5). It is more common in the fifth decade of life (6, 7). Our case was a male patient in his sixth decade of life. Many factors, such as radiation exposure, surgery, tuberous sclerosis, neurofibromatosis, Werner syndrome, Gardner syndrome, and immunosuppression, have been reported to be responsible for the development of sarcoma (3, 8). In our case, none of these factors predisposing to the development of LLMS were present.

Laryngeal leiomyosarcoma often has glottic localization (48%), followed by supraglottic localization (9). In our case, a 1.5-cm well-circumscribed round mass with a pedicle in the anterior ½ of the right vocal cord caused an obstruction in the respiratory tract.

The diagnosis of LLMS is clinically difficult because it is macroscopically impossible to differentiate it from laryngeal pedunculated polyps, fibromas, and cancers (2). Patients with
LLMS present with complaints of progressive hoarseness, dyspnea, or dysphagia, as in other laryngeal tumors (1). The duration of symptoms can prolong from several weeks to a year (6). In our study, the patient consulted our hospital due to hoarseness that had begun 3 weeks ago and dyspnea becoming more severe from the preceding week. Tumor can display a very fast progression. Some cases requiring emergency tracheotomy and even emergency laryngectomy have been reported (2, 10). Furthermore, in our case, rapid progression of tumor within days was observed.

The histological type of sarcoma, grade, and occurrence of metastasis are the most important factors determining prognosis and treatment regimen. Neck lymphatic metastasis is rare in head and neck LMS (2). Neck or distant metastasis was not detected on PER CT in our patient. Distant metastases are generally not recognized on first admission. The most common metastases are seen in the lung, liver, bone, and brain (6). CT and MRI provide data on the tumor size, local extension, vascularity, and neck nodal state (5).

Immunohistochemical evaluations are required for diagnosis. Leiomyocysts express SMA and desmine, but not vimentin and S-100. Rhabdomyosarcomas, melanomas, schwannomas, malignant fibrous histiocytomas, and sarcomatoid cancers should be considered in the differential diagnosis (2).

Primary treatment choice for LLMS is surgery (5). The best prognosis is obtained by completely removing the tumor with wide surgical margins (11, 12). In the absence of neck metastasis, neck dissection is not required (6). Chemoradiotherapy may have a role in local recurrence and residual tumor as an adjuvant therapy, but it is ineffective as a primary treatment (2).

Conclusion

Laryngeal leiomyosarcoma is rarely encountered, and its symptoms can be confused with those of other laryngeal tumors. In our case, because tumor restrictively held on the cord, clean surgical margins were obtained through cordectomy, and adjuvant chemoradiotherapy was administered. Our patient has completed 9 months without the disease.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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References