

Malignant Solitary Fibrous Tumor: Case Report

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Abstract

Soft tissue sarcomas (Sts) are rare tumors that make up about 1% of malignant neoplasms. In addition to their histological diversity, Sts can occur anywhere on the body but present predominance in end areas. We aim to describe a case of soft tissue sarcoma because it is a rare malignant neoplasm. We report the case of a male patient, 60 years old, with a previous history of lung adenocarcinoma that started with diffuse abdominal pain with progressive worsening requiring surgical approach presenting histopathological findings compatible with malignant solitary fibrous tumor. He underwent adjuvant chemotherapy but evolved with disease progression. Another surgical approach was performed but due to the extension of the disease, complete resection was not possible and local IMRT radiotherapy was then chosen. The patient has been in clinical follow-up since 2015 considering that the disease has been presenting a slight reduction in its size in the imaging studies.

Introduction

Soft tissue sarcomas (Sts) make up a heterogeneous group of malignant neoplasms with different morphological patterns of the mesenchymal lineage, representing about 1% of malignant neoplasms in adults [1]. Sts may occur in any part of the body, but most often originate in extremities (59%), trunk (19%), retroperitoneum (15%), or head and neck (9%) [2]. Moreover, there are more than 50 histological types of Sts, among the most common are Liposarcoma being the most frequent (20-30%) [3], Undifferentiated Pleomorphic Sarcoma (28%), Leiomyosarcoma (12%) and Synovial Sarcoma (10%) [4].

With regard to their epidemiology, they are rare tumors that appear to have no association with geographical and racial factors [5]. What can be observed is an increase in annual incidence according to age, being 20.7% in patients under 40 years of age and 27.6% in patients between 40 and 60 years of age and 51.7% in patients with over 60 years [6]. In the United States, the Annual Global incidence rate in USA is 6x1000 inhabitants¹⁰, the number of cases per year is averaging 8,0002, while in Brazil there are around 3,500 new cases per year. The low incidence leads to a late diagnosis and to inappropriate behaviors, as a consequence there are lower rates of global survival, and, often, worsening of quality of life [5].

It appears that soft tissue sarcomas do not appear to be the result of differentiation of benign soft tissue tumors, and despite the histological variety, Sts have similar clinical and pathological development [2]. Regarding retroperitoneal Sts, its location has direct implications in the diagnosis and treatment of sarcomas. Retroperitoneal soft tissue sarcomas are often asymptomatic or have nonspecific symptoms and are only diagnosed when they reach large proportions, and their relationship with adjacent organs often leads to the need for surgical resections (treatment of choice) that are often invasive [4,7].

Case Report

Patient ORC, 60 years old, with previous history of sublobar resection due to adenocarcinoma of the lung (pT1a N0), started a diffuse abdominal pain with progressive worsening, which after investigation with imaging was diagnosed in September 2010 with pelvic expansive lesion, submitted to total surgical resection R0, (CSF7 positive, CD34 positive, Bcl 2 positive, CD99 positive focally, Ki 67 ranging from 10 to 15%, focal positive actin, CD56 positive, CK7 negative, negative AE1 / AE3 negative, S100 negative, desmin negative, inhibin negative, calretin negative, c-kit negative) together with the histopathological findings compatible with malignant solitary fibrous tumor.

It evolved with rapid recurrence, with unresectable tumor, being submitted from December 2010 to January 2011 to chemotherapy with three drugs for 2 cycles (probable protocol Ifosfamide, mesna and epirubicin), evolving in February 2011 with sudden dyspnea being diagnosed with important mitral insufficiency, left ventricle and decreased ejection fraction to 59%, and submitted

to mitral valvuloplasty. He remained in clinical follow-up until December 2011. At that time he performed a new pelvic resonance with persistence of expansive formation in the right pelvic region measuring 7.3 x 5.4 x 5.0 cm.

In January 2012 a new course of chemotherapy with carboplatin + paclitaxel was performed for four cycles every three weeks, with no effective response.

It evolved with progression of disease with invasion of prostatic store and seminal vesicles on the right being submitted to a new surgical approach in May 2012, surgicalcalitor reduction, with ureteral lesion and need for ureter reconstruction, need for nephrostomy, double J placement and hospitalization in Therapy Unit Intensive by sepsis, and it is not possible to complete tumor resection due to adherence in the sacral region. Patient lost cancer follow-up for 2 years.

