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Synovial sarcoma of the larynx: case report and literature review

V. Boniver, P. Moreau and Ph. Lefebvre

Department of Otorhinolaryngology, Head and Neck Surgery, University of Liège, CHU Sart Tilman, Liège, Belgium

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Abstract. *Synovial sarcoma of the larynx: case report and literature review.* Synovial sarcoma is a rare mesenchymal malignancy which represents 8.5% of all soft tissue sarcomas and usually occurs in the lower extremities of young adults. Because the incidence in the head and neck region is very low, only 13 patients with endolaryngeal localisation have been reported so far. We present here a case of aryepiglottic synovial sarcoma. The tumour was completely resected, under suspension micro-laryngoscopy, using a CO_2 laser. No recurrence was observed three years after the surgery. Complete surgical excision is the treatment of choice. The role of chemotherapy and radiotherapy is still debated. However, this tumour has a poor prognosis because of the occurrence of distant metastasis.

Introduction

Synovial sarcoma is a rare mesenchymal malignancy which represents 8.5% of all soft tissue sarcomas occurring most often in the lower limbs of young adults. The occurrence in the head and neck varies from 0% to 16% according to the reported literature.[1] The usual sites involved in the head and neck are the prevertebral and hypopharyngeal regions. True endolaryngeal localisations of these tumours are very infrequent and were previously reported in only 13 patients. Synovial sarcoma usually occurs in patients between 20 to 40 years,[2] with men outnumbering women 3:2. Discovery of synovial sarcoma in the head and neck usually results from symptoms of hoarseness, dysphagia, pain and dyspnea. Despite its name, this lesion does not originate from synovial membranes but from pluripotent mesenchymal cells.[3]

Case report

A 30-years-old non-smoking woman presented to the clinic for sore throat. She had neither dysphonia nor dyspnea. A progressive pain located on the left side of the neck appeared three months before the consultation. The pain was enhanced during swallowing. She had no haemoptysis and never had complaints of epigastric pain. Indirect laryngoscopy and fibrolaryngoscopy were performed which revealed a mass arising from the left aryepiglottic fold (Figure 1). The mobility of the larynx was normal. The surface of the tumour was smooth and seemed to be covered by normal mucosa. No lymph nodes were palpable.

The patient was taken to the operating room for a panendoscopy. The tumour measured approximatively 2 cm in diameter and was pedonculated on the left aryepiglottic fold. It was firm and fixed. The remainder of the larynx and the hypopharynx was normal. A biopsy was performed during panendoscopy and the histological examination revealed a synovial sarcoma with a biphasic pattern.

A complete extension work up was made. Computed tomography (CT-scan) of the head and neck confirmed the pedonculated, well circumscribed tumour without lymphe node involvement (Figure 2). The chest CT-scan and the body positron emission tomography (PET) with ^{18}F-fluorodeoxyglucose didn't reveal any metastasis.

The tumour was removed with the use of an endoscopic CO_2 laser. Complete tumour exposition was possible in this young patient; an important condition for endoscopic resection. No tracheotomy was performed. The tumour was completely resected and all the margins were histologically safe with the nearest tumour at 1 mm in depth. The size of the tumour was $2.2 \times 2 \times 1.7$ cm (Figure 3).

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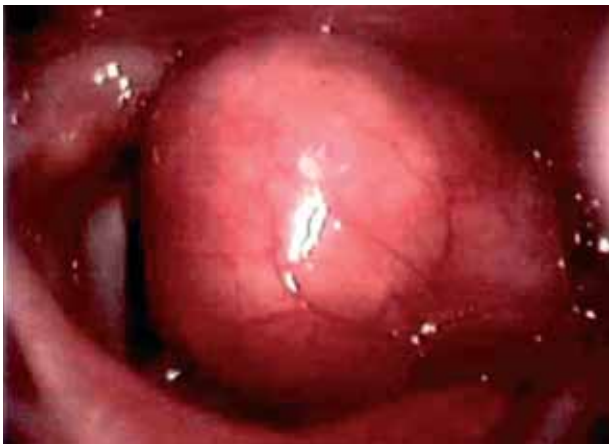

Figure 1
Preoperative view of the smooth tumour obliterating the left hemilarynx.

