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Thoracic Monophasic Synovialosarcoma in a 17 Year Old Girl: A Case Report

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Abstract **Case Report**

Synovialosarcomas are rare, malignant mesenchymal tumors common in adolescents and young adults. They preferentially affect the lower limbs. Much more rarely, at the level of the chest wall. Contrary to its name, synovialosarcoma does not develop from synovial tissue. We report the case of a 17-year-old adolescent girl, followed for monophasic synovialosarcoma of the chest wall, the diagnosis of which was confirmed by immunohistochemistry. Treatment consisted of neoadjuvant poly chemotherapy followed by concomitant radio chemotherapy and closing surgery. This observation confirms the value of multimodal management in the treatment of this type of pathology.

Keywords: Synovialosarcoma, Thoracic, Monophasic, Immunohistochemistry, Surgery, Radio chemotherapy.

INTRODUCTION

Soft tissue sarcomas are rare, malignant mesenchymal tumors of high grade malignancy, the 4th most common soft tissue sarcoma representing 5% to 10% of cases [1]. Synovialsarcomas usually develop in adolescent and young patients near the extremities of the joints [2] and are predominantly in males [3]. They present three histological aspects: the biphasic form; the fibrous monophasic form and the poorly differentiated form (PDSS) [4]. This can be confirmed by immunohistochemistry but a cytogenetic signature is required. Detection of a specific SYT-SSX fusion transcript, a product of the t(X;18) translocation (p11.2;q11.2), confirms the diagnosis [5]. The treatment is multimodal with surgery, chemotherapy and radiotherapy.

CLINICAL OBSERVATION

We report the clinical observation of a 17-year-old female patient, with no personal or family history, who consulted for the appearance of a supraclavicular mass that had been present for about 4 months and had progressively increased in volume without any other associated signs. The clinical examination noted a good general condition and a bulging ulcerated mass above the right clavicle with pus production. The radiological workup made of CT scan (Figure 1) and magnetic resonance imaging showed a process in the right supraclavicular area. A biopsy was performed, histological and immunohistochemical examination showed a monophasic synovialosarcoma in its cystic variant at least grade II (Figure 2 and Figure 3). Polychemotherapy with adriamycin and ifosfamide resulted in a reduction in tumor mass after 6 courses. A decision of the multidisciplinary consultation meeting preferred positron emission tomography to map the lesions and then concomitant radio-chemotherapy at a dose of 50 Gy to surgery considering the sub-clavicular vasculonervous relationships. An ablation of the tumor residue was performed after radiotherapy. Local recurrence and pleural metastasis was noted after an eighteen-month period, which was treated with palliative chemotherapy.

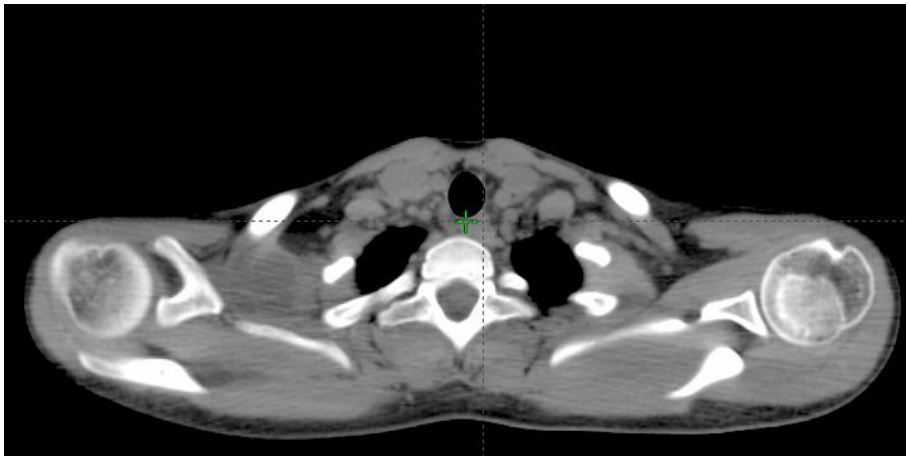

Figure 1: Transverse scanographic section showing the mass in the right upper clavicular region

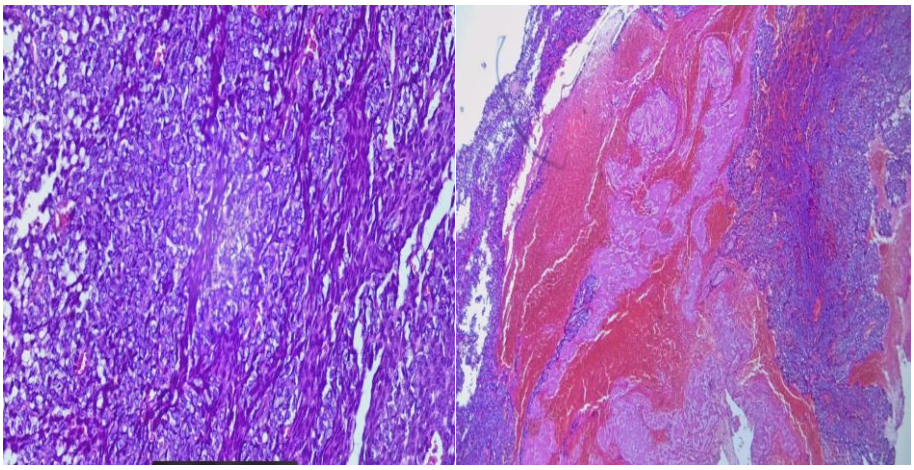

Figure 2: Histological appearance of monophasic synovialosarcoma: Monomorphic tumor cells with poorly defined cytoplasm, oval nuclei, homogeneous fine cytoplasm and richly vascularized. (Hx400 and Hx100)

