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Laryngeal pleomorphic rhabdomyosarcoma

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Abstract

Adult rhabdomyosarcoma of the larynx is a rare disease. Two cases of laryngeal rhabdomyosarcoma of the pleomorphic subtype are presented. One case was treated with surgery followed by chemotherapy and radiotherapy, the other by surgery alone. At present 20 months after treatment the patients are without local reccurrence or metastases. The pathology of the tumor, diagnosis, and treatment are discussed.

Keywords: Pleomorphic rhabdomyosarcoma; Laryngeal; Treatment; Prognosis

1. Introduction

Laryngeal cancer constitutes about 25% of all head and neck cancers. Over 95% of the malignant soft tissue neoplasms of the larynx are squamous cell carcinomas [1]. Primary laryngeal sarcomas are rare, representing less than 1% of all malignant tumors of the larynx. More than 50% of laryngeal sarcomas are fibrosarcomas, followed by chondrosarcomas, osteosarcomas, leiomyosarcomas, liposarcomas and rhabdomyosarcomas. While rhabdomyosarcomas account for 40% of all sarcomas found in the head and neck region [2] primary pleomorphic rhabdomyosarcoma of the larynx seems to be exceedingly rare and only a few case reports have been published so far. We report on two cases of laryngeal pleomorphic rhabdomyosarcoma.

2. Case 1

On admission in our department the 60-year old male patient presented with a 4 weeks history of hoarseness. He never smoked or consumed alcohol and his clinical history was otherwise unremarkable. Physical examination displayed a smooth mass arising from the left ventricular band with paralysis of the left vocal cord in paramedian position. CT scans of neck and thorax revealed a left transglottic tumor crossing the mid-line posterioly, infiltrating the left thyroid and arytenoid cartilage. No local or systemic metastases were detected (Fig. 1). Blood chemistry was within normal limits. Direct microlaryngoscopy showed a solid mass arising from the median septal wall of the left sinus piriformis protruding into the laryngeal lumen. The lesion was covered with normal mucosa without any ulcerations. A biopsy was taken and immunhistochemical stainining was positive for desmin, myogenin, sarcomeric aktin and negative for cyclin D1, and caldesmon. All findings were in agreement with the diagnosis of a pleomorphic rhabdomyosarcoma (Fig. 2). A total laryngectomy and a bilateral selective neck dissection due to the large size of the primary tumor was performed. The 3.8 cm tumor mass originated from the left true vocal cord, invaded the perilaryngeal soft tissues but did not affect the laryngeal cartilage. The neck dissection specimen contained 12 lymph nodes without metastasizing disease. Final pathology staging revealed an R0 resection of the tumor (pT3 pN0 cM0, G3) with very close margins (0.2 mm). Due to the high rate of local recurrence treatment was performed according to the CWS 96 protocol consisting of adjunctive chemotherapy with three cycles of Ifosfamid and Adriblastin followed by external radiotherapy (44.6 Gy covering the

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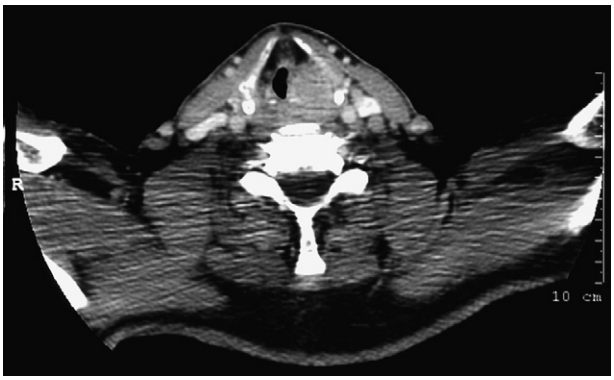

Fig. 1. Axial CT scan of the neck revealed a transglottic tumor crossing the mid-line, infiltrating the left thyroid and the arytenoid cartilage.

neck between skull base and the sternoclavicular joint and 60 Gy directly to the primary tumor site) and a complementary forth cycle of chemotherapy.