Figure 3
Resected tumour with the CO_2 laser

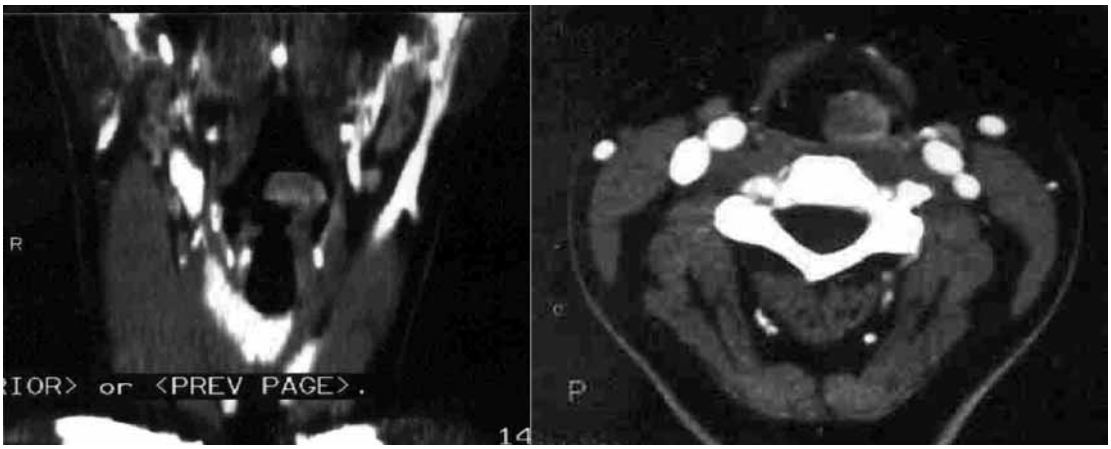

Figure 2
CT Scan showing a pedunculated mass on the left arytenoids

The nodule was covered by apparently normal mucosa and on section the tumour was grey and solid. At microscopic level, the tumour presented two patterns: an epithelial glandular like pattern and a mesenchymal pattern with spindle cells. The surface mucosa was focally infiltrated by tumour cells. Immunohistochemically the epithelial cells were positive for epithelial cells markers: cytokeratin KL-1 and EMA. The spindle cells expressed the mesenchymal marker vimentin. Both were positive for CD99 and CD117. The diagnosis of biphasic synovial

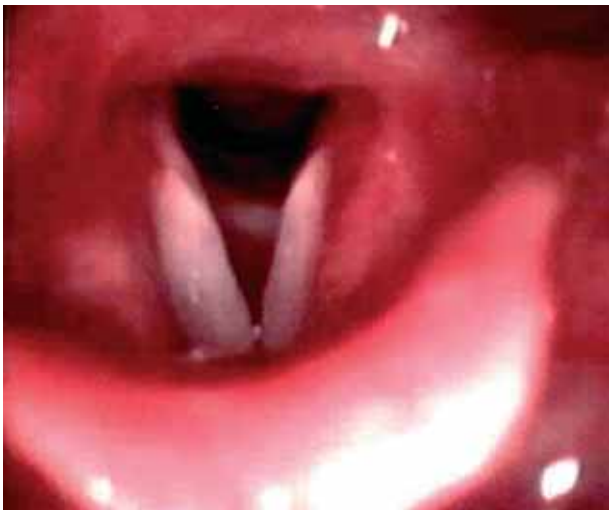

Figure 4
Postoperative view with minimal scar

Table 1
Endolaryngeal synovial sarcomas: reported cases

Reference	Age/Sex	Location	Therapy	Follow-up
Miller[4] Gatti[5] (same case)	23/F	Interarythenoid	Supraglottic laryngectomy Total laryngectomy	+ margins NED 12 years
Geachan[6]	24/M	Left arythenoid and left epiglottic fold	Partial laryngectomy Total laryngectomy	Recurrence after 4 years Lung metastasis after 6 years
Quinn[7]	76/M	Right subglottic	Extended frontolateral laryngectomy	NED 3 years
Kistmaniuk[8]	15/M	Left aryepiglottic fold and pyriform sinus	Extended total laryngectomy	NED 8 months
Kleinsasser[9]	58/F	subglottic	Partial laryngectomy	Recurrence after 7 years
Prusczynski[10]	28/F	Left aryepiglottic fold and false cord	Endoscopic resection + radiotherapy	NED 3 years
Ferlito[11]	28/M	Right aryepiglottic fold and epiglottis	Radiotherapy + supraglottic laryngectomy with neck dissection+ radiotherapy	NED 16 years
Danninger[12]	53/M	Right aryepiglottic fold and pyriform sinus	Total laryngectomy with neck dissection + radiotherapy	NED 16 months
Morland[13]	14/M	Left arythenoid	Tracheotomy + chemotherapy + radiotherapy Total laryngectomy	Recurrence after 3 years NED 2 years
De Itos[14]	27/F	Right aryepiglottic fold	Endoscopic resection Chemo + radiotherapy Partial laryngectomy	Recurrence after 3 months Persistent 3 months NED 9 months
Taylor[15]	68/F	Cricoid cartilage	Extended total laryngectomy + neck dissection	–
Papaspyrou[16]	16/M	Right aryepiglottic fold	Laser endoscopic resection + radiotherapy	NED 2 years
Bilgic[17]	24/M	Left aryepiglottic fold Epiglottis and left arythenoid	Hemilaryngectomy Total laryngectomy + neck dissection + radiotherapy chemotherapy	Recurrence 1 year Lung metastasis 20 months NED 3 years
present case	30/F	Left aryepliglottic fold	Laser endoscopic resection	NED 3 years

