Primary leiomyosarcoma of the nipple-areola complex with an unusual clinical presentation: A case report

Xavier Guedes, Meritxell Medarde, Tamara Parra, MJ Martinez, Enric de Caralt, Constanti Serra

ABSTRACT

Introduction: Primary leiomyosarcoma of the nipple is a rare disease that accounts for less than 0.1% of all malignant tumors of the breast. About 35 cases have been described in English literature.

Case Report: We report a case of a 46-year-old female presenting with a two-year suppurative squamous plaque-type lesion of the right nipple at first considered as an unusual Paget’s disease presentation. No adenopathies or associated lesions were identified. Core biopsy was informed as a possible leiomyosarcoma. It was decided to remove the lesion surgically by wide excision of the nipple-areola complex with adequate surgical margins with no lymph node dissection or adjuvant radiochemotherapy. Histopathology and immunohistochemistry confirmed the diagnosis of leiomyosarcoma. One year after the surgery there are no recurrences or distant metastasis.

Conclusion: More data and long-term follow-up is necessary to understand prognosis and develop an adequate treatment protocol for this type of neoplasms.
Primary leiomyosarcoma of the nipple-areola complex with an unusual clinical presentation: A case report

Xavier Guedes, Meritxell Medarde, Tamara Parra, MJ Martinez, Enric de Caralt, Constanti Serra

ABSTRACT

Introduction: Primary leiomyosarcoma of the nipple is a rare disease that accounts for less than 0.1% of all malignant tumors of the breast. About 35 cases have been described in English literature. Case Report: We report a case of a 46-year-old female presenting with a two-year suppurative squamous plaque-type lesion of the right nipple at first considered as an unusual Paget’s disease presentation. No adenopathies or associated lesions were identified. Core biopsy was informed as a possible leiomyosarcoma. It was decided to remove the lesion surgically by wide excision of the nipple-areola complex with adequate surgical margins with no lymph node dissection or adjuvant radiochemotherapy. Histopathology and immunohistochemistry confirmed the diagnosis of leiomyosarcoma. One year after the surgery there are no recurrences or distant metastasis. Conclusion: More data and long-term follow-up is necessary to understand prognosis and develop an adequate treatment protocol for this type of neoplasms.

INTRODUCTION

Since it was first described in 1968 by Waterworth [1], primary leiomyosarcoma of the breast has been discussed in about 35 reported cases in English literature. This entity is rare considering that it accounts for 5–10% of breast sarcomas, that only account for 0.1% of all malignant tumors of the breast [2]. From the reported cases, less than a third are located on the nipple-areola complex [3]. We describe the unusual clinical presentation, diagnosis and treatment of a 46-year-old female with a leiomyosarcoma of the nipple-areola complex and a review of the existing literature.

CASE REPORT

A 46-year-old female came to our clinic presenting with an induration of the right nipple after more than two years experiencing several episodes of suppuration of a punctate lesion in the left margin of the right nipple. The past medical history of the patient was without pathological findings and had no family history of malignancies in any first-degree relatives. She had a routine mammography performed four years before with no pathological findings. Physical examination only
revealed induration of the right nipple with increased sensitivity and a squamous plaque-type lesion (Figure 1). No axillary, supraclavicular or cervical lymph nodes were palpable. Ultrasonography did not find any suspicious lesions and assessed a BIRADS 1 category (Breast Imaging-Reporting and Data System of the breast 1: no images suggest breast malignant pathologies). A preoperative core biopsy of the indurated lesion was performed, and was described by the pathologist as a smooth muscle neoplasm with atypia that suggested a leiomyosarcoma with affected margins. Immunohistochemistry revealed actin (Dako- Clon 1A4, Denmark) and desmin (Dako-Clon D33, Denmark) positivity. The patient underwent a second surgical procedure for a wide excision of the nipple-areola complex with 4–5 cm safe margins (breast parenchyma) and without axillary lymph node dissection (Figure 2). Macroscopically, the tumor measured 1.6x1 cm located on the left margin of the nipple with superficial pale desquamative white patches. Histological examination revealed a leiomyosarcoma of the nipple, 1.6 cm in diameter, with hyper cellular areas composed of round cells, bundles of spindle cells with focal pleomorphic cells and 3 mitosis per 10 high-power fields (HPF) without necrosis and no affected margins (Figures 3–5). Immunohistochemistry confirmed actin and desmin positivity and the cell proliferation marker Ki-67 (Dako-Clon MIB-1, Denmark) showed an index of 15.

After surgery, a computed tomography (CT) scan of the chest and abdomen were performed discardiing distant metastasis. After one year, the patient is in good health with no evidence of local recurrence.

