Leiomyosarcoma of the larynx

B. M. Lippert · E. Schlüter · H. Claassen
J. A. Werner

Leiomyosarcoma (LMS) is a malignant tumor containing cells derived from smooth muscle. Anatomic sites principally affected are the uterus, gastrointestinal tract and retroperitoneal area [26]. LMS of the head and neck region are uncommon, with tumors extremely rare in the larynx. To our knowledge only 27 cases of LMS of the larynx have been reported in the literature [1, 3–10, 12–17, 19, 20, 23–25, 27, 28, 31, 32].

We now describe a case of laryngeal LMS of the left vocal fold and discuss the diagnostic problems had, as well as the therapy and prognosis associated with this rare tumor.

Case report

On 14 December 1994 a previously healthy 50-year-old white German officer presented with a 2-month history of hoarseness and was seen at the Department of Otorhinolaryngology, Head and Neck Surgery of the University of Kiel, Germany. Indirect laryngoscopy using a magnifying lens disclosed a pale, slightly polypoid, exophytic lesion in the anterior third of the left vocal fold. The anterior commissure was unobstructed and vocal fold mobility was seen to be intact. Palpation of the neck revealed no enlargements of cervical lymph nodes.

During subsequent direct microlaryngoscopy, excisional biopsy was performed (Fig. 1) using the CO₂ laser. Histologic examination of the tumor material by the Department of Pathology revealed a well-differentiated LMS, but with findings of residual tumor at the margins of excision. A second CO₂ laser procedure succeeded in removing all tumor residual. Further study included computer tomography of the brain, neck and thorax as well as neck and epigastric sonography, skeletal scintigraphy, gastroscopy and colonoscopy but did not reveal any signs of tumor dissemination or another primary tumor.

Routine follow-up examination in November 1995 showed redness of the left vocal fold. However subsequent microlaryngoscopy and biopsy showed no macroscopic or histologic signs of tumor recurrence (Fig. 2). At last follow-up (January 1997), the patient was free of symptoms and had no signs of tumor. At present he has a scarred vocal fold that permits nearly complete glottic closure and easily understandable speech.

Histopathology

The smooth polypoid biopsy specimen from initial surgery exhibited a typical stratified squamous surface epithelium. Situated immediately beneath the epithelium was a cell-rich tumor composed of interlacing bundles of smooth muscle fibers with elongated spindle cells. Some of the
nuclei were plump and cigar-shaped (Fig. 3), with a moderately coarse chromatin and 5 mitoses/10 high power fields (HPF) at a 200-fold magnification. Immunohistochemistry revealed moderate intracytoplasmic expression of the intermediary filaments vimentin and desmin. All of the tumor cells were markedly positive for smooth muscle antigen (Fig. 4), but were negative for neuron-specific enolase, S-100 protein and cytokeratin KL-1. Electron microscopic demonstration of myofibrils, pinocytotic vesicles and desmosomes confirmed the diagnosis of an LMS.

Discussion
Sites of LMS reported in the literature include the facial skin [26], nose and paranasal sinuses [18], oro- [11] and hypopharynx [30] and trachea [29]. LMS of the larynx was first reported by Jackson and Jackson [13] in 1939. these latter tumors usually occur in middle age, although Chizh [6] has described an LMS in an 8-year-old girl. Men are more often affected than women. The majority of the laryngeal tumors have been situated in the supraglottic area, while only a few cases of tumor confined to the vocal fold have been reported [2, 20, 24]. Symptoms are not characteristic and, as in the present case, do not differ from those of other laryngeal malignancies.