

# Management of A Paraglottic Synovialosarcoma: About A Case

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## Abstract

Cervicofacial synovialosarcoma is a malignant tumor. It poses a diagnostic and therapeutic problem. Synovialosarcoma has the strongest recurrence power among soft tissue sarcomas. These recurrences are more frequent during the first two years. Distant metastases are seen in 30% to 50% of cases and are mainly pulmonary. We report the observation of a young 38-year-old patient who was operated on in 2013 for synovialosarcoma oropharynx with radiotherapy postoperatively 6 years after the patient has two recurrences, one oropharyngeal and the other cervical.

**Keywords:** Paraglottic Synovialosarcoma Cervicofacial tumor.

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## INTRODUCTION

Synovialosarcoma is a rare malignant mesenchymal tumor, representing 5% to 10% of soft tissue sarcomas [1]. It mainly affects adolescents and young adults, with preferential limb damage [2]. Localization in the ENT sphere is very rare, representing less than 5% of tumors in this region [3] and essentially located at the level of the hypopharynx and the para-pharyngeal space. Standard treatment is based on broad and complete excision, alone or combined with post-operative external radiotherapy. The prognosis is generally poor due to local recurrences which are observed in 8% to 60% of cases [4]. Synovialosarcoma has the strongest recurrence power among soft tissue sarcomas. These recurrences are more frequent during the first two years. Distant metastases are seen in 30% to 50% of cases and are mainly pulmonary [2, 4, 5]. In this work, we report the observation of a young 38-year-old patient who was operated on in 2013 for synovialosarcoma oropharynx with radiotherapy postoperatively 6 years after the patient has two recurrences, one oropharyngeal and the other cervical.

## CASE REPORT

This is a 38-year-old patient, with no notable pathological history, who presented a year before her first consultation with a left earache associated withodynophagia. The clinical and fibroscopic examination

revealed a budding lesion in the posterior wall of the oropharynx without associated cervical adenomegaly. The extension assessment including cervical magnetic resonance imaging (MRI) objectified a heterogeneous tissue lesion process of 6 cm at the level of the posterior wall of the oropharynx, extended below the hypopharynx, with iso signal in T1 compared to muscle, and hyposignal in T2, without associated cervical lymphadenopathy. A chest CT scan as part of the remote extension checkup was normal. A monoblock resection of the tumor without lymph node dissection was performed. The anatomopathological study of the operating room has objectified a fusocellular mesenchymal proliferation whose appearance was in favor of sarcoma. The immunohistochemical complement revealed a positive labeling of tumor cells with cytokeratin, EMA, vimentin and PS 100, with an absence of labeling with AML CD34 and Desmine. The FNCLCC grade II synovialosarcoma diagnosis was accepted. The patient received adjuvant radiotherapy at a dose of 70 Gy, the patient was monitored clinically and by cervico-thoraco-abdomino-pelvic CT imaging.

6 years later, the patient has a left laterocervical mass, for which she underwent a cervicotomy under general anesthesia. The result of which was an extemporaneous study and definitive anapath came back in favor of synovialosarcoma in a second cervical localization limit's excision were healthy, for this second cervical localization no

radiotherapy. 7 months after the patient presented odynophagia with installation of dysphagia with solids of progressive installation, the clinical examination did not find a palpable cervical mass, oropharyngeal examination objectified a tissue mass hidden by the base of the tongue. The patient benefited from an injected cervical scanner which returned in favor of a process on the glottis subglottic stage in posterior extending from the first cervical vertebra to seven cervical vertebra, increasing after injection of contrast agent measuring 37 \* 39 \* 62mm without mass effect on the jugulocarotid axis (Figure 1).

The assessment completed by a direct laryngoscopy and a hypopharyngoscopy with general anesthesia which showed a tissue lesion at the expense of the left aryepiglottic fold not invading the glottic plane nor the sub glottic plane, the base of the tongue and the valleculae were healthy. Hypopharyngoscopy could not find involvement of the two piriform sinuses

or the oesophageal mouth, a biopsy made addressed to the pathology study returning in favor of synovialosarcoma.

