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Primary tracheal leiomyosarcoma

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Abstract
We present the clinical course of a 56-year-old female patient with a primary tracheal leiomyosarcoma. The diagnostic approach and pathological classification of this seldom described tumour remains extremely difficult. We discuss the symptoms as well as the diagnostic and therapeutic procedures, including multimodal chemotherapy with organ-preserving surgery leading to complete remission.

Key words: Leiomyosarcoma; Trachea; Surgery, operative; Chemotherapy, adjuvant

Introduction

Leiomyosarcomas are malignant tumours showing smooth muscle differentiation. They are rarely found in the trachea, with only 20 cases reported in the literature since 1950. We report on the clinical course of a 56-year-old female patient with a leiomyosarcoma of the cervical trachea.

Case report

A 56-year-old, previously healthy female presented to the ENT outpatient department after two months of progressive dyspnoea and stridor. A flexible fibre-optic laryngotracheoscopy revealed a subglottic mass almost completely obstructing the upper airways. Acute dyspnoea during the examination required tracheostomy. The subsequent rigid tracheoscopy demonstrated a fleshy polypoidal tumour inserting in the right paramedian area of the posterior tracheal wall and spreading caudally over a length of 1.5 cm from 1 cm below the glottic plane.

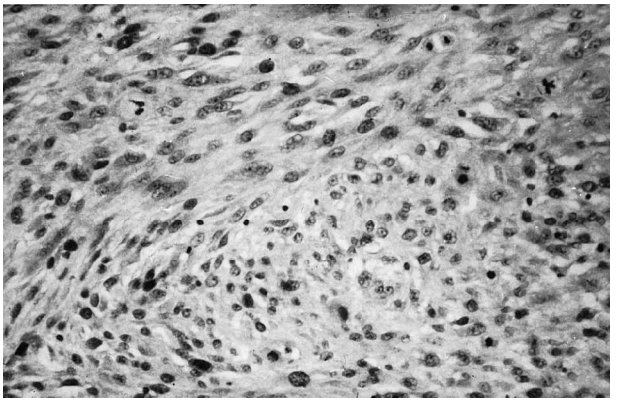

Fig. 1

Classical spindle-cell component of the leiomyosarcoma. The figure shows some atypical mitoses (H & E; × 250-fold magnification).

This enabled a transglottic removal of the tumour. Histopathological examination of the surgical specimen disclosed an ulcerated moderately differentiated leiomyosarcoma with spindle-shaped cells and some epithelioid cells. The tumour was positive for vimentin and negative for cytokeratin. The tumour cells expressed in part smooth muscle actin and desmin. The resection margin showed microscopic tumour invasion (R1 resection). In the postoperative staging (cervicothoracic computed tomography (CT), bone scintigraphy, abdominal ultrasound) no further tumour manifestation was found, thus yielding a UICC $pT_1N_0M_0$ stage.

We decided to administer neoadjuvant polychemotherapy according to the EVAIA protocol[1] followed by a potentially curative laryngectomy. This involved adminis-

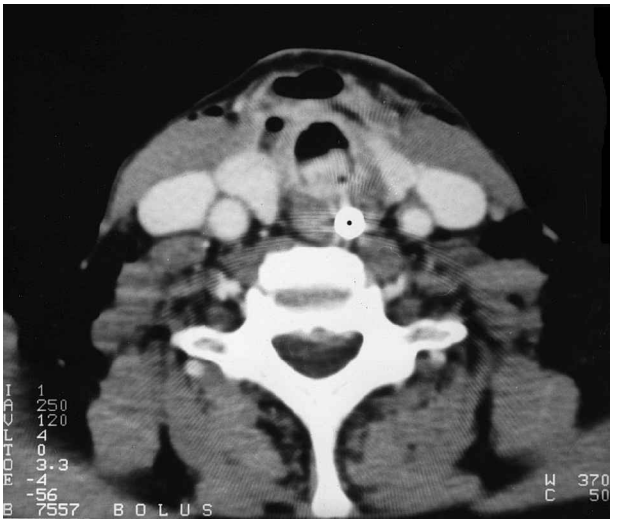

Fig. 2

Cervical CT: The scan shows a subcutanous emphysema after tracheostomy. The posterior tracheal wall shows a subglottic mass partially blocking the lumen of the trachea. A gastric tube in the oesophagus can be seen. There is no indication of regional lymph node involvement.

