Primary Breast Sarcoma: A rare pathology in women from the National Cancer Institute (INCan) of Mexico

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Abstract

Background: Primary breast sarcoma (PBS) is an extremely rare type of tumor accounting for 0.2% to 1% of all breast cancers and less than 5% of soft-tissue sarcomas. A non-epithelial breast tumor compromised of mesenchymal mammary tissue, PBS has a difficult diagnosis and no standardized treatment.

Objective: The principal aim of this study was to review the Breast Clinic data of patients diagnosed and treated for soft-tissue breast sarcoma at the National Cancer Institute in Mexico City (Instituto Nacional de Cancerología, or INCan).

Methods: A 10-year retrospective review identified only 8 patients diagnosed with PBS. All the available clinical and imaging studies (BI-RADS) were examined, as were the treatments.

Results: Independently of the tumor size, radical mastectomy was practiced on seven patients. After surgical resection, adjuvant therapy was administered to five of these patients. Half of the patients presented distant metastasis. Unfortunately, the five-year overall survival is unknown.

Conclusions: The frequency of PBS in our Institute is much less than that found in the literature. Many clinical characteristics were similar to previous reports, although the treatment was different. The appropriate diagnosis of PBS should allow for the standardization of treatment and improvement of overall survival.

Introduction

Primary breast sarcoma (PBS) is a rare type of cancer arising from the mesenchymal tissue of the breast. It accounts for 0.2% to 1% of all breast malignancies and less than 5% of all soft-tissue sarcomas [1-3]. The annual incidence, estimated at 40 new cases per 10 million women, has remained constant. The pathological classification produces controversy because the disease consists of a heterogeneous group of malignancies, as seen in soft-tissue sarcomas in other parts of the body. Accordingly, PBS includes malignant fibrous histiocytoma, fibrosarcoma, stromal sarcoma, spindle sarcoma, liposarcoma, leiomyosarcoma, osteosarcoma, chondrosarcoma, lymphomas, malignant cystosarcoma phylloides and rhabdomyosarcoma, of which the more common subtypes are angiosarcomas, malignant fibrous histiocytoma and stromal sarcoma [4-6]. Although retrospective single institution experiences have been reported, diagnosis and treatment are based on limited data [7-10]. Unlike epithelial breast cancers, there is still no consensus on the optimal management of PBS. We herein document a retrospective review of PBS using Breast Clinic data from the National Cancer Institute in Mexico (Instituto Nacional de Cáncerología, or INCan), with the aim of describing clinicopathologic features, management and prognosis. The present information is compared to reports from other institutions dedicated to the treatment of cancer.

Material and methods

Founded in 1946, the National Cancer Institute in Mexico is a tertiary care hospital that serves a population of 20 million in Mexico City and offers consultation services to other hospitals in different regions of the country. From 2003-2008, a computer program was developed, and all records were transferred onto an electronic medical record (eMR). Currently, approximately 4,500 patients with breast cancer are treated every year in the INCan, representing 11.7% of the total number of cancer cases in this Institute. We accessed the database and retrieved all cases with a diagnosis of PBS from 2000 to 2010 (10 years), finding 8 patients out of the almost 40,000 having some type of breast malignancy. The H&E tissue sections were examined again to confirm the histopathological diagnosis. The clinical records were retrospectively reviewed to identify clinical characteristics at the time of diagnosis regarding age, family history of breast cancer, duration of the symptoms, clinical tumor size, existence of axillary lymph nodes and/or distant metastasis, and type of surgery and adjuvant treatments (chemotherapy and/or radiotherapy).

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Results

These are summarized in Table 1. The medical reports of the patient's evidence a median age at diagnosis of 50.8 years (range 29-75 years). The clinical progression was calculated from the date of the diagnosis to the date of the first sign of change, with a median follow-up of 7.2 months (range of 1-13 months). Clinically, all patients presented a palpable mass and progressive swelling with or without pain. Palpable axillary lymphadenopathy existed only in two cases. The right breast was affected in 5 cases and the left breast in 3. Lesion size was determined by measuring the maximum diameter, calculating the median clinical tumor size in 7 patients as 3.38 cm (range 3 to 8 cm).

In one patient the size was unknown. Standard medio-lateral oblique and cranio-caudal mammograms had been made with supplementary ultrasound images for all patients. All available images were interpreted by a team of radiologists. The findings were classified according to the American College of Radiology Breast Imaging Reporting and Data Systems (BI-RADS) lexicon [11,12]. Modified radical mastectomy and axillary lymph node dissection was the surgery of choice for one patient.

Another patient initially presented metastasis to the axillary lymph nodes and lung at the time of diagnosis and was inoperable. Six patients underwent a mastectomy and 5 of them were given adjuvant therapy. The median cycle of chemotherapy was 6 months (range 4-8). None of these patients with PBS received adjuvant radiation therapy. A distant metastasis developed in 4 patients, the most common site being the lungs and then bone and adipose tissue (consistent with current knowledge about the clinical course of soft-tissue sarcomas). Overall survival was calculated from the date of diagnosis to the date of death (for any reason) or of last contact. Four patients died of the disease and its complications (two died of lung metastasis), one of unknown cause, and three did not return for treatment (reason unknown).

Discussion

PBS is a rare disease entailing malignant tumors of the breast. Previous reports reveal that its true prevalence is difficult to determine. Between 2000 and 2010 in our institution, PBS had a prevalence of 8 in 40,000 cases of breast cancer (0.02 %), well below the rate evidenced in the literature. Given the rare nature of this malignancy, mammography and ultrasound play only a limited role in diagnostics. According to the reports with mammography, most lesions are round or oval masses with a well-circumscribed margin [13-15]. None of the masses herein observed had spiculated edges or micro-calcifications. The magnetic resonance imaging (MRI) of breasts is not a routine procedure in primary breast sarcoma but may be considered for patients with clinically advanced pathogenesis (including axillary lymph nodes positive to cancer or clinical signs or symptoms of metastasis). Previous studies demonstrate that breast sarcomas are heterogeneous hypointense on T1-weighted images and hyperintense on T2-weighted images (with intensity enhancement after the administration of contrast medium).

Histological grading is not an accurate prognostic factor. Although the first steps of tumorigenesis may be different in PBS and other sarcomas, this classic distinction is of minimal importance from a therapeutic point of view. The international consensus is that mastectomy can still be considered the gold standard. Surgical excision to clear margins is the procedure of choice, while the dissection of axillaries is not always indicated given the rareness of lymph node involvement [16-21]. Unfortunately, the role of adjuvant chemotherapy remains uncertain in the scientific literature [22,23], while the role and the timing of radiation and chemotherapy (pre-surgical vs. post-surgical) has varied [24-26]. Lung, bone and liver are the most common sites for metastasis. Hematic dissemination is the predominant via, although some reports suggest that lymphatic migration may also occur [27-30]. Recent single-institution case series demonstrate that tumor size and margin status are the best prognostic factors for PBS. A tumor size >5 cm is the only significant prognostic indicator of poor survival. The 5-year overall survival rate of PBS is similar to that found for non-breast sarcomas, with a very poor prognosis and no specific treatment guidelines.

Conclusions

PBS requires a multidisciplinary team involving an oncologist, breast surgeon, radiologist, pathologist and chemotherapy oncologist for determining the treatment that offers the best probability of patient survival. According to previous reports, it is unclear which treatment can optimize long-term survival.

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Competing interests

The authors declare that they have no conflicts of interest.

References
