Management of Neuroendocrine Ductal Carcinoma In Situ (DCIS) of the Breast

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Introduction

Neuroendocrine tumors are defined as tumors that express neuroendocrine markers, specifically synaptophysin and/or chromogranin in at least 50% of tumor cells. To date, neuroendocrine cancers of the breast are extremely rare with reports that they account for less than 1% of all neuroendocrine tumors and less than 0.1% of all breast cancers. Limited literature exists to guide treatment decisions tailored to neuroendocrine tumors of the breast, despite the possibility that they may be associated with an overall worse prognosis.

Methods

We present a rare case of neuroendocrine ductal carcinoma in situ (NE-DCIS) which was discovered upon work up for bloody nipple discharge. In this case, NE-DCIS was managed with the standard recommended treatment regimen for ductal carcinoma in situ (DCIS).

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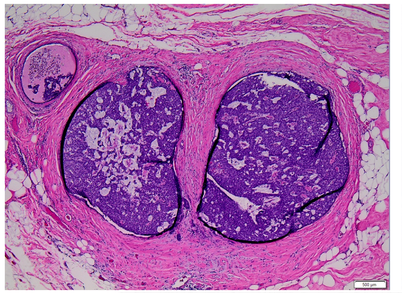
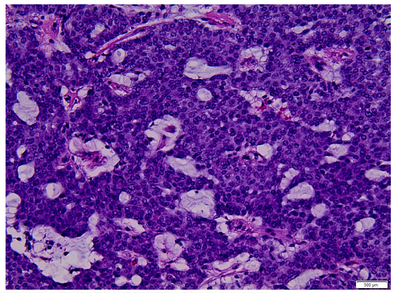
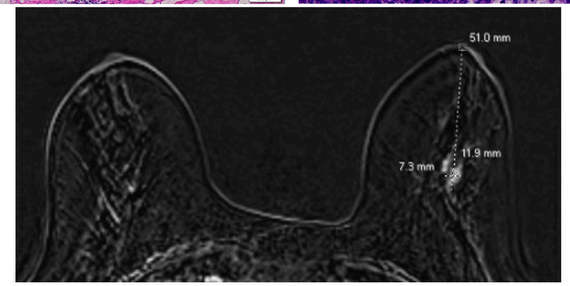
Results

Our patient was a 72-year-old Caucasian female who presented to breast clinic with 3 months of spontaneous, unilateral bloody nipple discharge from her left breast. Bilateral diagnostic mammograms were performed and showed single duct spontaneous bloody nipple discharge without definitive mammographic or ultrasound correlate. Magnetic resonance imaging (MRI) was then performed, as well as an MRI-guided biopsy, which resulted in a linear enhancement measuring 1.2 x 0.7 centimeters (cm) in the lower, central, middle third of the left breast. Pathology revealed severely atypical intraductal proliferation involving an intraductal papilloma and suspicious for intermediate grade ductal carcinoma in situ (DCIS) with neuroendocrine differentiation. Patient then underwent needle-localized left partial mastectomy with onco-plastic closure. Final pathology confirmed NE-DCIS composed of papillary type DCIS with estrogen receptor (ER) positivity at 94% and progesterone receptor (PR) positivity at 89% with neuroendocrine features staining positive for synaptophysin in 100% of cells. Final negative margins were less than 1 millimeter (mm) and patient is currently scheduled to undergo re-excision with intraoperative radiation and plans for post-operative Tamoxifen therapy.

Imaging







Conclusion

We risk underdiagnosis due to controversy of non-operative management with surveillance compared to surgical excision for intraductal papilloma. Due to lack of available literature, there are no guidelines specific to NE-DCIS. We revert to recommended standardized treatment regimen for DCIS, despite the possibility there may be an associated worse prognosis. We advocate for future prospective study in the guidance of management of this rare malignancy.