Synovial cell sarcoma: A rare laryngeal tumor

Le synovialosarcome: Une tumeur laryngée rare

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RÉSUMÉ

Le synovialosarcome est une tumeur des tissus mous se développant principalement au niveau des membres inférieurs, près des articulations. La tête et le cou en sont des localisations extrêmement rares. Le larynx est le site d'atteinte le moins fréquent, avec seulement près de 20 cas publiés à ce jour. Pour cette raison, le traitement de ces tumeurs reste encore controversé, bien qu'il devrait suivre les lignes directrices établies pour les autres localisations. Nous rapportons les cas d'un patient âgé de 37 ans, porteur d'un synovialosarcome primitif du larynx, traité chirurgicalement par une laryngectomie carrée.

Mots-clés

Synovialosarcome, larynx, immunohistochimie, laryngectomie, radiothérapie

SUMMARY

Synovial cell sarcoma is a soft tissue tumor that occurs predominately in the lower limbs near the joints. Lesions of the head and neck are extremely rare. The larynx is the least frequent site with only about 20 cases reported in the literature. Treatment of these tumors is controversial and should follow the guidelines for other tumor sites. We report the case of a 37-year-old man with primary laryngeal synovial cell sarcoma, who was surgically managed by a narrow field laryngectomy.

Key-words

Synovial cell sarcoma, Larynx, Immunohistochemistry, Laryngectomy, Radiotherapy

Synovial cell sarcoma is an aggressive mesenchymal tumor, which occurs most commonly in the lower limbs accounting for around 10% of soft tissuessarcoma(1). Only 3-9% of this histological form occurs in head and neck(2), most commonly in the pharynx where it presents as a slowly enlarging mass. The larynx is the least frequent site for synovial cell sarcoma (1, 2, 3). Its clinical behavior is aggressive for either high or low-grade lesions. Lymphnode metastasis is rare and when it occurs it is generally associated with high-gradetumors (4). Local recurrencesare usual. Immunohistochemistry is the key to histological diagnosis.

This report reviews the clinical presentation, diagnosis and the therapeutic option chosen in a case of laryngeal synovial cell sarcoma in a 37-year-old man.

CASE REPORT

A 37-year-old man presented to our service with an enlarging anterior neck swelling and hoarseness of the voice worseningfor 9 months, but without pain or dysphagia. Foreign body sensation in the throat, snoring and recent dyspnea were also mentioned. The patient has no remarkable medical history. He was a heavy smoker, an occasional alcohol consumer and works as a driver. An ill-defined-5cm- mass was noted anterior to the thyroid cartilage, but no lymph nodes waspalpated.

Suspension laryngoscopy has shown anirregular red and bloody laryngeal mass arising from the subglottis, obliterating 50% of the larynx.

Multiple biopsies were obtained. CT scan revealed a 46x38mm soft tissue mass, extending to the glottis and sub glottis, destroying the thyroid cartilage. Histopathology and immunohistochemistryrevealed a biphasic synovial cell sarcoma with epithelioid cells intermixed with fusiform cells.

A narrow field laryngectomy, total thyroidectomy and bilateral modified neck dissection were performed. The patient was referred for post-operative radiotherapy.

DISCUSSION

Only 3% of synovial sarcoma occur in the head and neck, with about 20 laryngeal localizationreported (5).Sex ratio is about 4/1(m/f),the majority are young adults(mean age of 37 years) (2).The supra glottis is the most frequent site with about 94,1% of the cases reported (2,6,7), this is followed by the sub glottis with 5,9%, no cases involved the glottis. Synovial cell sarcomas can present with dysphagia, cervical swelling, snoring, stridor and hoarseness of the voice.They are thought to arise from pluripotent mesenchymal cells that have the ability to differentiate into epithelial and mesenchymal lineages(8). The underlying defect in synovial cell sarcoma is primarily a specific chromosomal translocation t(X; 18) (p11, q11)(1). Histological diagnosis is difficult and immunohistochemistry is essential.There are two main subtypes monophasic and biphasic: both have proliferating spindle cells and branching, dilated, thin-walled blood vessel within a heterogeneous collagenous stroma(9). Monophasic subtypes usually contain only spindle cells. However, biphasic forms contain glandular structures in addition to all other histological features. CT and MR are non-specific in diagnosis (4) but help in planning the treatment strategy.PET-CT is useful in follow up and in detecting distant metastasis.