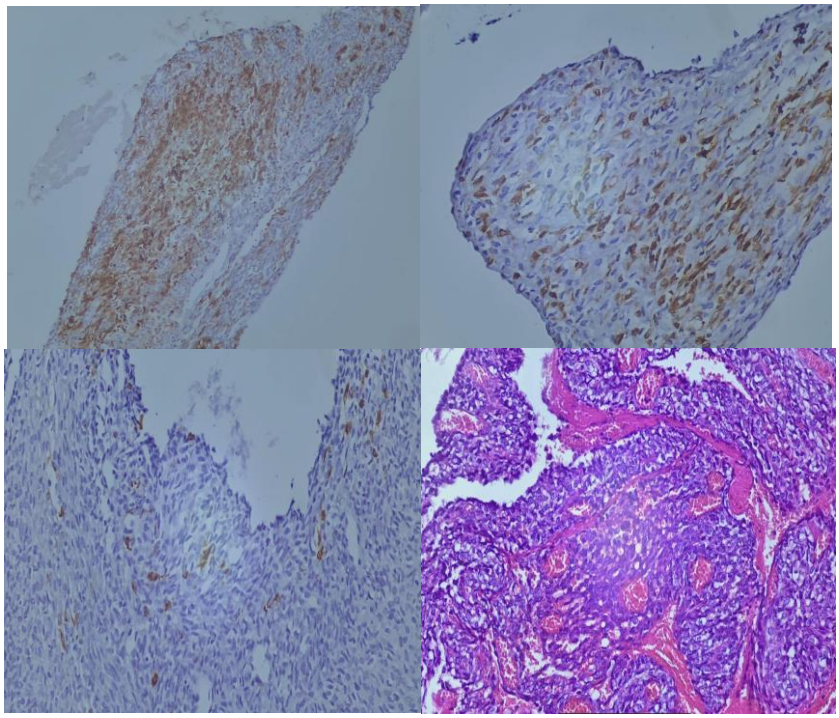

Figure 3: Histological appearance of monophasic synovialsarcoma: Positive expression of EMA, Cytokeratin 7, Cytokeratin AE1 / AE3 and hemangiopericytic vascularization

DISCUSSION

Synovialosarcoma, the fourth high-grade histological variety of sarcoma, is a rare tumor [6]. It develops from a pluripotent mesenchymal cell with synovial differentiation [7] and is mostly found in adolescents and young adults with a male predominance and a sex ratio of 2.1 [8]. Our patient is a female, 17 years old. The localizations are the periarticular regions (80% of cases) and the soft tissues (5-10% of cases) [7]. Thoracic involvement occurs in 8% of cases [9].

It presents as a soft mass, progressively increasing in size over several months or years [10], with curvature of the wall, when it develops outwardly. It may be asymptomatic [9] or responsible for chest pain, progressively increasing, as in our observation for several months [10, 11]. Radiologically, CT scans allow a better appreciation of the presence of micro-calcifications, to specify the endo- and exo-thoracic extension of the tumor [7]. On magnetic resonance imaging, approximately 90% of synovialosarcomas are well limited with a capsule-like appearance; the presence of lobulations or septa is common [12]. In 80% of cases, tumors are T2 heterogeneous with fluid, solid, or fibrous tone signals [7]. On pathological examination, the tumor is oval or rounded in shape, sometimes multi-nodular, often well delimited and encapsulated, pale, whitish or grayish in color, and with a soft consistency. Three subtypes of synovialosarcoma can be distinguished: the monophasic form (31%), the biphasic form (33%), and the undifferentiated form (36%) [7]. On immunohistochemistry, synovialosarcomas express Epithelial Membrane Antigen (EMA) and cytokeratins in 90% of cases, CD99 in 60% of cases and S100 protein in 30% of cases [13]. In our patient, the immunohistochemical study showed the negativity of S100 protein, cytokeratin, vimentin and EMA positivity. By identifying by Fish or RT-PCR the presence of a specific fusion transcript SYT-SSX, resulting from the translocation t(X;18) (p11.2; q11.2), the cytogenetic analysis of the tumor cells confirmed the diagnosis of synovialosarcoma [5].

Several prognostic factors have been suggested which are mainly: age, location, tumor size, high mitotic index and Ki-67 greater than 10% [14, 15]. Among these, in our patient, we find size, age and thoracic location. Treatment is multimodal combining surgery and radio-chemotherapy [7]. The combination of chemotherapy with radiotherapy is not standard, it potentiates the response of radiotherapy to allow resection of the mass [7]. The recurrence rate of locoregional or metastatic disease at two years is 50%. These recurrences are more frequent during the first two years and affect more regional lymph nodes, lung, bone and liver (16). The prognosis is generally poor due to local and metastatic recurrence [17].

CONCLUSION

Synovialosarcoma of the chest wall, a rare malignancy; its treatment is multimodal based on extensive, complete surgery and neoadjuvant or adjuvant chemotherapy and external radiotherapy. The prognosis is poor due to frequent local and metastatic recurrences. Our observation highlights the interest of multidisciplinary collaboration in the treatment of this tumor.

Conflict of Interest: The authors declared no conflict of interest.

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