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Skin-nipple-sparing mastectomy: The first approach in primary myxoid chondrosarcoma of the breast



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ABSTRACT

The primary mammary chondrosarcoma corresponds to less than 0,5% of the mammary malignancies. For the period ranging from 1967 to 2014, only 18 cases were reported in the literature.

A 41 year old woman found a hard nodule on her external right superior quadrant/axillary prolongation through breast self-examination. The vacuum-assisted core biopsy (VACB) revealed "high grade extra-skeletal myxoid chondrosarcoma". A skin-nipple-sparing mastectomy with the insertion of a mammary expander was performed. A protocol of adjuvant radiotherapy was also indicated.

Until 2013, the gold standard was the radical mastectomy. By 2014, there were two cases of conservative approach to quadrantectomy. To our knowledge, this represents the first case in the literature in which a skin-nipple-sparing mastectomy has been performed on a primitive mesenchymal neoplasm of the breast. Such an oncoplastic approach was performed considering the young age of the woman, to assure the surgical radicality and a better quality of life to the patient.

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1. Introduction

Breast sarcomas are a highly heterogeneous group of tumours [1], most of which are malignant fibrous histiocytoma, fibrosarcoma, liposarcoma and, less commonly, angiosarcoma, rhabdomyosarcoma, dermatofibrosarcoma, osteosarcoma, chondrosarcoma, desmoid tumours and other very rare malignancies.

Pure primary chondrosarcoma is one of the extremely rare types of sarcomas of the breast, corresponding to less than 0,5% of all breast malignant tumours. The surgical approach is the biggest concern. Here, we report a case that recently occurred to our institution with the aim of discussing the therapeutic strategies that we adopted.

2. Case report

A 41 year old woman came to our university hospital after having noticed a painless lump by palpating her right breast. Physical examination revealed a hard lump with irregular margins, fixed at the right external superior quadrant/axillary prolongation and measuring about 2,5 cm. Ultrasonography showed a hypoechoic lump, with a polylobated shape and, on defined hyperechoic mar-

gins, inside of which we noticed intralesional vascular areas on colour-Doppler test, that measured 2,8 × 1,7 cm. We also noticed a bilateral axillary lymphadenopathy. Because of the suspicious ultrasonographic features of the lump and of the patient's MRI, which underlined "in the axillary prolongation a massive lump with annular enhancement, whose curve of signal/time intensity showed rapid wash-in and following plateau", we suggested the patient to undergo a core-biopsy. The biopsy report confirmed the features of suspicion already detected.

Consequently, we performed a vacuum-assisted core biopsy (VACB) of the lump. The histological examination revealed large areas of necrosis, a great number of atypical round or spindled small cells, sometimes joined in small groups, with a high mitotic division rate, in a background consisting of amorphous basophilic, myxoid or vaguely chondroid substance. An immunohistochemical study showed a strong and widespread positivity for vimentin; only focal positivity for pankeratin, and negativity for CK7, CK5/6, S100, ER, PR, EMA, and Her2. Histology revealed a mesenchymal-type neoplasm and in particular an "extra-skeletal myxoid chondrosarcoma", likely in "high degree" variant cells.

We consulted an oncologist for an opinion. Considering the absence of metastasis (verified on body CT and on bone scintigraphy), he recommended the patient to undergo a surgical operation. The patient was subjected to a skin-nipple-sparing mastectomy with the insertion of a breast expander also after having gone through the technique of sentinel lymph node and extemporane-

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Fig. 1. The whole breast was removed by exploiting the diamond-shaped incision on the skin projection of the tumour.



Fig. 2. A submuscular case for the insertion of the breast expander (300 cc) was created. The submuscular case was then closed through the creation of a flap, the serratus anterior muscle was sutured with the inferior-lateral margin of the pectoralis major muscle. The expander was filled with 150 cc saline solution. It was not necessary to carry out a counter-lateral mastopexy considering the symmetry of the breasts.

ous histological examination of back-nipple and of the margins of the resected lesion: all of which were negative.

In particular, with regard to the surgical technique, the whole breast was removed by exploiting the diamond-shaped incision on the skin projection of the tumour (Fig. 1). A submuscular case for the insertion of the breast expander (300 cc) was created (Fig. 2). The submuscular case was then closed through the creation of a flap (the serratus anterior muscle was sutured with the inferior-lateral margin of the pectoralis major muscle). The expander was filled with 150 cc saline solution. It was not necessary to carry out a counter-lateral mastopexy considering the symmetry of the breasts.

The specimen, consisting of the mammary gland (15 × 9 × 5,5 cm), was partially covered with diamond skin (8 × 3,5 cm) and included a whitish compact neoplasm with



Fig. 3. During the follow-up in the 4th, 8th, and 12th month after the operation, the patient seemed to be free from local relapse, metastasis, and postoperative complications and showed also a very good final aesthetic result.



Fig. 4. One year after the operation, the expander was removed, and, an anatomic (400 g) mammary prosthesis was inserted on the right breast, and mastopexy was performed on the left breast.

polycyclic margins and large areas of necrosis next to the axillary prolongation (3 × 2,5 × 2,5 cm). The final histological examination confirmed that the diagnosis was already formulated by core-biopsy: extra-skeletal myxoid chondrosarcoma. No neoplastic infiltration was found in the lymph nodes and the resection margins were R=0.

