Case Report

Pancreatic metastasis of mesenchymal chondrosarcoma

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ABSTRACT

Mesenchymal chondrosarcoma is an aggressive tumor with chondrogenic differentiation that typically develops in skeletal sites, such as craniofacial bones, ribs, ilium, femur, and the vertebrae. It counts less than 3% of primary chondrosarcomas, and about a third of these tumors develop extraskeletal sites such as the meninges, and soft tissue. We present a case of a 53-year-old female, that was diagnosed with mesenchymal chondrosarcoma in the left thigh and submitted to surgical excision. The patient's case was evaluated by a multidisciplinary team, and it was decided to maintain vigilance. In the 5 years, follow-up presented several pulmonary metastases that were submitted to pulmonary resections. Two years later, during imaging control, an abdominal computed tomography (CT) scan was performed and revealed a pancreatic mass of 3 centimeters, located in the pancreatic tail. The patient was submitted to an explorative laparoscopy and a distal pancreatectomy was performed. The biopsy gave the definitive result of the metastasis of mesenchymal chondrosarcoma.

Keywords: Mesenchymal chondrosarcoma, Pancreas, Metastasis

INTRODUCTION

Mesenchymal chondrosarcoma (MC) is an aggressive, rare tumor with chondrogenic differentiation, first described in 1959 by Lightenstein and Bernstein. The peak incidence is in the second and third decade of life, and equally, affects men and women. It counts less than 3% of primary chondrosarcomas, that typically develop in skeletal sites, most commonly from craniofacial bones, ribs, ileum, femur, and vertebrae. A small percentage of these tumors develop extraskeletal sites such as the meninges, and soft tissue.1 There are some case reports of intra-abdominal metastasis.

Clinically, patients with MC suffer from pains due to the space-occupying effect caused by the growing tumor.

The classic histologic finding of MCs is a biphasic tumor composed of sheets and irregular islands of hyaline cartilage or branching thin-walled vasculature, whereas the cartilage component contains hypertrophic chondrocytes with distinct nuclei but without pleomorphism.2 However, despite the protracted clinical course, it is characterized by developing metastases and local recurrence at a high rate and finally high mortality.3

We describe a clinical case of female suffering from MC who developed a single pancreatic metastasis 7 years after the initial diagnosis.

CASE REPORT

In February 2010, a 53 years-old female was treated in our hospital for a lower limb mass. The patient was submitted to surgery and histopathological results confirmed mesenchymal chondrosarcoma.

The patient's case was evaluated by an oncologic multidisciplinary team, and it was decided to maintain vigilance.
At 5 years follow-up, imaging showed 3 lung metastases, with 0.2, 0.3, and 0.4 cm each. The patient was evaluated in Cardiothoracic Surgery, to remove these lung metastases.

Despite these treatments, abdominal computed tomography (CT) imaging, at 7-year follow-up, demonstrated a pancreatic mass of 3 centimeters, and the patient was referenced to our service. Laparoscopic distal pancreatectomy with spleen preservation was performed. The histopathology of the excised specimen proved to be the one of metastatic lesions of MC. The pancreatic resection margin was not involved by tumor, and metastases were not identified in peripancreatic lymph nodes. Microscopically, the tumor was composed of sheets of small, round to oval cells with stippled chromatin, scant cytoplasm, and “hemangiopericytoma-like” vasculature surrounding scattered islands of well-differentiated hyaline cartilage.

The post interventional course was prolonged due to pancreatic fistulae, that we managed conservatively. The patient left the hospital after 15 days of treatment.

In 2019, other lesions of metastatic origin in both lungs were found and the patient was submitted to thoracotomy and 3 lesions were excised. The patient remains asymptomatic and to date with no signs of apparent recurrence.

**DISCUSSION**

MC is a rare primitive appearing tumor with cartilaginous differentiation that predominantly develops in skeletal sites. Despite these tumors tend to affect patients in the second to third decade of life and 10-year overall survival rates are lower (27 to 44%), our case showed another pique of incidence and longer survival rate. This tumor entity is rare and randomized clinical trials examining the effect of chemotherapy in metastatic and non-metastatic mesenchymal chondrosarcoma are lacking. Generally, the therapy consists of radical surgery and probably chemotherapy. However, a clear treatment strategy has not been defined.

Despite the metastatic disease, these patients have a possible survival for years, so surgical treatment is recommended. The prognosis is poor because new metastasis may occur and the patient already was submitted to a lot of surgeries. In the literature, metastases are an independent predictor of death in a 44% 10-year-overall survival rate, in extraskeletal MC.

Some cases reports were published concerning abdominal metastasis from Mesenchymal chondrosarcoma in the kidney, uterus, and a unique case in the spleen. Relatively to the pancreas, 4% of the pancreatic masses are secondary tumors, and metastases of MC is relatively rare.

**CONCLUSION**

When detecting a mass of the pancreas in patients with a medical history of MC, a metastasis of this tumor entity should be taken into consideration. Further development of novel agents for treatment is essential for improving the prognosis of this type of tumor.

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**REFERENCES**