North American Spine Society Journal (NASSJ) 15 (2023) 100243

Contents lists available at ScienceDirect

North American Spine Society Journal (NASSJ)

journal homepage: www.elsevier.com/locate/xnsj

Clinical Case Studies

Multidisciplinary management of recurrent synovial sarcoma of the chest wall

Santiago A. Endara, MD[a], Juan S. De la Torre, MD[b], Fernando J. Terán, MD[c], Juan Pablo Alarcón, MD[d,*], Carla E. Tovar, MD[e]

[a] Cardiovascular and Thoracic surgeon, Hospital Metropolitano, Quito, Ecuador
[b] Spine Surgeon, Hospital Metropolitano, Quito, Ecuador
[c] Oncologist surgeon, Hospital Metropolitano. Quito, Ecuador
[d] Orthopedic surgery senior resident, Universidad Internacional del Ecuador, Quito, Ecuador
[e] General Surgery resident, Universidad Internacional del Ecuador, Quito, Ecuador

ARTICLE INFO

Keywords:
Synovial sarcoma
Multidisciplinary management
Local recurrence
Adjuvant therapies

ABSTRACT

Background: Synovial sarcoma (SS) is part of soft tissue sarcomas (STS). An incidence between 5% to 10% is estimated. The origin is mesenchymal mainly affecting the extremities. Being even rarer at the chest level and vertebral body, representing around 1%. Histologically, it consists of 3 variants: monophasic, biphasic, and poorly differentiated. Surgical resection is a priority when it comes to multidisciplinary management. The prognosis of patients with SS over the years has improved markedly.

Purpose: Understand and evaluate the multidisciplinary management of SS considering that the SS has a lowe prevalence and highly malignancy.

Study Design: We present a case of a 31-year-old male who has a history of monophasic synovial sarcoma diagnosed in 2019 and underwent surgery. Patient came back after two years without symptoms and posterior to a control MRI we observed a local recurrence of SS.

Methods: The literature was reviewed with a focus on best clinical and surgical strategy for recurrence of SS.

Results: The patient recovered well with return to his normal daily activities. The review of the literature shows us the importance of the multidisciplinary management for the optimal clinical and surgical approach of SS recurrence.

Conclusions: SS represents a unique variant of STS, with malignant and metastatic potential. Being a rare pathology, an adequate multidisciplinary management is essential when providing optimal care for the patient.

Introduction

Synovial sarcoma (SS) is part of the soft tissue sarcomas (STS) [1]. The literature on its incidence is somewhat scarce due to few reported cases and the lack of follow-up. An incidence between 5% to 10% of all STS is estimated. Differing from these by its relative appearance at the early age (between 15 and 35 years) [2] at diagnosis and clinical presentation [3]. The name is somewhat ambiguous because the first reported cases were located around the distal femur and proximal tibia, mistakenly believing that its origin was synovial [4]. Currently it has been determined that its origin is mesenchymal, mainly affecting the extremities with a similar affectation in both sexes [5].

Histologically, it consists of 3 variants: monophasic, biphasic and poorly differentiated. The monophasic variant consists of spindle cells while the biphasic variant consists of spindle and epithelial cells [6]. Pathologically, SS is characterized by a translocation t(x:18)(p11.2q11.2) which is present in 95% of cases [7]. Different genetic fusions have been investigated, the main one being SYT-SSX1, but none of its known variants correlate with prognosis [8].

The clinical presentation of SS differs from the typical STS; the growth is slow, and the symptoms appear approximately 2 years after its appearance [9]. The clinical and surgical management of SS must be multidisciplinary and individualized for each patient. Considering that the complete excision of the tumor with negative margins is the main

FDA device/drug status: Not applicable.

Author disclosures: ***SE:*** Nothing to disclose. ***JDLT:*** Nothing to disclose. ***JPA:*** Nothing to disclose. ***CT:*** Nothing to disclose. ***DG:*** Nothing to disclose.

