Case report: rare breast tumor—Schwannoma of the breast

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Abstract: Schwannomas are rare, slow growing nerve sheath tumors, one-third of which arise from the head and neck. We report the case of a 37-year-old female who presented with a slow growing, non-tender, left-sided breast mass designated as a Bi-Rads 4 on imaging of the breast warranting further biopsy. A core needle biopsy was obtained of the lesion and appropriately identified as a schwannoma of the breast. Schwannomas have a characteristic histologic appearance of Antoni A, Antoni B, and Verocay bodies with characteristic S100 and SOX-10 staining—all characteristics of which this lesion did possess. The patient did have a history of neurofibromatosis type 2, which previously had manifested as left-sided neck lesions confirmed as schwannomas by pathology following surgical excision of the masses. Schwannomas of the breast are very rare, accounting for only 2.6% of schwannomas. The schwannoma of the breast in this case was appropriately treated with surgical excision and confirmation of the diagnosis on pathology with the entirety of the specimen. In most cases, no further surgical intervention is warranted subsequently. In this case report, we highlight the relevance of consideration of a schwannoma as a breast mass, particularly in patients with a history of neurofibromatosis type 2.

Keywords: Schwannoma; breast; rare; neurofibromatosis type 2 (NF2); case report

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Introduction

A schwannoma, a slow growing tumor made up of Schwann cells which develops in the nerve sheaths of the peripheral nervous system, is extremely rare in the breast. While the cause of this tumor is not known in most cases and occurs spontaneously, some correlation has been identified with genetic disorders such as neurofibromatosis type 2 (NF2), schwannomatosis, and Carney complex. Surgical excision is typically the method of treatment in schwannomas, although radiosurgery may be utilized for schwannomas of the head. Benign schwannomas may be observed, while malignant schwannomas may be treated with immunotherapy and chemotherapy in addition to surgical excision. During excision, if the schwannoma is not removed in entirety, there is a risk of recurrence (1). We present the following case in accordance with the CARE reporting checklist (available at https://dx.doi.org/10.21037/tbcr-21-17).

Case presentation

We report the case of a 37-year-old female who came for evaluation after discovering a nontender, stable left sided breast mass. The patient did have a history of NF2. Historically, the patient's NF2-related lesions were always associated with tenderness and previously manifested as left-sided neck schwannomas, confirmed on pathology following surgical excision. The patient's family history was pertinent for a grandmother with post-menopausal breast cancer. On physical exam, the patient did have a palpable, non-tender breast mass with no associated tissue texture changes, overlying skin changes, nipple retraction, nipple discharge, or palpable lymphadenopathy.

A bilateral 3d mammogram was obtained, with evidence of an oval, circumscribed mass measuring 1.5 cm × 1 cm × 1.5 cm with internal flow at the 5 o'clock position, 8 cm from the nipple (*Figure 1*); a supplemental limited left sided

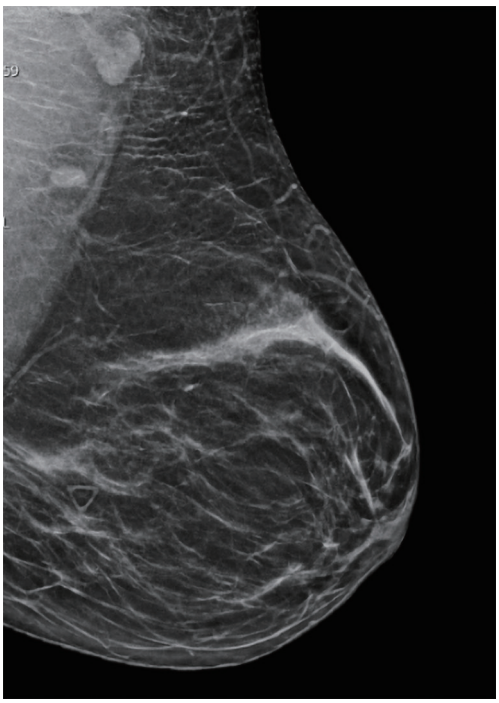

Figure 1 Focal asymmetry (1.2 cm × 1 cm).