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tration of four three-day cycles of both ifosfamide 2 g/m^2 and Vepesid®-16 150 mg/m^2 with vincristine 1.5 mg/m^2 once per cycle and adriblastin 20 mg/m^2 and actinomycin 0.5 mg/m^2 for two alternating three-day cycles each. There was a total of four cycles after one, four, seven and 10 weeks.[1,2] Histological restaging after four cycles of chemotherapy revealed no more evidence of tumour tissue. We, therefore, decided against the laryngectomy originally planned and carried out percutaneous irradiation of the primary tumour region including the regional (paratracheal) lymph nodes up to a total dose of 66.6 Gy, after closure of the tracheostomy. After a period of 48 months from the time of diagnosis the patient is still free of recurrence.

Discussion

Malignant tracheal tumours are found very rarely with an incidence of one case per one million population. Histologically, most of them are squamous cell or adenoid cystic carcinomas,[3] whereby only 20 tracheal leiomyosarcomas have been reported in the literature since 1950.[4–23] Leiomyosarcomas can occur in all tissues containing smooth muscle cells, but the most common localizations are the extremities, the gastrointestinal tract, the retroperitoneal space and the urogenital tract. Head and neck incidence amounts for only 15 per cent of all cases.[24] Pulmonary leiomyosarcoma may occur at any age, and is much more common in females.[13] Due to their low incidence the prognosis for these tumours remains unknown. However, for all primary tracheal tumours, Morrison and Salkeld[3] found the mean five-year survival rate to be 30 per cent. They described a five-year survival rate for intraluminal tumours of 50 per cent, in contrast to an 18 per cent five-year survival rate with extratracheal extension.

Tracheal tumours usually manifest themselves with nonspecific clinical signs such as persistent coughing, sanguineous sputum, increasing dyspnoea occasionally associated with supra- and infrasternal retraction. The differential diagnosis includes inflammatory diseases of the larynx and trachea or the lung. Tracheobronchoscopy, including histological confirmation by biopsy, is the diagnostic method of choice. Since tracheal leiomyosarcomas are extremely rare, no data is available on the probability or localization of metastases. The tumour, as is common for most sarcomas, spreads via the haematogenous route.[13] In consideration of the anatomical lymphatic and venous tracheal drainage, however, we advocate cervicothoracic CT prior to therapy for exclusion of peritracheal lymph-node involvement and lung metastasis.[25] In our practice all tumours of the upper aerodigestive tract have further staging by bone scintigraphy and abdominal ultrasound.

Total resection of malignant tracheal tumours is generally recommended if primary reconstruction appears possible.[25] A laryngectomy and tracheal resection should be performed for a high tracheal tumour or laryngeal infiltration.[20] The majority of hitherto published patients with tracheal leiomyosarcomas underwent radical excision and lymphadenectomy.[8] For leiomyosarcomas in other localizations, more recent studies recommend a multimodal procedure consisting of radical excision, irradiation and polychemotherapy.[1] Recurrence-free survival for chemotherapy-sensitive soft tissue sarcomas was 64 per cent in the CWS-E study with treatment following the EVAIA scheme and subsequent irradiation. The survival rate was 79 per cent for a mean observation period of 46 months.[1] The five-year survival rate after surgical therapy alone is 26 to 76 per cent depending on the sarcoma differentiation, limiting factors being local recurrence and distant metastases.[24]

Our patient, who underwent primary emergency endotracheal tumour extirpation, had an inadequate resection margin with residual tumour. To achieve a tumour clearance, a laryngectomy and partial tracheal resection would have been necessary. Instead, we treated the patient with multimodal chemotherapy following the EVAIA protocol of the CWS-E study and subsequently irradiated the tumour bed.

Neither clinical nor histological examination has detected any residual tumour after the completion of therapy. The patient has been recurrence-free after a follow-up period of 48 months. To these authors' knowledge, this will be the first complete remission of a primary tracheal leiomyosarcoma after treatment by organ-preserving surgery with subsequent chemotherapy and percutaneous irradiation.

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