PAROSTEAL OSTEOSARCOMA WITH MYOCARDIAL METASTASIS 13 YEARS AFTER FOLLOW-UP


PURPOSE: To report the case of a woman with a diagnosis of grade II (low grade) parosteal osteosarcoma with the occurrence of myocardial metastasis 13 years after resection, and to present a review of the existing literature on the subject.

METHODS: Description of the case and review of the literature.

CONCLUSION: The review leads to the conclusion that the occurrence of metastasis from parosteal osteosarcoma can occur in up to 38% of the cases, in spite of its relatively low aggressiveness. However, myocardial metastasis of a parosteal osteosarcoma is an event that was not found in the literature.


INTRODUCTION

Parosteal osteosarcoma (POS) is an uncommon, slow-growing variant of osteosarcoma (OS) that occurs on the surface of the bone, and its diagnosis is obtained using specific criteria. Its evolution is relatively benign, it is usually treated by wide resection with good results, and it often does not require chemotherapy.

The metastatic form of the disease may occur in up to 38% of the cases a few months or many years after the initial treatment. The low-grade type of POS seldom metastasizes; on the other hand, the dedifferentiated variant has high potential for distant metastasis, with the lung being the most common site. However, 14 cases of patients with cardiac OS metastases whose primary tumors were of the conventional variety in all cases are reported in the literature. This report describes a case of POS in a female patient who was initially treated by wide resection and developed late myocardial metastasis, and it also presents a review of the literature with regard to several aspects related to the occurrence of cardiac metastases in patients with OS.

DESCRIPTION OF THE CASE

A 26-year-old, white, female patient presented a slow-growing tumor in the right arm over a period of 5 years. The tumor was resected at another service, with no available information on surgical pathology (SP) findings.

Six months after the resection, there was a relapse of the lesion, at which time the patient sought our service.

After a simple x-ray that showed characteristics suggestive of POS (Fig. 1), the patient was biopsied, with the corresponding SP findings confirming a grade II POS (Fig. 2). The patient underwent wide resection of the proximal humerus and implantation of a non-conventional polyethylene endoprosthesis (Fig. 3). Chemotherapy was not indicated since it was a low-grade lesion. Thirteen years later at the annual follow-up exam, imaging exams revealed thoracic nodules, one of which was located in the cardiac topography (Figs. 4 and 5). When the thoracotomy was performed, 2 pulmonary nodules
were resected, at which time a 6 cm nodule was also discovered in the myocardial region in the atrioventricular septum next to the marginal and diagonal arteries, which was considered unresectable due to its location.

The SP examination of the lesions yielded grade II POS material, with characteristics similar to those of the primary tumor (Fig. 6).

The authors chose not to administer adjuvant treatment and decided on a strict follow-up protocol, with simple x-rays and thorax tomography at 3- and 6-month intervals, respectively (Fig. 7).

The patient is faring well 3 years after the surgery without additional symptoms or signs of tumor growth (Fig. 7).

DISCUSSION

POS is a malignant tumor of the bone tissue that was first described by Greschckter & Copeland in 1951, with an incidence corresponding to 1% of all bone tumors and 4% of OS. The World Health Organization de-
FINES GROWTH ON EXTERNAL SURFACES, MEDULLAR INVASION OF UP TO 25%, HIGH GRADE OF DIFFERENTIATION, AND LOW GRADE OF MALIGNANCY AS ITS DIAGNOSTIC CRITERIA. THE HIGHEST INCIDENCE OF POS OCCURS IN THE THIRD AND FOURTH DECADES OF LIFE PROMINENTLY IN WOMEN, AND THE INCREASE OF LOCAL VOLUME AND PAIN ARE ITS CLINICAL MANIFESTATIONS. THE DIAGNOSIS IS SUGGESTED BY CLINICAL AND RADIOGRAPHIC FINDINGS AND CONFIRMED BY BIOPSY AND SP FINDINGS. HISTOPATHOLOGY SHOWS THE PRESENCE OF 3 COMPONENTS, CONSISTING OF FIBROUS, OSSEOUS, AND CARTILAGINOUS TISSUES, WITH 3 GRADES OF HISTOLOGICAL DIFFERENTIATION. GRADES I AND II ARE CHARACTERIZED AS LOW GRADE, ALSO CALLED CONVENTIONAL POS. HOWEVER, POS MIGHT HAVE A HIGH-GRADE COMPONENT (GRADE III). IN SUCH CASES, THE TERM DEDIFFERENTIATED HAS BEEN USED. THERE CAN BE DEDIFFERENTIATION OF A TUMOR FROM LOW TO HIGH GRADE; THIS IS A COMPLEX PROCESS THAT COULD BE RELATED TO SEVERAL LOCAL RELAPSES.