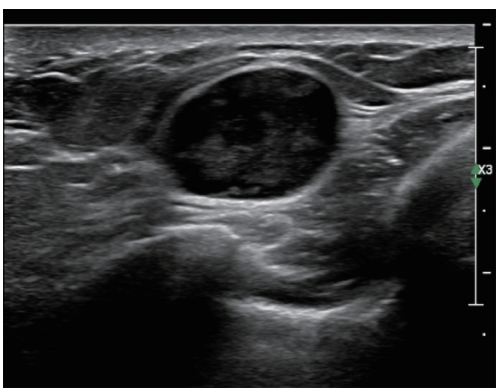

Figure 2 Sonographic evidence of oval-shaped, well-demarcated, hypoechoic solid mass in the left breast at the 5'oclock position 8 cm from the nipple measuring 1.5 m × 1 cm × 1.5 cm.

ultrasound was obtained with evidence of a hypoechoic, oval mass with internal flow (*Figure 2*). The lesions of investigation were determined to be a Bi-Rads 4 lesions, warranting biopsy.

A vacuum-assisted core needle biopsy demonstrated spindle cell proliferation and revealed Antoni A and Antoni B areas; Verocay bodies were additionally noted. The lesional cells were positive for S100 and SOX-10; and negative for AE1/3, desmin, smooth muscle actin, and epithelial membrane antigen (EMA); CD34 showed patchy staining.

Tissue sections showed a spindle cell proliferation with Antoni A and Antoni B areas, which are alternating interlaced bundles of spindle cells and hypocellular areas, respectively. Verocay bodies, areas of density in schwannomas between palisading bodies of spindles cells in Antoni A regions, were noted (2). The lesion cells were also positive for S100 and SOX-10, notable markers for neural crest cell derived tumors (3); and the tissue was negative for AE1/3, desmin, smooth muscle actin, and EMA; CD34 shows patchy staining. The overall histological findings as well as staining pattern were consistent with a schwannoma. This case was peer reviewed with a second pathologist for intradepartmental consultation and diagnosis confirmation.

Given the results of the biopsy, recommendations for surgical excisions were made. Surgical excision of a 4.2 cm × 2 cm × 2.8 cm mass was undertaken, and sent for further pathology evaluation. Pathology of the complete specimen was consistent with biopsy features, indicating a benign schwannoma of the left breast with negative margins.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Schwannomas are slow growing tumors of the nerve sheath, which typically arise from the peripheral nerves or spinal nerve roots. The most common locations of this tumor include the head, neck, and extensor surfaces of the extremities; breast schwannomas account for only 2.6% of schwannomas, most ranging from 7 mm to 11 cm (4,5). Very limited findings of schwannomas of the breast are reported, with our review of the English literature discovering 25 previously described cases (6,7). Approximately 1/4 of the cases were identified as a schwannoma on preoperative diagnosis, and notably 1/3 of patients with a documented schwannoma of the breast have been male (6), while schwannomas may grow large, they are not associated with a risk of lymphatic spread. For this reason, it would not be indicated to do a sentinel lymph node biopsy in conjunction

with the excisional biopsy (8).

The majority of schwannomas tend to appear as well defined, nonspecific, round or oval densities on mammogram, and well defined, solid, hypoechoic masses on ultrasound (9). The findings in our cases were mostly consistent with these findings as described above. However, the diagnosis could not simply be made by radiologic characteristic; in a majority of cases, the diagnosis of schwannoma required excisional biopsy after core needle biopsy for appropriate diagnosis.

Our patient did have NF2, which is an autosomal dominant disorder characterized by multiple peripheral and central tumors; while different types of neurofibromatosis exist, type 2 is characterized by bilateral vestibular schwannomas, cutaneous schwannomas, and meningiomas. It is caused by a loss of function mutation of the *NF2* gene, located on band 22q12 and codes for merlin; merlin itself is a tumor suppressor as well as a cell membrane protein (10). While vestibular schwannomas are characteristic of NF2, individuals do possess a higher risk of schwannomas developing in other regions of the body (11).

Conclusions

In conclusion, based on the results that we have obtained through the review of the literature as well as our case, we recommend excision when the consideration of schwannoma as a possible diagnosis for breast mass arises. We also illustrate the relevance of including schwannoma as a differential diagnosis for breast masses, particularly in patients with a history of NF2.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://dx.doi.org/10.21037/tbcr-21-17

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://dx.doi.org/10.21037/tbcr-21-17). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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