NED: no evidence of disease.

sarcoma was confirmed. The tumour presented a KI67 index of 10%.

The patient did not receive adjuvant therapy. The postoperative course was uneventful and the patient didn't complain of any pain or dysphagia anymore. Fibrolaryngoscopy returned to normal (Figure 4). There is no evidence of recurrence 3 years after surgery.

Discussion

Synovial sarcoma is a rare malignant tumour that has no benign counterpart. Only 13 cases with endolaryngeal origin are described in the literature. Table 1 summarizes the clinical data for all the endolaryngeal synovial sarcomas reported so far including the present study.

The most common complaints are dysphagia, increasing hoarseness and shortness of breath on exertion.[2] In most of the cases, the tumour arises from the aryepiglottic fold.[4,6,10-12,14,16,17] The tumour

appears as a mass covered by an intact mucosa and sometimes pedonculated. Histologically, the tumour often presents a biphasic pattern consisting of a spindle cell stroma and foci of epitheloid cells forming clefts and acini. Calcification may be seen. The monophasic form of synovial sarcoma is uncommon and its diagnosis is difficult. When only the spindle cell component is present, the tumour can be misdiagnosed as a fibrosarcoma. The presence of vascular structure, when the epithelial cells predominate, may be misleading and prompt an erroneous diagnosis of adenocarcinoma metastasis. Immunohistochemistry helps for the diagnosis as the tumour stains for vimentin and cytokeratin. A KI67 index of 10% or more is considered highly proliferative.[18] Cytogenetically, the tumours are characterized[14] by the reciprocal translocation t(X;18) (p11.2; q11.2). This may be helpful in case of a monophasic form of the tumour. Synovial sarcomas have generally a poor prognosis. The survival rate is not any better than in the extremities with a 40% 5-year survival rate.[2] One of the most important prognostic indicators of survival is tumour size. Patients with a primary tumour larger than 4 cm in diameter have a poorer outcome than those with smaller tumour.[3,6,14,18,19] As in the case of other soft-tissue sarcomas, surgery is the treatment of choice. Excision must be done widely and margins proven to be negative. In the reported cases of laryngeal synovial sarcomas, no lymph node metastasis were described.[11,17] Neck dissection is not indicated without lymph node involvement. Distant metastasis may develop several years after the initial diagnosis and pulmonary metastasis is the usual cause of death.[3,5] Postoperative radiotherapy for synovial sarcoma of the extremities has not been found to increase survival.[3] In head and neck localisation, postoperative radiation is often a part of the treatment but proof of its efficiency on local control or distant metastasis is lacking. Radical surgical procedure may be difficult in head and neck area. However radiation therapy should not be considered as a salvage therapy for functional structure preservation. The role of chemotherapy is controversial. Ifosfamide has been investigated but it is difficult to distinguish its real beneficial effect and what represents ideal sequencing of treatment modalities.[14] Two other cases treated by endoscopic resection with postoperative radiation have been reported[10,16] but in the present case the treatment was endoscopic resection alone. We made the choice of only surgical treatment to keep maximal chances to detect early local recurrence and the possibility of external surgery and radiotherapy if necessary.

Conclusion

Synovial sarcoma is a rare tumour in the head and neck region. However we believe that in selected cases endoscopic resection with laser removal is a valuable treatment. The tumour must be completely accessible and the quality of the resection depends on the surgeon's experience. Three patients have been treated so far with this technique. The indication is a pedonculated tumour less than 4 cm in size with perfect view of its implantation site and normal larynx mobility. This seems to be safe for local control allowing a good laryngeal function.

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Vincent Boniver, M.D.
ENT Department
CHU Sart Tilman B35
B-4000 Liège, Belgium
E-mail: vincent.boniver@docs.be