Most cases manifest themselves at the time of diagnosis as tumors of local growth, and occurrences of initial metastatic disease are rare, which demonstrates the insidious development of this type of tumor.

Treatment of POS is based upon the specific stage of the tumor and on whether it is intracompartmental or extracompartmental (Iα or Iβ). In intracompartmental tumors (Iα), in which cortical and medullar invasion of the bone tissue does not occur, a transparietal resection can be performed, with better functional results and low rates of local relapse, as long as strict controls of the resection margins are observed. In extracompartmental cases (Iβ), it is necessary to use wide resections, followed by reconstitution with homologous grafts or endoprostheses. Amputation is rarely indicated, and it is restricted to the cases of large volume with several relapses.

Later evolution to metastatic disease can occur in up to 38% of the patients, and metastasis is generally associated with cases of grade III POS, medullar invasion, and several local relapses. The occurrence of metastases with conventional tumors (low grade) is less frequent, and it is restricted to less than 20% of the cases. The main site of metastases is the lung, with the abdomen and the central nervous system being reported less frequently.

The period of latency between the primary tumor and the occurrence of metastases can vary from a few months to many years; survival following metastasis varies from 6 to 32 months, with an average of 16 months.

The presence of cardiac metastases is relatively rare, occurring in 3% to 10% of the cases, of which just 10% will present clinical or radiological manifestations that may suggest the diagnosis. The main histological types of the cardiac metastases are...
Parosteal osteosarcoma with myocardial metastasis
Santos-Machado TM et al.

purposes, is the production of bone tissue, which can be radiographically demonstrated as a circumscribed lesion with osseous density in the projection of the cardiac region. Trans-esophageal echocardiography confirms the lesion, permitting visualization of the extent of involvement as well as any eventual secondary hemodynamic repercussions. Final diagnosis is provided by the surgical pathology exam.

To the best of our knowledge, the occurrence of cardiac metastases from POS is not described in the literature. In addition, POS is a low-grade tumor with a low probability of causing systemic disease, which is one of the reasons for diagnosis after such extended periods of latency. Campanacci reports 9 occurrences of metastases in 41 cases of POS (21.9%), 4 of which were discovered after latency periods of over 10 years (11.5-23 years). Of those cases, 3 were grade II, 2 presented involvement of the medullar channel, and all of them had undergone at least 2 manipulations.

The previously described characteristics of low aggressiveness also account for the favorable prognosis for this patient, even without resection, which is considered by thoracic surgeons to be of high morbidity potential, and without adjuvant treatment. One option would be heart transplantation, but that was not indicated in our case, since there was no evidence of symptosis; additionally, having to keep the patient under immunosuppression adds additional risk of morbidity. The choice in this case was the follow-up based on clinical and radiographic parameters.

No evidence of cardiac or systemic symptoms has been found up to the time of the last evaluation 4 years later, confirming the low aggressiveness of the lesion.
RESUMO


OBJETIVOS: Os autores relatam um caso de uma paciente do sexo feminino, com diagnóstico de osteossarcoma parostal grau II (baixo grau), que evoluiu com a ocorrência de metástase miocárdica 13 anos após a ressecção do tumor e incluem uma revisão da literatura sobre o assunto.

MÉTODOS: Descrição do caso e revisão da literatura.

CONCLUSÕES: A revisão leva à conclusão que a existência de metástases oriundas do osteossarcoma parosteal pode ocorrer em até 38% dos casos, a despeito da sua relativa baixa agressividade. No entanto, a metástase miocárdica constitui um evento não relatado na literatura.


REFERENCES


Parosteal osteosarcoma with myocardial metastasis
Santos-Machado TM et al.


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