The patient had an endoscopically excision of the whole tumor, then extemporaneous pathologic examination confirmed the diagnosis of synovialosarcoma. Then we did a cervical approach on the old incision of (2013). We did not find any tumor. So It was not a recurrence but a new localisation of paraglottic synovialosarcoma.

There were no post operative complication. The patient was referred to medical oncology for a chemotherapy according to the protocol based on DOXORUBICINE and ISOFOFAMIDE: DOXORUBICINE 75mg / m<sup>2</sup> per day for 5 days and ifosfamide 1.8mg / m<sup>2</sup> per day for 5 days, the same protocol repeated on day 21.

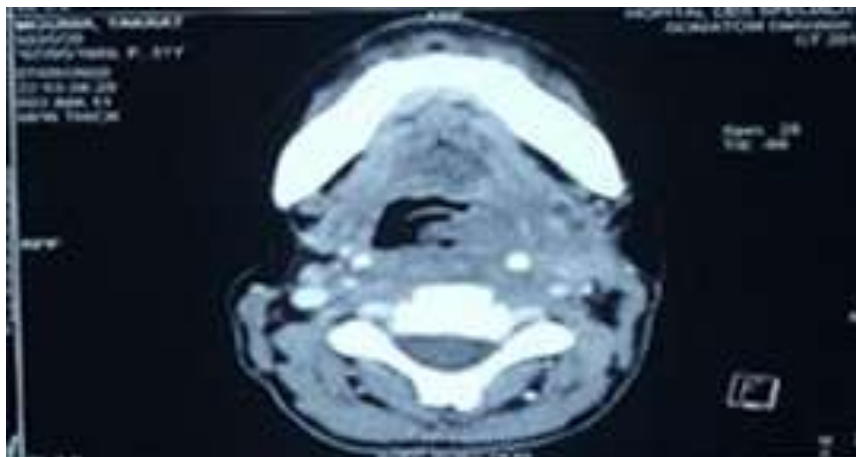


Fig-1: Axial sections of the cervico-thoracic scanner carried out in 2020 for the last oropharyngeal recurrence showed the paraglottic extension

## DISCUSSION

Synovialosarcoma is a rare, high-grade tumor of connective tissue malignancy, often occurring next to the joints. This tumor does not originate from synovial tissue, but rather from pluripotent mesenchymal cells near or even away from joint surfaces [6, 9]. Synovialosarcoma mainly affects the lower limbs. Cervico-facial localization is rare. She sees herself in less than 10% [9].

According to literature reviews, almost 100 cases have been reported [6, 9]. The most common locations are the pharynx, parapharyngeal space and the joints between the cervical vertebrae [6]. Latero-cervical localization is only reported by a few publications. Classically, synovialosarcoma is seen in young people. It occurs in 90% of cases before the age of 50 [7, 9].

Some infantile forms have been described [10]. The tumor mainly affects humans [9].

Synovialosarcoma most often presents after a lag time of 1 to 2 years in the form of a soft mass, slowly increasing in size often simulating a benign process [11].

Pain is often present and may be the only functional sign, due to the invasion or compression of adjacent structures. Signs of sensory and / or motor dysfunction are rarer. The imagery is not specific. Usually synovial sarcomas manifest as well-defined solid masses on CT or MRI scans [9-11].

Infiltration of adjacent soft tissue is a less common finding. The lesions can be homogeneous or heterogeneous, depending on the degree of hemorrhage or necrosis. The contrast enhancement is variable but it is often moderate. About 30% of synovialosarcomas contain calcifications which may be apparent on CT [4, 5]. The presence of calcifications is correlated with a good prognosis. In our case, there were no calcifications on the imagery [11].

The treatment of choice is complete excision with wide margins, but it is rarely feasible at the cervico-facial level [12]. Lymph node involvement is exceptional. Therefore, lymph node dissection is not recommended in the absence of lymphadenopathy [13]. The synovial sarcoma of the head and neck has an aggressive character and a poor prognosis. Several factors were assessed for prognosis, the most important of which were tumor size and deep extension at the time of primary treatment, with survival varying inversely with these two factors [14-16].

