CLINICAL VIGNETTE

Granular Cell Tumors of the Breast: A Breast Carcinoma Mimic

Malena SC Law, MD and Gloria S Kim, MD

A 52-year-old G4 P3013 postmenopausal, non-smoking female presented to her primary care physician's office with right axillary pain. She noticed discomfort for several months and thought this was due to a new deodorant. She then noticed a lump and presented for evaluation. She had no significant past medical history and no history of hormone replacement therapy, abnormal mammograms or breast biopsies. She denied any skin changes or nipple discharge. On physical examination, a palpable, firm, round, mobile mass approximately 1 cm in diameter in her right axilla was noted, without skin changes or nipple discharge. No other breast masses were noted.

Diagnostic mammogram revealed a suspicious 12 mm x 9 mm spiculated mass. Targeted ultrasound confirmed a solid mass at the site of the clinically palpable lesion. Ultrasound-guided core-needle biopsy, which revealed granular cell tumor and she was referred to general surgery for wide local excision.

Granular cell tumors are rare neoplasms that originate in the Schwann cells of the peripheral nervous system[1]. They can arise anywhere in the body but most commonly occur in the oral cavity or head and neck region, with only 5-6% of cases in the breast [2,3]. They occur more commonly women. Granular cell tumors of the breast are usually benign, but can be a diagnostic challenge mimicking breast cancer clinically, mammographically and sonographically. They are more common in premenopausal women and in blacks[1]. They can occur at any age, with average age of 40 years. A review of 159 cases of granular cell tumors of breast reported an age range of 15 to 74 years with 9.8% cases in males[4]. Less than 1% of cases occurred in adolescents[3].

Granular cell tumors of the breast typically present as painless, firm or hard palpable masses that average 1 to 2 cm in size. They more commonly are found in the upper inner quadrant of the breast (whereas, breast carcinomas more frequently arise in the upper outer quadrant). The tumors grow slowly and usually present as a solitary mass. In addition to their firm texture on physical exam, they can also present with dimpling of the skin, fixation of the mass to the skin, and even ulceration, thus adding to the mimicry of breast carcinoma[3].

Sonographically, they can appear as ill-defined solid masses or circumscribed masses with posterior acoustic enhancement. On mammogram evaluation, they can appear as spiculated masses with margins that appear infiltrative. They frequently can have associated microcalcifications with patterns that can be suggestive of ductal carcinoma in situ[2].

Histologically, the tumors appear to have sheets or nests of loosely infiltrating polygonal cells or large round cells with abundant eosinophilic granular cytoplasm[3]. Immunohistochemical analysis is positive for S-100 protein, PAS (periodic acid Schiff), neuron specific enolase (NSE), and CEA[1]. They tend to be negative for estrogen and progesterone receptors[3]. Granular cell tumors are felt to originate from Schwann cells and arise from intralobular breast stroma[3]. They can form cords that extend into adjacent breast tissue[3]. This pattern of growth leads to the dimpling and infiltrative appearance of the skin over the tumor[3].

Rarely, granular cell tumors can be malignant (less than 1% of all granular cell tumors are malignant). Features that are more concerning for malignancy include: large tumor size (greater than 4cm), rapid growth, local invasion, presence of necrosis, increased mitotic activity (>2 mitosis per 10 high-power fields, and cellular and nuclear pleomorphism[3]. Malignant granular cell tumors are categorized as high-grade sarcomas and are associated with a high rate of metastasis and short survival time[5].

Currently, definitive management includes only wide surgical excision[3]. Chemotherapy has no role in management of granular cell tumors and adjuvant radiation is typically only given if the tumor is malignant[3]. Local recurrence rates range from 2-8%[5]. Local recurrence typically occurs within the first year after surgery; however, it can occur even as late as 10 years after excision[3]. With malignant granular cell tumors of the breast, distant metastases are reported as high as 60%[5]. Due to the high rate of local

recurrence and remote risk of distant metastases, regular follow up is essential.

Granular cell tumors of the breast can be diagnostically challenging with clinical and radiographical resemblance to breast carcinoma. Core needle biopsy is essential as the management of a benign granular cell tumor of the breast involves wide excision of the tumor versus lumpectomy or mastectomy or radiation for breast cancer.

REFERENCES

1. **Gogas J, Markopoulos C, Kouskos E, Gogas H, Mantas D, Antonopoulou Z, Kontzoglou K**. Granular cell tumor of the breast: a rare lesion resembling breast cancer. *Eur J Gynaecol Oncol*. 2002;23(4):333-4. PubMed PMID: 12214737.
2. **Scaranelo AM, Bukhanov K, Crystal P, Mulligan AM, O'Malley FP**. Granular cell tumour of the breast: MRI findings and review of the literature. *Br J Radiol*. 2007 Dec;80(960):970-4. Epub 2007 Oct 16. PubMed PMID: 17940129.
3. **De Simone N, Aggon A, Christy C**. Granular cell tumor of the breast: clinical and pathologic characteristics of a rare case in a 14-year-old girl. *J Clin Oncol*. 2011 Aug 1;29(22):e656-7. doi: 10.1200/JCO.2011.35.9448. Epub 2011 Jun 6. PubMed PMID: 21646617.
4. **Boulat J, Mathoulin MP, Vacheret H, Andrac L, Habib MC, Pellissier JF, Piana L, Charpin C**. [Granular cell tumors of the breast]. *Ann Pathol*. 1994;14(2):93-100. Review. French. PubMed PMID: 8198646.
5. **Fanburg-Smith JC, Meis-Kindblom JM, Fante R, Kindblom LG**. Malignant granular cell tumor of soft tissue: diagnostic criteria and clinicopathologic correlation. Am J Surg Pathol. 1998 Jul;22(7):779-94. Erratum in: *Am J Surg Pathol* 1999 Jan;23(1):136. PubMed PMID: 9669341.

Submitted on August 9, 2013