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Case report

Radiotherapy for granulocytic sarcoma of the breast—Case report and review of the literature

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ABSTRACT

Granulocytic sarcomas are rare tumors that can present in innumerous locations; thus there is very little clinical experience with these cases. Therefore there is no consensus on which is the best treatment for patients with this malignancy.

The authors present a case of a female with a granulocytic sarcoma of the breast and review the literature for the role of radiotherapy in the management of this clinical entity.

1. Introduction

Granulocytic sarcomas (GS) are rare manifestations of acute myeloid leukemia (AML). They consist of immature myeloid cells that proliferate producing a clinically evident tumor.[1,2] These normally occur associated with AML, nevertheless very rarely they might present as isolated malignancies.[1,3]

Isolated GS of the breast (i.e. without medullary evidence of leukemia) are somewhat rare, and frequently associated with a dismal prognosis prompting treatment optimization.[3–5]

In the following section a case of an isolated GS of the breast is presented.

2. Case report

Thirty-five years old female with no relevant medical history, on auto-examination of the breasts found a tender nodule on the left breast.

Physical examination revealed a 25 mm nodule on the upper external quadrant of the left breast, without any other relevant findings.

The patient was submitted to an ultrasonography and a mammography. These showed a solid heterogeneous nodule, measuring 19.5 mm × 22 mm × 21 mm on the upper external quadrant of the left breast and a small lymph node on the ipsilateral axilla.

A tru-cut biopsy of this lesion was performed. Pathological analysis was inconclusive; however GS and myofibroblastoma were referred as the most likely diagnoses; lymphoma, carcinoma and melanoma were excluded.

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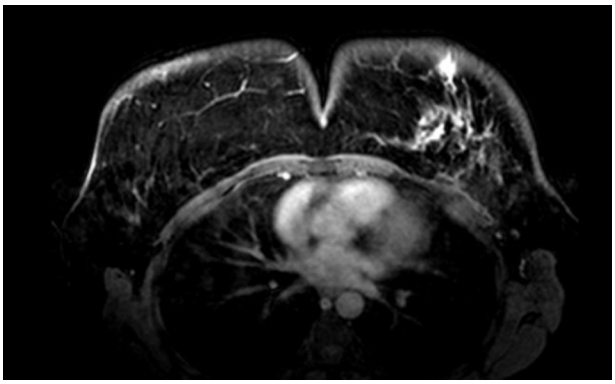

Fig. 1 – Post induction chemotherapy MRI.

Therefore the patient underwent tumorectomy. The final histologic diagnosis was GS with the following immunophenotype: myeloperoxidase+ CD45+, CD68+, CD34+, CD31+, CD43+, Cam 5.2−, CD117−, CD3−, CD20−, TdT−, CD10−.

As part of the workup a bone marrow study was conducted, it revealed a normocellular marrow with no excess of immature cells, some atypia and signs of dysmyelopoiesis.

The patient underwent chemotherapy treatment: induction with idarrubicin and cytarabine, followed by consolidation with 4 cycles of cytarabine. There were complications during the consolidation phase: febrile neutropenia, pulmonary aspergillosis, pyelonephritis and severe enterocolitis (with need for parenteral nutrition).

A post induction chemotherapy magnetic resonance (MRI) showed persistent disease (Fig. 1)

Another MRI, performed after completion of the entire chemotherapy regime, revealed a heterogeneous enhancement with approximately 60 mm (Fig. 2)

On a follow-up MRI, 3 months post chemotherapy, there was no evidence of disease.

After resolution of the adverse reactions, the patient underwent radiotherapy (RT). Radiation was delivered to the left breast with a 3D conformal (multi-leaf collimator) technique: 30 Gy in 2 Gy fractions (1 fraction per day, 5 days per week), with 6 MV photons, in a total of 21 days (Fig. 3). There were no interruptions of the treatment or any severe reactions. By the end of the treatment the patient had grade 2 dermatitis of the breast skin (CTCAE v4.03).