**DISCUSSION**

We report a case of a primary leiomyosarcoma of the nipple which only accounts for less than 0.1% of all malignant neoplasms of the breast. Most patients are female, though there are at least four reported cases...
of male subjects with primary leiomyosarcoma of the breast [4]. It usually appears in postmenopausal women between the ages of fifty to eighty years and in some rare cases it can appear in young girls [5]. Most cases describe a painless slow-growing nodular mass. In our case the patient described a punctate lesion that underwent several episodes of suppuration that gradually turned into a plaque-type lesion. No reports of this type of clinical presentation were found on previous literature. At first it was thought to be a rare case of Paget’s disease because of the similarity to the eczema that appears on the nipple on this disease.

The importance of describing these rare cases is to avoid the risk of not including this neoplasm in the differential diagnosis of neoplasms of the breast, specially sharing similarities with a wide range of benign and malignant neoplasms such as leiomyomas, peripheral nerve sheath tumors, malignant phyllodes tumors, malignant fibrous histiocytoma, dermatofibrosarcomas, spindle cell malignant melanoma and other sarcomatoid carcinomas [6]. Mammography and ultrasonography can often lead to misdiagnosis of these tumors because of the difficulty in differentiating them from benign lesions such as fibroadenomas, phyllodes tumors or intracystic papillomas. Even fine-needle aspiration cytology (FNAC), core-needle or surgical biopsies are not enough to make a definitive diagnosis of leiomyosarcoma. A complete excision of the tumor, immunohistochemistry, and detailed histologic examination is needed for correct diagnosis [3]. Most cases are not diagnosed preoperatively for these reasons.

Microscopically, the tumors are composed of atypical and hyperchromatic spindle-shaped cells arranged in interdigitating fascicles and the cytological characteristics are hyperchromasia in the nuclei, pleomorphism, and mitosis [7]. The key to discern leiomyomas from leiomyosarcomas is the presence of mitosis, and in previous literature the mitosis of these tumors ranged from 2 to 21 per 10 HPF [8, 9]. Immunohistochemical profile of smooth muscle actin, vimentin and desmin positivity can be helpful but not pathognomonic of the tumor [5]. There is still controversy on the origin of this tumor. In the nipple-areolar complex it might originate from the smooth muscle bundles that surround the lactiferous ducts and the arrector pili muscle at the periphery of the areola. Outside this region it is believed to originate from the smooth muscle cells of the vascular walls in the mammary parenchyma [3, 6].

As for any unusual or infrequent type of tumor, there is no clear consensus on the best treatment modality. Since 1968, it has been treated surgically either by simple excision, modified radical or radical mastectomy. Some cases included axillary dissection and others preferred the use of adjuvant radiochemotherapy. Though there were cases that reported metastasis or local recurrences, most authors agree that there is not enough evidence that supports the idea of lymphatic spread and nodal metastasis as features associated with these neoplasms, similarly to other breast sarcomas [10–12]. Usually, tumors of the nipple-areola region are treated as other skin sarcomas, with wide local excision leaving 3 to 5 cm safe margins, and those involving mammary parenchyma undergo simple or modified radical mastectomy with no lymph node dissection. Adjuvant radiochemotherapy is still an option considered by some authors [6]. Better prognosis has been associated with adequate surgical margins and with low cellular pleomorphism [1, 13]. Wong et al. review of previous cases suggests that leiomyosarcoma of the nipple-areola complex has less local recurrences or metastases compared to those located on mammary parenchyma, though most cases had no long-term follow up. In general, the prognosis of patients with leiomyosarcoma of the breast is better than that of other sarcomas of the breast [14].
CONCLUSION

Still there is not enough gathered data on this type of neoplasms. We describe a case with a different clinical presentation thus supporting the idea that we need more cases to develop a better understanding of this disease. Long-term follow-up is necessary to understand prognosis thus to find the need for less or more aggressive treatments such as wider surgical resections or the use of adjuvant radiochemotherapy, always considering the added risk of causing more damage or developing second malignancies.

*********

Author Contributions

Xavier Guedes – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Meritxell Medarde – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Tamara Parra – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

MJ Martinez – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Enric de Caralt – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Constanti Serra – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© Xavier Guedes et al. 2014; This article is distributed under the terms of Creative Commons attribution 3.0 License which permits unrestricted use, distribution and reproduction in any means provided the original authors and original publisher are properly credited. (Please see www.ijcasereportsandimages.com/copyright-policy.php for more information.)

REFERENCES

Edorium Journals: An introduction

Edorium Journals Team

About Edorium Journals
Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission
We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?
In less than 10 words - we give you what no one does.

Vision of being the best
We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services
We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review
All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review
All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version
Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status
From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks
You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.

Four weeks
After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.

Mentored Review Articles (MRA)
Our academic program “Mentored Review Article” (MRA) gives you a unique opportunity to publish papers under mentorship of international faculty. These articles are published free of charges.

Most Favored Author program
Join this program and publish any number of articles free of charge for one to five years.

Favored Author program
One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program
Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence
We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...
We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

CONNECT WITH US

Edorium Journals: On Web
Browse Journals