Most authors indicate adjuvant radiotherapy for better locoregional control. The value of chemotherapy is not yet well defined [11, 13]. Its efficacy on soft tissue sarcomas has not yet been demonstrated, but some tumors have significant sensitivities, it is usual in high grade soft tissue sarcomas of malignancy in addition to surgery. There is no standard neoadjuvant chemotherapy protocol in STM. Since the main objective in this situation is the response rate, preference should be given to multidrug therapy protocols (doxorubicin combination + ifosfamide). The optimal dose is 75mg/m<sup>2</sup>. Other molecules are also effective but to a lesser degree: dacarbazine, cyclophosphamide, cisplatin, anthracycline and epirubicin.

Many association protocols have been proposed, the cytotoxic molecules of choice are those giving the best response rate. Either protocol: doxorubicin + ifosfamide or Either protocol: doxorubicin + ifosfamide + dacarbazine [16].

The use of systematic adjuvant chemotherapy remains controversial in the treatment of adult STM, indeed the studies are contradictory. It does not represent a standard. If there is evidence that adjuvant chemotherapy improves survival without local relapse and relapse without metastasis, its impact on overall survival has not been demonstrated. This approach can be offered to selected patients at high risk (grade 2-3, > 5 cm). If adjuvant chemotherapy: use of a combination comprising doxorubicin and ifosfamide in optimal doses (4 to 6 cycles) [17-23].

For palliative chemotherapy: It is intended for metastatic patients for whom a curative program cannot be offered. The standard chemotherapies continue today to use doxorubicin and ifosfamide. The response rates obtained with doxorubicin at the 75 mg / m<sup>2</sup> dose as "monotherapy" vary from 8 to 30% depending on the study; they seem lower in the most recent studies, however most centers report a standard response rate of around 20% in current practice. Ifosfamide gives response rates of 6 to 15% with doses varying according to studies from 5 to 14 g / m<sup>2</sup>, as has also been described with doxorubicin above 60 mg / m<sup>2</sup>, a dose effect seems likely for ifosfamide, response rates

appear to be higher for doses greater than 9–10 g / m<sup>2</sup>. Large doses of ifosfamide are however burdened with severe toxicity, particularly haematological, but also digestive and sometimes renal, even neurological. If the combination of these two agents gives response rates of the order of 50% in many trials and therefore constitutes in many countries a standard treatment - with, moreover, multiple variants - there is no To date, no randomized trial showing indisputably that multidrug therapy allows a significant increase in survival with a median survival time which peaks in all trials, around 10 to 12 months.

The current trend is therefore to reserve the DOXORUBICINE – IFOSFAMIDE combination for subjects in whom an increased response rate can translate into a real benefit, for example in the case of isolated potentially resectable pulmonary metastases or in the case of symptomatic diseases for which a few additional percentages of tumor regression can change the patient's daily life. This type of combination must also be proposed with a view to primary chemotherapy, because the tumor response conditions the surgical possibilities for patients with limited operability, but neoadjuvant chemotherapy should ideally be practiced in clinical trials. Several chemotherapies have been compared to DOXORUBICINE, including other anthracyclines [16, 21].

The development of a new targeted therapy is based on the identification of a target, specific for a subgroup of tumors and involved in the phenomena of carcinogenesis and / or tumor growth. Several studies have already shown the effectiveness of certain targeted therapies in STM.

The search for agents targeted against a tumor begins by assessing the "signature" of the tumor, its uniqueness. In sarcomas with known characteristic cytogenetic abnormalities, one begins by determining the character and the translocation or fusion protein function.

Synoviosarcomas have a characteristic translocation of chromosome 18 and X, with one of the two variants of the SSX portion observed in more than 90% of cases. Suppression of the SYT-SSX protein or interference with its downstream targets could selectively inhibit the growth of synoviosarcoma cells. Unfortunately, there are unanswered questions about the biology of the fusion product, therefore targeted therapy steps remain to be studied.

## CONCLUSION

Cervicofacial synoviosarcoma is a malignant tumor. It poses diagnostic and therapeutic problems. The prognosis is generally poor due to local recurrences which are observed in 8% to 60% of cases [4]. Synoviosarcoma has the strongest recurrence power

among soft tissue sarcomas (STM). These recurrences are more frequent during the first two years. In our case, the recurrence was very rapid. Distant metastases are seen in 30% to 50% of cases and are mainly pulmonary, hence the need for close monitoring.

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