Granular cell breast cancer: A rare misleading lesion

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Case report

A 42-year-old multiparous woman with no particular past personal family history was referred after a right breast lesion ACR class 5 was found on ultrasound in a routine breast assessment. She had no palpable abnormalities and her axillary fossae were clear.

Mammography showed distortion of the architecture in the supero-external quadrant of the right breast, apparent on CT films (Fig. 1) and appearing on ultrasound as a highly attenuated 6 × 5 mm hypoechogenic mass, long axis perpendicular to the deeper planes with poorly demarcated, indistinct outlines (Fig. 2).

Ultrasound-guided microbiopsies were taken and showed no carcinomatous cells, although there were findings consistent with a granular cell tumour. As a result of the inconsistency between anatomical and radiological findings, a lumpectomy was performed after identifying the tumour with a metal suture (Fig. 3). Histological and immunohistochemical examination confirmed that this was a benign granular cell tumour, measuring 15 mm along its long axis. The tumour was not demarcated and was infiltrating around epithelial mastosis structures (Fig. 4). In the absence of signs of malignancy, it was reclassified as ACR 2 and breast ultrasound follow-up in 2 years was organized.

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Figure 1. Subtle architectural distortion of the supero-external quadrant of the right breast, which is difficult to detect on postero-anterior (a) and lateral-internal (b) views, because of the BI-RADS classification type III breast density. Additional lateral tomosynthesis (c) confirms this lesion as class ACR 5.

Discussion

The granular cell tumour (GCT) was initially described by Abrikossoff in 1926 as a “granulo-cellular myoblastoma”. It is a rare, usually benign lesion, the histogenesis of which has long been discussed and remains uncertain [1]. It is currently assumed to be neuronal in origin, originating from Schwann cells [2] as it expresses protein S100. This hypothesis is also supported by the absence of acid gliofibrillar protein and neurofilaments.

GCT is a ubiquitous tumour, mostly affecting soft tissues, particularly the oral cavity, in 70% of the cases [3].

Rarer sites, such as the gastrointestinal tract (esophagus, anal margin), tracheobronchial tree and glandular organs (parotids and thyroids), kidney and eye orbit have been reported [4]. The age at the onset of the lesion is generally between 20 and 60 years old and it has a female predominance (M/F: 1/2). Cases in children and multifocal tumors have been described.

Breast locations are rare (approximately 5 to 6% of all GCT) and represent approximately 1/1000 of breast tumors. African-American and premenopausal women are more commonly affected [5]. The tumor is generally unique and tends to be found in the upper quadrants, particularly the supero-external quadrant. It is usually an asymptomatic nodule under 3 cm in size.

Figure 2. Ultrasound appearances of the ACR 5 mass in the supero-external quadrant of the right breast, showing an irregular hypoechoogenic mass with poorly demarcated outlines, measuring 6 × 5 mm.

Figure 3. Macroscopic appearances of the lumpectomy specimen, showing the star-shaped lesion.
The mammography and ultrasound appearances of the GCT vary. They may appear as round well-demarcated masses, suggestive of a benign lesion, such as a cyst or fibroadenoma. They may also have disorganized architecture, with an area of convergence of fibrous and connective tissue sheets, suggestive of infiltrating carcinoma. Microcalcifications are usually not present. The abnormality is sometimes difficult to detect in a breast because of the density of the breast tissue and the heterogeneous distribution of the connective tissue and glandular matrix. The lesions were identified with computed tomography in this case.

Histologically, the lesion is well demarcated and not encapsulated. It is made up of round cells with granular eosinophilic cytoplasm. The nuclei are small and do not display atypia. The tumour cells are either isolated or grouped in clusters and may partly infiltrate the surrounding connective tissue in a star-shaped arrangement, explaining the occasionally worrying mammographic appearances. A key factor in the histopathological diagnosis is immunohistochemical expression of protein S100, neuron specific enolase (NSE) and vimentin [6].

Abrikossoff tumors must be investigated in order not to miss a malignant lesion, particularly, if the microbiology results are inconsistent with the mammography and ultrasound appearances. An unequivocal diagnosis can only be made after the lesion has been completely excised.

One to three per cent of the tumors are malignant [4]. This may be suggested clinically if it is over 4 cm in size, if it contains heterogeneous necrotic areas, or rapid clinical or radiological changes develop. Histology then shows multiple cellular atypia, areas of necrosis, with many mitoses and a high ki67 proliferation index. The diagnosis of malignant degeneration is supported by the presence of lymph node or visceral metastases, which are histologically identical to the primary tumour.

Surgical excision remains the treatment of choice to obtain an unequivocal diagnosis. The excision margins should be wide to ensure that no signs of malignancy are present [7].

**Conclusion**

The granular cell tumour, or Abrikossoff tumour, is a rare benign neurogenic tumour, which is found ubiquitously and predominantly in the ear-nose-throat (ENT) region. It is uncommon for it to be located in the breast. The lesion can be confused with an infiltrating malignant tumour on mammography and ultrasound. An unequivocal diagnosis can only be made from immunohistochemical analysis of the whole lesion. It carries a good prognosis after surgical excision. Malignant forms of the tumour have been described and its excision margins therefore need to be wide.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

**References**