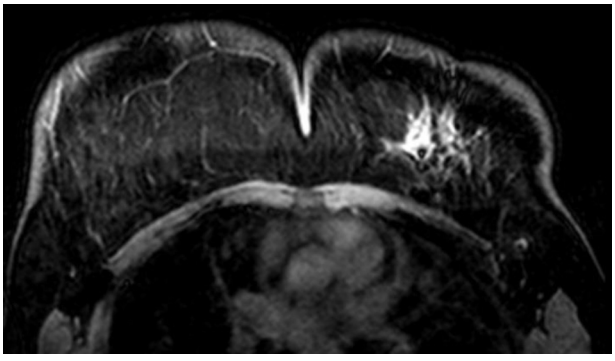

Fig. 2 – Post chemotherapy MRI.

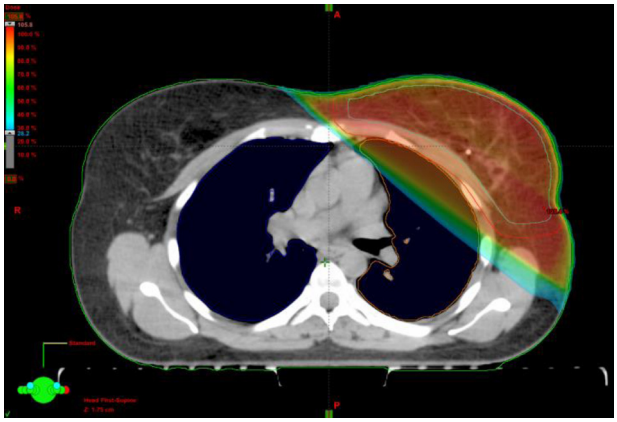

Fig. 3 – RT dose distribution (color wash).

At 26 months follow-up the patient remains asymptomatic and there is no evidence of relapse of the disease.

3. Discussion

Granulocytic sarcomas, or chloromas, are rare solid tumors, composed of immature myeloid cells, which occur in extramedullary locations.[1,2] This malignancy was first described by Allan Burns in 1811.[6] It was not until 1853 that the term chloroma was coined by King A.[7]; it derives from the Greek *chloros*, meaning green, due to a green tint that myeloperoxidase gives these cells. However, the presence, concentration, and oxidative status of this enzyme vary among tumors, giving cells different colors, 30% are white, grey or brown. Therefore the term Granulocytic Sarcoma, introduced by Rappaport in 1966[8] is nowadays the most used one. This initially referred only to tumors of granulocytic origin, as GS most frequently develop in patients with AML.[1,3] This association was firstly noted by Dock and Warthin in 1902.[9] Nevertheless, it can also be associated with chronic myeloid leukemia, myelodysplastic syndrome, chronic idiopathic myelofibrosis, hypereosinophilic syndrome and polycythemia vera hence the current broader use of the term granulocytic sarcoma.[1,3,4,10,11] These tumors present more frequently concomitantly or after the diagnosis of AML, there are, however, some cases where GS antedates AML.[1–5,10–13]

Some have argued that there is a slight male predominance, however many series fail to show any difference in gender.[1–4,10,11] Age of incidence ranges from 2 to 81 years (mean 33–48 years) depending on the series.[1–3] In a large autopsy series published by Liu et al. that comprised 338 cases of myeloid leukemia (both acute and chronic) of which 23 had GS, age at onset was the only factor with statistical significance, with younger patients having a higher risk of developing GS (under 15 years-old 13.6%; 15–44 y 6.7%; 45–59 y 4.4%; 60 y 0%).[11]

Granulocytic sarcomas can virtually occur anywhere in the body, however it more frequently arises in the skin, subperiosteal bone (cranium, paranasal sinuses, sternum, ribs, and vertebrae), soft tissues of the head and neck, orbit, central nervous system and lymph nodes.[1–4,10,11,13,14] The breast