Figure 1: Operatory specimen: dark laryngeal tumor

Figure 2: Operatory specimen: dark well limited laryngeal tumor with an important sub-glottic extension

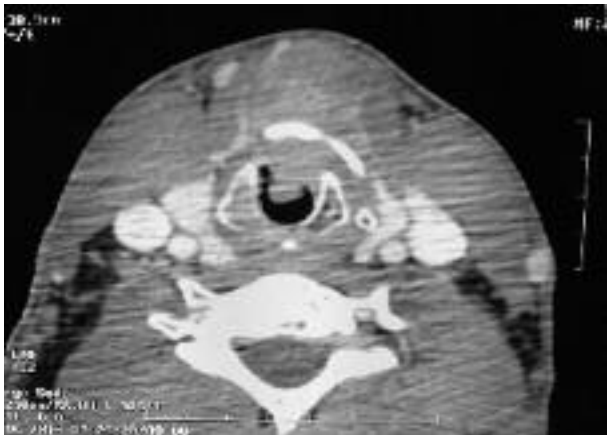

Figure 3: : Axial CT scan with contrast: soft tissue mass extending to the glottis and sub glottis destroying the thyroid cartilage

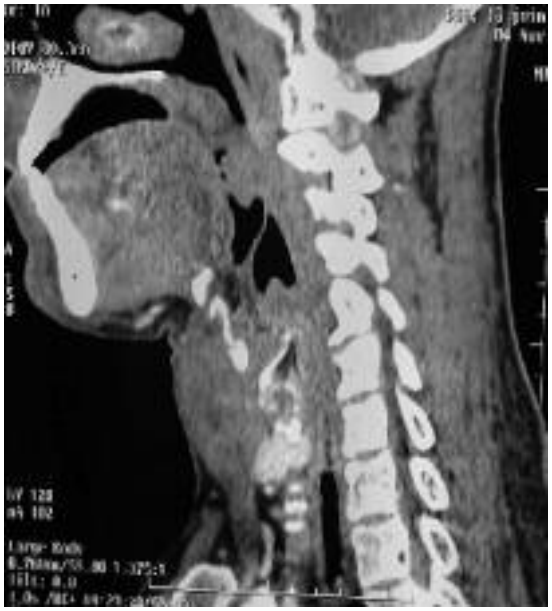

Figure 4 Sagittal CT scan with contrast: soft tissue mass destroying the thyroid cartilage with prelaryngeal space involvement

Most authors recommend that all synovial sarcomas should be treated as high-grade sarcomas(1). However, there are no established treatmentguidelines. Surgery is the main option: either partial or total laryngectomies were proposed for most of thepatients. Neck dissection is not systematic because lymphnode metastases are rare. Laser CO2 is an alternative that was associated with favorable results for the lower volume tumors. In fact, disease-free survival time was between 15 months, 2 and 3yearsin three cases operated with laser(10,11,12). Chemotherapy is controversial: for some it has no place in the treatment, others recommend it as adjuvant treatment with several agents such as ifosfamide (13, 14). Radiotherapy is reported to be of benefit in local disease management. It is administrated mainly in sarcomas greater then 5cm(1).The probability of recurrence is between 30% and 40% with 50% of distant metastasis(4) (lung for the most).The 5-year survival ranges between 23.5% and 45%(4).

CONCLUSION

In conclusion, laryngeal synovial cell sarcoma is a rare entity. Immunohistochemestry is necessary for diagnosis. Surgery is the essential treatment. It is associated with poor prognosis because of the aggressivebehavior and the common recurrence.

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