* Corresponding author. Department of Orthopedics and Traumatology, Hospital Metropolitano, Av. Mariana de Jesús s/n y Nicolas Arteta, Quito, 170521, Ecuador.
E-mail address: juan_pa1089@hotmail.com (J.P. Alarcón).

https://doi.org/10.1016/j.xnsj.2023.100243
Received 13 April 2023; Received in revised form 14 June 2023; Accepted 5 July 2023
Available online 21 July 2023

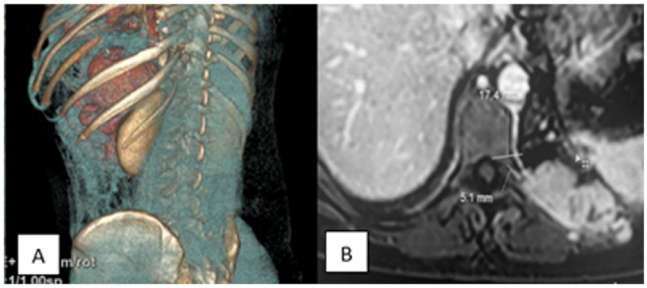

Fig. 1. (A). Tumor mass at the level of the left posterior chest wall, with involvement of the pleura and adjacent diaphragm. (B). Extension of the tumor mass towards the T9 vertebra left transverse process.

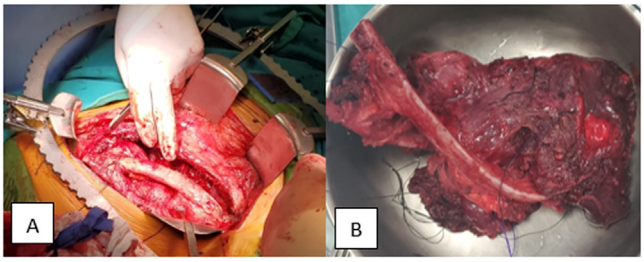

Fig. 2. (A). Left thoracotomy. (B). En bloc resection of Left posterior chest wall tumor mass.

priority in the treatment followed by chemotherapy and adjuvant therapy [10].

Case Report

A 31-year-old male patient presented to a private hospital in Ecuador with history of localized pain on the chest wall and sensation of dyspnea after several studies (referred by the patient) he was diagnosed with monophasic synovial sarcoma in 2019 and went to surgery by thoracotomy, decortication, partial resection of the left ninth rib, and fragmented tumor resection; performed by another group of surgeons in another hospital. Adjuvant chemotherapy was given for 7 cycles (last therapy on July 2020) with ifosfamide and adriamycin. After that, the patient abandoned treatment and medical follow-up. He went back to consultation in June 2022 denying symptoms. Based on sarcoma management guidelines a body CT scan and MRI were performed, in which a tumor mass with bilobed morphology measuring 3.7×3.0×5.3 cm (Fig. 1A) with involvement of the adjacent pleura was reported at the level of the left posterior chest wall with extension to the diaphragm, lung and erosion of the posterior ninth costal arch and left transverse process (Fig. 1B) of the T9 vertebral body. No foraminal or spinal canal involvement was appreciated. Taking into account the age of the patient, the localized progression of the tumor mass; surgical resolution was decided by multidisciplinary management with cardiothoracic, oncology and spine surgery through: left thoracotomy (Fig. 2A), en bloc resection of the chest wall tumor (Fig. 2B), with diaphragm resection, left lung lobectomy and posterior reconstruction of the chest wall and diaphragm (Cardiothoracic surgeon); by anterior approach, a left T8-T9 hemilaminectomy was performed + spinal cord decompression + block pedicle osteotomy T8-T9 + ligation of emerging nerve roots, after which chest wall reconstruction was performed. The patient was repositioned to perform arthrodesis and posterior instrumentation T6-T12, bilateral foraminectomy T6-T12 + facetectomies T6-T12. (Spine surgeon) (Fig. 3A and B).

Patient was transferred to the intensive care unit for pain management and hemodynamic control. After an adequate response to pain and physiotherapy, discharge was decided by the service to subsequently perform adjuvant chemotherapy (Oncologist) and controls by outpatient department. The histopathological result of the tumor mass reported a

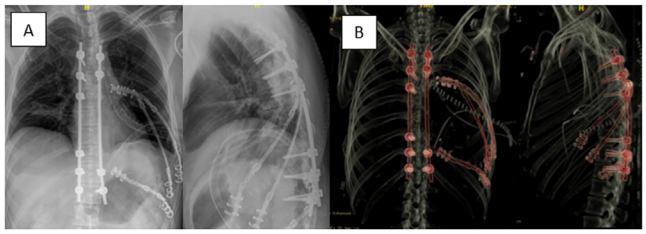

Fig. 3. (A and B). Arthrodesis and posterior instrumentation T6-T12 with open transpedicular screw fixation with the objective of provide immediate stability of the thoracic spine, prevents further risk of complications and allows early mobilization of patient.

recurrent monophasic synovial sarcoma without lymphovascular invasion with tumor-free borders at the bone, soft tissue, muscle, lung, and diaphragm levels. (Stage IB AJCC staging system). Eleven months after the tumor resection, the patient complained of mild intensity left intercostal pain and dyspnea with moderate activity. The patient has remained under regular check-ups since the post-surgical period, no local recurrences or metastatic spread have been detected. He is currently receiving adjuvant chemotherapy. The patient presents a Karnosfky scale of 80 points, that is, he can carry out normal activities, with mild symptoms of the disease. In addition, he presents an ECOG 1 scale, that is, he presents symptoms that prevent him from performing hard work, although he performs normally in his daily activities and in light work.

