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LEIOMYOSARCOMA OF THE LARYNX: A COMPLEX DIAGNOSIS.

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CASE REPORT

An 84-year-old man was referred to our institution for dysphonia present for 2 months. Fiberoptic laryngoscopy demonstrated a polypoid lesion of the right true vocal fold with normal motility.

Excisional biopsy of a submucosal thickening of the right vocal fold with vocal ligament calcification was performed. Histology revealed squamous cell epithelium without malignancy. Post-operative endoscopic follow up was regular.

Six months after surgery, dysphonia and dyspnea occurred. Flexible laryngoscopy demonstrated right hemilaryngeal paralysis, edema and partial airway obstruction. Necrotic tissue and purulent secretions were visible at the anterior commissure, right vocal fold and ipsilateral vestibule. A second microlaryngoscopic surgery included multiple biopsies and debridement of the necrotic tissue. Histological report was negative for malignancy, and purulent chondritis was diagnosed. Intravenous antibiotic therapy was administered with improvement of infection, inflammation, dysphonia, and dyspnea. The patient was discharged.

Two months later, a new sudden and severe dyspnea occurred. He was admitted to the E.R., and oropharyngeal intubation was necessary because of respiratory failure.

CT-scan showed thickening of the right laryngeal wall with wide fluid collection (48-43 mm diameter) that almost completely obstructed the airway, with erosion of the right thyroid and cricoid cartilage (Figure 1).

Debridement of the necrotic tissue and drainage of the purulent collection was performed with direct laryngoscopy. Culture was positive for *Staphylococcus epidermidis* and *Candida albicans*. Intravenous antibiotic, antimicotic and corticosteroid therapy was administered, and other 3 surgical debridements and drainage were performed, but without clinical or

radiological improvements. Moreover, patient developed signs of sepsis. Since no improvement had been achieved, total laryngectomy was planned even without a positive histologic biopsy. Intraoperative findings showed trans-glottic extension of the ulcerated material with full-thickness infiltration of the laryngeal wall and external spread through the thyroid cartilage, with invasion of pre-laryngeal tissues (Figure 2). The patient's clinical condition improved, and he started oral feeding without pharyngo-cutaneous fistula. Morphological and immunohistochemical features were consistent with leiomyosarcoma (FNCLCC Grade 3). (Figure 3).

Multidisciplinary consultation suggested adjuvant radiation therapy (RT), but pre-irradiation staging CT-scan showed rapid local recurrence and lung metastasis. The poor clinical condition of the patient contraindicated 1st line chemotherapy. At 3-month-follow-up, the patient was alive and receiving best supportive care.

DISCUSSION

The first case of laryngeal leiomyosarcoma (LLMS) was reported by Jackson et al. in 1939.[1] After that, fewer than 50 cases of LLMS have been reported in international literature,[2] including an 87-year-old man with laryngopyocele and LLMS who was treated with total laryngectomy.[3]

Simultaneous presence of laryngeal carcinoma, laryngopyocele, and laryngomucocele was described by Cassano et al[4], and Mitro et al[5], but no leiomyosarcoma was found in these patients or in cases reviewed in similar articles.

Purulent chondritis of laryngeal cartilages (PCLC) is a very uncommon abscess that grows between inner and outer perichondria of the laryngeal framework. To date, only 5 cases have been described in the literature[6-8], and none of them was associated with neoplasm.

In our case, radiological suspicion of laryngeal abscess and no clear radiological signs of malignant tumor were detectable. Laryngeal biopsies were negative for malignancy, and cultures were consistent with PCLC, but combined antibiotic therapy, surgical drainage and debridement did not improve the clinical condition, and the patient developed sepsis. Fortunately, after total laryngectomy, his clinical condition improved quickly with complete sepsis resolution.

Etiology of LLMS is still unclear. A possible factor is an abnormal post-operative healing process.[9] In our case, the patient developed worsening dysphonia and dyspnea, six months after his first vocal fold biopsy with negative findings.

Treatment guidelines for LLMS are lacking due to its rarity. First line treatment should be tumor resection with wide margins, reserving radiotherapy as adjuvant treatment or in case of recurrence or persistent tumor.[10,11] Chemotherapy usually is suggested for distant metastases[12].

Even though if LLMS is a very rare laryngeal malignancy, it should be considered when dealing with laryngeal abscess, especially with cartilage involvement.

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FIGURE LEGENDS

Figure 1. CT image showing an almost complete erosion of the laryngeal framework cartilages due to LLMS. Residual airway is minimal, therefore an emergency oropharyngeal intubation is needed.

Figure 2. Intraoperative picture showing the almost complete erosion of laryngeal cartilages with necrotic material substituting normal laryngeal tissue.

Figure 3. Histopathology of the lesion: SMA (Smooth Muscle specific Actin) positivity demonstrates the smooth muscle nature of the neoplastic population. Written histological report shows a malignant neoplasm with spindle cells, high mitotic index (10-14 mitosis x 10HPF) and large areas of tissue necrosis (>50%).