In a multidisciplinary internal meeting, radiotherapy was recommended, but, not chemotherapy, and it was recommended to perform a tight oncological follow-up for the first five years, after which the risk of recurrence or metastasis should reset. It was decided, with the consent of the patient, to administer five cycles of standard radiotherapy (50 Gy). During the follow-up in the 4th (Fig. 3), 8th, and 12th month after the operation, the patient seemed to be free from local relapse, metastasis, and postoperative complications and showed also a very good final aesthetic result. One year after the operation, the expander was removed, and, an anatomic (400 g) mammary prosthesis was inserted on the right breast, and mastopexy was performed on the left breast (Fig. 4).

3. Discussion

Primary mammary sarcomas originate from a supporting interlobular mammary stroma; they don't arise directly in the chest wall from the underlying bone or cartilage, as instead the secondary breast sarcomas do, involving the breast secondarily (for example the angiosarcoma of the breast which could develop as a consequence of breast irradiation) [1].

In particular, it seems to be very important to consider mammary sarcomas as entities separated from the more common breast carcinomas and from mixed tumours, such as metaplastic carcinoma and malignant phyllodes tumour, with whom they come in differential diagnosis. These tumours show a similar clinical presentation, but they deeply differ in their biological behaviour, requiring thus a different approach [2]. Both the metaplastic carcinoma [3] and the malignant phyllodes tumour [4] show a mixed histological pattern with both epithelial and mesenchymal elements, therefore, an extensive sampling of the tumour should be made in order to reveal a right diagnosis [5]: pure chondrosarcoma is microscopically characterized by chondroid lacunas in which numerous chondroblasts exhibit cellular atypia [6], and it is also characterized by the absence of epithelial areas. Only 18 cases of primary breast chondrosarcoma have been reported in the literature between 1967 and 2014. Until 2013 the gold standard was a radical mastectomy [7], and conservative breast surgery was never part of the management of the 16 reported cases; in 2014, two cases of quadrantectomy were reported in the literature [8,9], but the benefits of radical mastectomy compared with a more conservative surgery have not been mentioned. Mastectomy was associated with lymphadenectomy in 4 cases, associated with grafting in one case [2]; it was preceded by neoadjuvant chemotherapy (with partial response) in one case [6] and was followed by adjuvant radiotherapy in three cases. Moreover, in two cases, this rare kind of tumour occurred to male [10,11] patients and, in one case, it was focally infiltrated into the surrounding skeletal-muscle [2].

Surgery remains the mainstay of treatment for most sarcomatoid tumours. Despite the possibility that adjuvant therapy may decrease local and systemic recurrence rates of somatic sarcomas, the literature is lacking in significant information regarding the benefits of chemo or radiotherapy in mammary sarcomas, because of the rarity of this disease and of the small number of cases reported [2,6]. The present case report describes a new case of primary mammary chondrosarcoma: by analysing the nature of the neoplasm, its dimensions, its suprafascial localisation, the necessity of wide margins of resection, the dimension of the breast, and the young age of the patient, and considering the absence of extra-lymph nodal and lymph nodal metastasis, it was decided to perform a skin-nipple-sparing mastectomy with the insertion of a mammary expander, rather than a definitive prosthesis, to allow the patient to undergo adjuvant radiotherapy. This surgical approach and the adjuvant radiotherapy were chosen with the consideration that the rare histological "myxoid chondrosarcoma" pattern of the tumour makes it chemoresistant [8], and that hormone therapy has no rule in the treatment of pure mammary sarcomas. This fact is supported by immunohistochemical studies: the mesenchymal elements are positive both for S-100 and vimentin, but negative for common epithelial markers, such as cytokeratins and oestrogen and progesterone receptors [7,11].

Another aspect of the management of this case is the execution of sentinel lymph node technique [12,13]: studies have pointed out that sarcomas do not have tendency to spread via lymph node; consequently, the axillary dissection that would undergo patients to further pains seems to be useless. However, since it is important to remember that these neoplasms enter in differential diagnosis with metaplastic carcinoma and malignant phyllodes tumour [14,15], we have preferred to perform sentinel lymph node biopsy

in a preventive line in the eventuality of an incorrect temporary histological diagnosis (VACB): if this had happened, we would have had to operate again the patient.

The present treatment strategy that we chose is in line with the last two cases reported in the literature, which opt for a more conservative approach, since radical mastectomy does not offer a higher efficacy.

4. Conclusion

We extol the present case, because it is the first time in which a skin-nipple-sparing mastectomy was performed on a primary mesenchymal breast tumour, a very rare example of tumour if we consider its nature and location. This oncoplastic approach seems feasible, safe, and reliable, considering the young age of the patient. It assures her not only the surgical radicality, but also a better quality of life. All the work has been reported in line with the SCARE criteria [16].

Conflicts of interest

There were no conflicts of interest, sources of financial support, corporate involvement, patent holdings, etc. involved in the research and preparation of this case report.

Sources of funding

There were any source of funding, sponsors for collection, analysis and interpretation of data; in the writing of the manuscript; and in the decision to submit the manuscript for publication.

Ethical approval

For our case report the scientific ethics committee approval was required, the consent of the patient expressing his consent to our medical records.

Consent

All the case reports was written keeping the anonymity of the patient and not by entering sensitive information, but in our medical records has been given consent for privacy.

Author contributions

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