3. Case 2

A 64-year-old man was admitted to our department with a 6-month history of dyspnea on exertion and a 2-week antibiotic-resistant hoarseness. Besides smoking (35 PY) neither excessive alcohol consumption nor other relevant clinical history could be recorded. Indirect laryngoscopy demonstrated a polyploid mass arising from the left vocal cord, without any restriction of mobility. On palpation and sonography no lymph nodes were detected in the neck region. Chest X-ray and blood chemistry were within normal limits. On direct microlaryngoscopy the mass was seen to originate from the left vocal cord extending into the subglottis. Normal mucosa without any ulceration was covering the lesion. The tumor (Ø1.7 cm) was removed by endoscopic lasersurgery (CO_2-laser). Histopathology displayed a diffuse cellular tumor with numerous large round and few spindle-shaped cells. A pleomorphic rhabdomyosarcoma was diagnosed by immunhistochemical staining as described above. The following computed tomography scans of the neck revealed no contrast enhancement of the larynx as sign of malignancy. Neither bone erosion nor pathologic lymphatic nodes were found. However a hypodense area of the left thyroid lobe 2 cm in diameter was noted. Abdominal sonography did not display any pathological findings. After 1 week a endoscopic lasersurgical follow-up resection of the left vocal cord (from the anterior commissura to the processus vocalis) and a

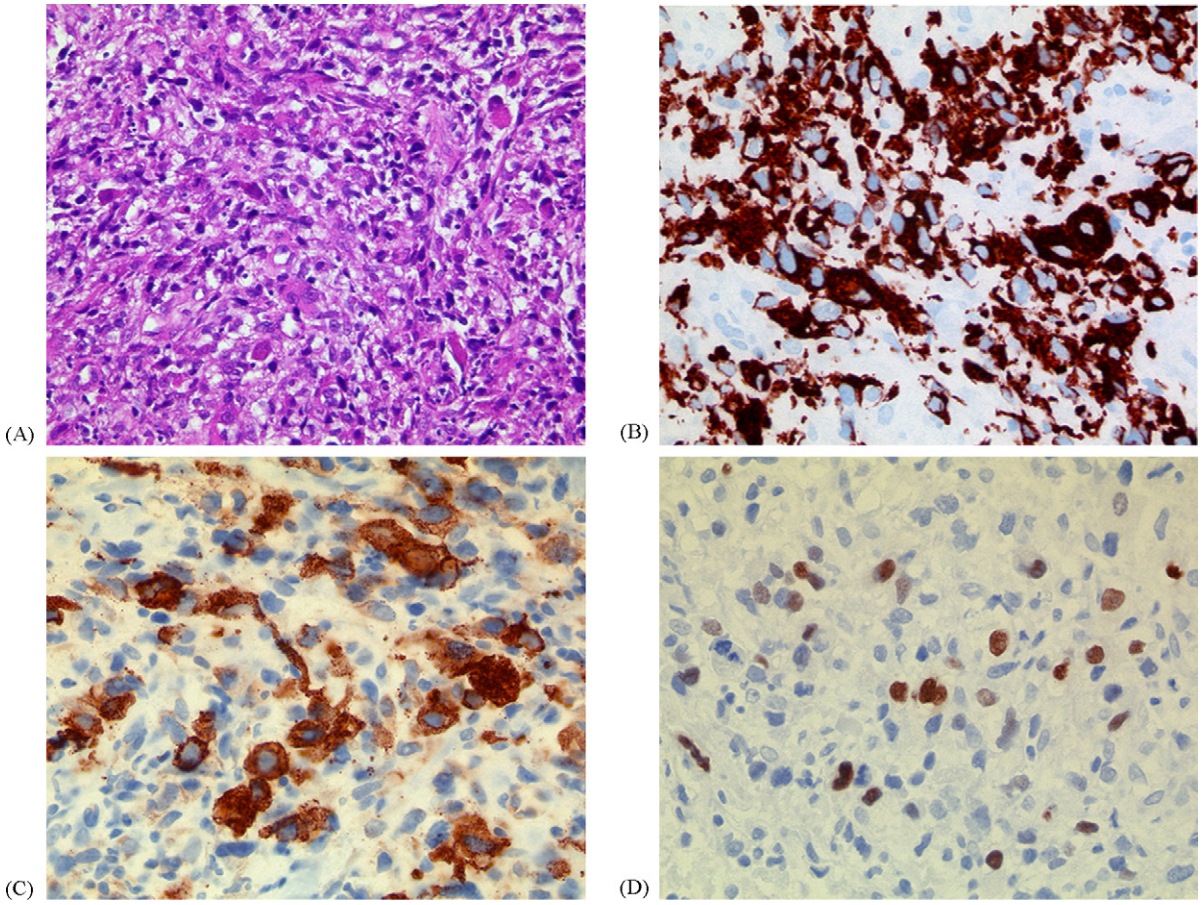

Fig. 2. Photomicrographs of the rhabdomyosarcoma from case 1 consisting of spindle and pleomorphic tumor cells (A, H&E, objective 40×) staining positive for antibody to desmin (B, objective 40×), sarcomeric actin (C, objective 40×, nuclear staining) and myogenin (D, objective 40×, cytoplasmatic staining).