When in November 2014 through new imaging tests, pelvic resonance showed increased lesion with the presence of massive solid expansive lesion with areas of internal cystic degeneration located in the topography of the seminal vesicles, notably on the right but invading the prostate store, measuring 12.5 x 12.0 x 9.0 cm with heterogeneous signal in T1 and T2 weights and intense and heterogeneous enhancement by gadolinium, without pelvic or inguinal lymph node enlargement.

In this patient, neoadjuvant chemotherapy was instituted for 3 cycles of epirubicin and ifosfamide, after rapid recurrence, but the patient presented cardiotoxicity due to the treatment and after an interval without treatment and disease progression, carboplatin and taxol were prescribed in 4 cycles, but the same did not respond. A surgical approach was proposed after this chemotherapy, due to no effective response, and it was not possible to complete resection of the tumor, and after a new analysis of histopathological and immunohistochemical material, determining the present histological type of Malignant Solitary Fibrous Tumor.

Due to previous major mitral insufficiency and chronic atrial fibrillation, a new surgical approach was contraindicated and only IMRT radiotherapy was performed for 30 fractions from February to March 2015.

Initiated clinical follow-up at our institution since May 2015 and was chosen for clinical observation since the disease has been presenting a stable disease in its dimensions seen in serial MRI of the abdomen.

Discussion

The patient in question initially presented, in 2010, a pelvic retroperitoneal sarcoma of histological type hemangiopericytoma, according to the immunohistochemistry (IHQ) presented to us. This type of tumor, first described in 1942 by Stout and Murray, accounts for 5% of all Sts [1,5]. It is a vascular neoplasm, originated in the pericytes (Zimmerman cells), present in the whole body, wrapped in the arterioles, capillaries and venules, they exert a contractile action on these vessels. Tumors of this cellular type have a predilection for the musculoskeletal system, often found in the lower extremities and trunk, mainly retroperitoneal region [7,8]. It does not have higher prevalence between the sexes and can affect any age, however it is more frequent between the third and sixth decade of life [8]. They

present insidious growth and the symptomatology is due to the compression of adjacent organs and structures [7]. Its diagnosis is through histopathological study and the tumors can be classified into benign, borderline and malignant (> 4 x 10 campos of mitosis) [9], that is, according to the histological characteristics such as mitotic activity, nuclear atypia and cellularity, however, it is very difficult this distinction between benign and malignant [10,11]. The treatment of choice is complete surgical resection of the tumor, radiotherapy is indicated after surgery to reduce the risk of local recurrence and chemotherapy is advised in cases of metastasis, in some cases partial remission may be done [7].

Although most TFS show indolent behavior, with very low risk of recurrence or metastasis, some tumors behave aggressively. Although commonly described as predominantly intrathoracic tumors, approximately 50 to 70 percent are located outside the thorax. Radiographic findings in the transverse image (Computed Tomography [CT], magnetic resonance imaging [MRI]) are characteristic but not pathognomonic. Contrast enhanced computed tomography (CT) [12] will demonstrate a well-circumscribed, often lobulated, hypervascular tumor often with areas of necrosis especially when large T2-weighted Magnetic Resonance Imaging (MRI) typically reveal a well-defined mass that is markedly in homogeneous with large areas of bright signaling reflecting extensive areas of necrosis [12].

Conclusion

Solitary fibrous tumor (TFS) comprises a histological specimen of rare soft tissue neoplasms demonstrating fibroblastic differentiation including hemangiopericytoma. Are variably cellular and composed of cells with oval to spindle-shaped nuclei with minimal cytoplasm and intervening collagen bands arranged in pattern less distribution with are as highly rich in tumor cells while other areas are more hypocellular with higher percentage of stromal collagen most TFS behave indolently and do not resort locally or remotely. A number of risk stratification models have been devised in attempts to more accurately predict the behavior of TFS based on a combination of clinical, pathological and anatomical criteria. Management of TFS should be discussed in a multidisciplinary tumor advice with sarcoma specialists who have experience with the disease. Postoperative radiotherapy (adjuvant) (RT) and chemotherapy were not well studied in TFS. However, given the lack of data supporting the benefit and favorable outcome of surgery in most cases, we suggest that no adjuvant RT or chemotherapy is sought for the majority of patients with completely resected TFS. The use of adjuvant RT or adjuvant / neoadjuvant chemotherapy for incomplete or recurrent resection of TFS is best decided case by case, in the context of a multidisciplinary discussion.

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