Discussion

Synovial sarcoma is a rare malignant neoplasm that represents around 5% of STS [11]; It is mainly located in the extremities, being even rarer a primary level at chest level and vertebral body, representing around 1% [12]. In order to differentiate it from other types of STS, it is imperative to perform a biopsy guided by tomography or magnetic resonance imaging. Immunohistochemistry in 90% of cases expresses the epithelial membrane antigen (EMA) and Bcl2, 60% CD99 and 30% SM100 [13].

The first surgical procedures carried out in the context of the SS were salvage surgeries, that is, carrying out gross amputations; by which very low recurrence rates were obtained but sacrificing the functionality of the affected limb. With the advent of new surgical techniques and adjuvant methods, the so-called focused surgery was later proclaimed, but the recurrence rate was not taken into account, which is why the currently objective is to perform a wide excision of the tumor with negative margins in order to reduce the risk of local recurrence rate, procedure performed on our patient.

Taking into account the age of our patient, the absence of symptoms and the local recurrence evidenced in the complementary studies; It was decided to perform complete resection of the tumor mass through left thoracotomy with resection of 3 ribs, diaphragm, lung, left transverse process of T9 vertebral body, diaphragmatic and chest wall reconstruction, left hemilaminectomy T8–T9 and resection of the vertebral pedicle in order to obtain negative tumor borders, it was necessary to perform a facetectomy of the affected segment as the entry point to the pedicle area, leaving the affected segments unstable, for this reason it was necessary to perform a posterior vertebral instrumentation with the purpose of avoiding a progression to hyper kyphosis, compression of the nerve roots of the intervened segments or spinal cord injury.

Current use of postoperative chemotherapy has positive results, which is reserved for tumors larger than 6 cm. At present, there is no indication for the initiation of chemotherapy in tumors smaller than 5 cm, our patient is receiving chemotherapy [14]. Different studies have shown that adjuvant radiotherapy decreases local recurrence rates [15].

Synovial sarcoma has high rates of recurrence and metastases, mainly to the lung. Zhang et al, [16] has determined that the number of

recurrences is a risk factor for local recurrences and distant infiltration, which influences the patient's survival time. Our patient required lung resection due to infiltration.

The prognosis of patients with SS over the years has improved remarkably, reporting survival rates between 59% and 75% at 5 years due to a better availability of tools for diagnosis and early treatment [17,18]. Kyriazoglou et al, [19] in a retrospective study report that the average survival rate is 96.7 months at diagnosis with a disease-free survival of 26 months. It should be emphasized that there are key prognostic factors such as: tumor size (less than 5 cm), anatomical location, staging (less than Grade 3), age (less than 65 years), negative surgical margins, chemotherapy and adjuvant radiotherapy that should be taken into account when discussing the prognosis [20]. Thus, an unplanned surgical excision, advanced age, neurovascular invasion, mitoses per field greater than 10/mm2 have been related to a worse prognosis [21]. A review by Xiong et al, [22] determined that the biphasic variant has the highest survival rates at 5 and 10 years of follow-up . Krieg et al. demonstrated that local recurrence occurred after a mean of 3.6 years (range 0.5–15 years) and metastases occurred at a mean of 5.7 years (range 0.5–16.3 years).

Among the most important prognostic factors (age, initial tumor size, surgical resection method, histological subtype, metastasis) especial attention it must given to the local recurrence (LR). We mainly emphasize LR, taking into account that it is part of our case. It has recently been reported that LR may affect the prognosis of patients with STS. But there are limited studies on this topic. Berg et al, [26] reports that the mortality risk per year with a local recurrence of SS is 3.6. Zhang et al, [16] report that more than half of the patients with a history of local recurrence experience another episode (HR 2148 in the first year after surgery) and 82% of the patients with multiple recurrences develop distant metastases. For this reason, constant monitoring of this type of patients is mandatory to act in a timely manner.

Multidisciplinary management is the most appropriate clinical and surgical strategy when dealing with this pathology; with the main objective of achieving disease-free survival and reducing rates of local recurrence and metastasis [23]. Pang et al, report a diagnostic accuracy rate of STS without a multidisciplinary team of 90.84% while with a multidisciplinary team diagnostic accuracy power increase to 95.42%. The pathological tissue subtypes are diverse and have overlapping histological features. The advantage of the multidisciplinary team (MDT) discussions in improving diagnostic accuracy is evident. For certain difficult cases, through preoperative MDT discussions, radiologists and pathologists can communicate with orthopedic surgeons and advise the most suitable site for biopsy, which may reduce the probability of invalid specimens and improve the quality of specimens. The accuracy of relapse diagnosis by the MDT reached 100%, and the MDT improved the accuracy of a recurrence diagnosis STS. Reported rates of residual sarcoma in the re-resected specimen after unplanned resections are uniformly high, ranging from 24% to 74% [24]. In contrast, the residual rate of re-excision after MDT discussion was 81.2%, higher than that reported in the literature [25]. These data may indicate to a certain extent that multidisciplinary management plays an active role when making decisions in the context of adequate treatment of recurrent SS.

There are no supported studies that refer to the best treatment in terms of the subtypes of synovial sarcoma. Therefore, we fully abide by international driving guidelines. Considering that an optimal surgical approach and a complete tumor resection with negative free margins are the primary objectives to achieve the highest possible survival rate knowing that we are dealing with a very malignant tumor class.

After 11 months of continuous follow-up periods, the patient has not presented alarming clinical symptoms that lead us to a progression of his disease. Functionally active ECOG 1. Complementary imaging studies have not revealed local recurrence or distant metastasis. He is currently functionally active without limitations on his daily life. Taking into account that the 5-year survival rate varies between 60% to 70% in adulthood, periodic evaluations of the patient will be necessary to detect in the shortest possible time the possibility of local recurrences or distant metastases because they are highly malignant tumors.

In Ecuador, unfortunately, there is no database on the management of this pathology due to different factors: socioeconomic problems, inadequate infrastructure, lack of specialists, ignorance or simply the omission of working together. It is the first report of a case of recurrent synovial sarcoma in the chest wall successfully managed in our country based on current management guidelines and with special emphasis on multidisciplinary management.

Conclusion

Synovial sarcoma represents a unique variant of STS, with malignant and metastatic potential. Being a rare pathology, an adequate multidisciplinary management is essential when providing optimal care for the patient. To date, the spearhead of treatment is the resection of the tumor mass with negative margins, with a prior guided biopsy being a priority for its diagnosis. Monophasic SS is the most prevalent and to date the one with the best prognosis and recurrence rate at 5 years is the biphasic lineage. It is imperative to carry out serial follow-ups by outpatient department in order to detect in time local recurrences or metastatic extension, mainly to the lung. Radiotherapy and chemotherapy both are necessary in high-risk patients with SS. The combination of surgery and radiotherapy remains the cornerstone of local control meanwhile adjuvant chemotherapy is necessary to prevent metastatic extension. The prognosis depends on quality management by an experienced multidisciplinary team.

Declarations of competing interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] Gazendam AM, et al. Synovial sarcoma: A clinical review. Curr Oncol 2021;28(3):1909–20. doi:10.3390/curroncol28030177.
[2] Faur CI, et al. Synovial sarcoma of the extremities: A literature review. Appl Sci 2021;11(16):7407. doi:10.3390/app11167407.
[3] WHO. Classification of tumours editorialSoft Tissue and Bone Tumours. 5th ed. Lyon, France: IARC Press; 2020. ISBN 978-92- 832-4502-5.
[4] Sistla R, Tameem A, Vidyasagar JV. Intra articular synovial sarcoma. Indian J Pathol Microbiol 2010;53:115–16.
[5] Aytekin MN, Öztürk R, Amer K, Yapar A. Epidemiology, incidence, and survival of synovial sarcoma subtypes: SEER database analysis. J Orthop Surg 2020;28:2309499020936009.
[6] Jo VY, Fletcher CDM. WHO Classification of soft tissue tumours: An update based on the 2013 (4th) edition. Pathology 2014;46:95–104.
[7] Stegmaier S, Leuschner I, Poremba C, et al. The prognostic impact of SYT-SSX fusion type and histological grade in pediatric patients with synovial sarcoma treated according to the CWS (cooperative weichteilsarkom studie) trials. Pediatr Blood Cancer 2017;64:89–95.
[8] Sun Y, Sun B, Wang J, et al. Prognostic implication of SYT-SSX fusion type and clinicopathological parameters for tumor-related death, recurrence, and metastasis in synovial sarcoma. Cancer Sci 2009;100(6):1018–25.
[9] Chotel F, Unnithan A, Chandrasekar CR, Parot R, Jeys L, Grimer RJ. Variability in the presentation of synovial sarcoma in children. J Bone Jt Surg Br Vol 2008;90:1090–6.
[10] Von Mehren M, Randall RL, Benjamin RS, et al. Soft tissue sarcoma, version 2.2016, NCCN clinical practice guidelines in oncology. J Natl Compr Cancer Netw 2016;14:758–86.
[11] Thway K, Fisher C. Synovial sarcoma: defining features and diagnostic evolution. Ann Diagnost Pathol 2014;18(6):369–80.
[12] Shah UB, Joshi S, Ghorpade SV, Gaikwad SN, Sundrani RM. Primary pleuropulmonary synovial sarcoma. Indian J Chest Dis Allied Sci 2010;52:16972.
[13] Lino-Silva LS, Flores-Gutiérrez JP, VilchesCisneros N, Domínguez-Malagón HR. TLE1 est exprimé dans la majorité des sarcomes synoviaux pleuropulmonaires primaires. Virchows Arch 2011;459(6):615–21.
[14] Nakamura T, Saito Y, Tsuchiya K, et al. Is perioperative chemotherapy recommended in childhood and adolescent patients with synovial sarcoma? A systematic review. Jpn J Clin Oncol. 2021;51:927–31.
[15] Song S, Park J, Kim HJ, et al. Effects of adjuvant radiotherapy in patients with synovial sarcoma. Am J Clin Oncol 2017;40:306–11.
[16] Zhang H, Huang W, Feng Q, Sun W, Wang C, Zhang J, Huang K, Yu L, Qu X, Chen Y. Clinical significance and risk factors of local recurrence in synovial

sarcoma: A retrospective analysis of 171 cases. Front Surg 2022;8:736146. doi:10.3389/fsurg.2021.736146.
[17] Aytekin MN, Öztürk R, Amer K, Yapar A. Epidemiology, incidence, and survival of synovial sarcoma subtypes: SEER database analysis. J. Orthop. Surg. 2020;28:2309499020936009.
[18] Smolle MA, Parry M, Jeys L, Abudu S, Grimer R. Synovial sarcoma: Do children do better? Eur. J. Surg. Oncol. 2019;45:254–60.
[19] Kyriazoglou A, Timmermans I, De Cock L, et al. Management of synovial sarcoma in a tertiary referral center: A retrospective analysis of 134 patients. Oncol Res Treat 2021;44(5):232–41. doi:10.1159/000515112.
[20] Ferguson PC, Griffin AM, O'Sullivan B, et al. Bone invasion in extremity soft-tissue sarcoma: Impact on disease outcomes. Cancer 2006;106:2692–700.
[21] Lewis JJ, Antonescu CR, Leung DH, et al. Synovial sarcoma: A multivariate analysis of prognostic factors in 112 patients with primary localized tumors of the extremity. J. Clin. Oncol. 2000;18:2087–94.
[22] Xiong L, Chen Z, Zhou Y, Li H, Xiao T. The survival and prognosis analysis of synovial sarcoma subtypes: A surveillance, epidemiology, and end results population-based analysis. Int. Orthop. 2020;44:2779–86.
[23] He Huayu, Yang L, Peng Y, et al. The value of Multidisciplinary Team (MDT) management in the diagnosis and treatment of primary intrathoracic synovial sarcomas: A single-center experience. J Thor Dis 2021;13(2):600–12. doi:10.21037/jtd-20-2887.
[24] Venkatesan M, Richards CJ, McCulloch TA, Perks AG, Raurell A, Ashford RU. East Midlands Sarcoma, S. Inadvertent surgical resection of soft tissue sarcomas. Eur. J. Surg. Oncol. 2012;38:346–51.
[25] Pang Ke, Guo x, Liu T, et al. The role of a multidisciplinary team in the diagnosis and treatment of bone and soft tissue sarcomas: A single-center experience. J Pers Med 2022;12(12):2079. doi:10.3390/jpm12122079.
[26] Bergh P, Meis-Kindblom JM, Gherlinzoni F, et al. Synovial sarcoma: Identification of low and high risk groups. Cancer 1999;85:2596–607. doi:10.1002/(SICI)1097-0142(19990615)85:123.0.CO,2-K.