Radiologic evaluation and imaging-pathologic correlation of uncommon tumor of the breast

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Learning objectives

1. To show several rare breast tumors.
2. To describe the most relevant clinical and pathologic features.
3. To recognize the appearances of this tumors on radiologic images (ultrasound, mamography, and magnetic resonance).
Background

Unusual breast tumors suppose a challenge in image diagnosis. Knowing its radiologic features, helps us to realize an early presumptive diagnosis , that later will be confirmed by pathology.

We will show and describe the radiological findings of some rare breast tumors. We have also added a radiopathologic correlation and a brief literature review of each of them.

We have classified them into two groups, in order to facilitate the exhibition: benign tumors and malignant tumors.

A) BENIGN TUMORS:

1. GRANULAR CELL TUMOR: (Fig.1)

Granular cell tumor (GCT) of the breast is a rare neoplasm that accounts for 5% to 15% of all GCTs and occur most frequently in middle-aged women. It is usually benign and be derived from perineural cells on the basis of their immunophenotypic and ultrastructural characteristics. In 1-2% can be either malignant. In these cases, are presented as a large masses which have rapid growth.

They often mimic breast carcinoma on clinical examination and radiological characteristics (spiculated nodules without calcifications at mammography and solid, irregular nodules with acoustic shadow by ultrasound).

Sometimes is associated with intraductal carcinoma.

2-8% recurrences.

2. HIBERNOMA: (Fig.2)

Hibernomas are extremely rare benign soft tissues tumors, derived from brown fat. Most hibernomas occur in sites where brown fat persists beyond fetal life (usually in the interscapular region or thigh), but they also occur in sites where brown fat is usually absent. There are few cases reported. In the breast is still more rare. There are asymptomatic unless they have rapid growth.
Radiological findings:

- Mammography: may be normal or isodense compared to breast fatty tissue.
- Ultrasound: hyperechogenic and highly vascularized lesion.
- MR: On T1 and T2 weighted imaging, it is similar to subcutaneous fat. Gadolinium: early and marked contrast enhancement. The enhancement kinetics curves shows a rapid initial rise, followed by a drop-off whit time (washout) in delayed phase (Type 3 curve).
- Microscopically, the tumor is characterized by large multivacuolated cells with scanty granular eosinophilic cytoplasm and eccentric nuclei, univacuolated cells, with peripheral nuclei, and smaller round cells with granular cytoplasm.

The differential diagnosis includes: hemangioma, lipoma, angiomyolipoma, liposarcoma.

3. SCHWANNOMA (Fig.3)

Schwannomas or neurilemmomas are benign neoplasms arising in Schwann cells. The most common locations include the head, neck, trunk and flexor surface of the extremities, the breast being a highly unusual location. (there are very few cases reported), and are mostly benign. These tumors may arise sporadically or in the context of neurofibromatosis. On the mammography show features of benign nodules (well-circumscribed, oval shaped nodule without microcalcifications) and ultrasound imagin will tend to reveal a well-defined hypoechoic solid mass, usuallly with posterior enhancement.

B) MALIGNANT LESION.

1. TUBULAR CARCINOMA. (Fig. 4)

Tubular carcinoma of the breast is a subtype of invasive ductal carcinoma it tends to be a less aggressive type that responds well to treatment. It isn't likely to spread outside the breast and is considered to have a very good prognosis. Accounting for around 2% of all breast cancers. The average age of diagnosis for tubular carcinoma is the early 50s.

Tubular carcinomas are usually small (about 1 cm or less). It can be multicentric (28%) and bilateral (12-14%).

2. MUCINOUS CARCINOMA. (Fig 5)
It tends to occur in older women where a prevalence of as much as 7%.

In mucinous carcinoma, mucin becomes part of the tumor and surrounds the breast cancer cells.

Even though mucinous carcinoma is an invasive breast cancer, it tends to be a less aggressive type that responds well to treatment. Mucinous carcinoma is less likely to spread to the lymph nodes than other types of breast cancer.

Usually, at mammography the presence of mucin results in a low-density and relatively well-defined lobular mass. Sometimes they may have partly faded or obscure margins.

3. MEDULLARY CARCINOMA. (Fig.6)

It is a rare sub type of breast cancer and accounts for ~ 2 - 7 % of all breast cancers.

They tend to occur more frequently in younger women (46 - 54 years)

Clinically, these tumours are characterised by rapid growth and therefore often manifest as palpable masses, mobile and soft.

At mammography, most frequent forms of presentation are well defined masses without calcifications and by ultrasound hypoechoic mass with distal enhancement.

4. SIGNET RING CELL CARCINOMA. (Fig.7)

Signet-ring cell carcinoma of the breast accounts for between 1-4% of all breast cancer and tend to occur more frequently in older women.

It is a variant of the in situ- lobulillar carcinoma, poorly differenciated.

Usually there is lymph node involvement and metástasis. The prognosis for breast signet rings cell carcinoma is generally poorer than for other breast cancer.

Histologically characterized by proliferation of cells that accumulate mucin in big intracytoplasmic vacuoles and displace the core to the periphery.

Asymmetric density or areas of architectural distortion is the most frequent form of presentation by mammography.

5. MICROPAPILLARY CARCINOMA. (Fig.8)
Invasive micropapillary breast carcinoma is a very aggressive form of breast cancer, with a very high rate of lymph node metastasis and high rate of recurrences. These represents under 2% of breast cancers, frequently associated with ductal carcinoma. Clinical and mammographic features were not different from those of ordinary invasive carcinoma of the breast.

Mammographic studies tend to show a high density, irregular or round solid mass, with spiculated margins, and microcalcifications present only at 50% of all the cases. These microcalcifications tend to be either pleomorphic, punctate, or both, and their distribution tends to be segmental or clustered.

Ultrasound images show a solid hypoechoic mass with irregular or microlobulated margins, usually with homogenous echotexture and posterior acoustic shadowing.

6. BREAST SARCOMA. (Fig. 9, Fig.10 and Fig.11)

Sarcomas are malignant tumors that occur in the body's soft tissues such as fat, muscle, nerves and blood vessels. Breast sarcomas in particular develops in the connective tissue within the breast. These are a very rare form of breast cancer that accounts for just 1% of all breast cancer cases.

The typical clinical presentation is a unilateral breast mass that grows in size more rapidly than epithelial breast cancer. The size of these tumors is variable, ranging from less than 1 cm to larger than 40 cm. Metastases typically occur hematogenously (lung, bone marrow and liver). Axillary lymph node involvement is rare.

Mammography and ultrasound are nonspecific. The majority presented as noncalcified oval masses with indistinct or circumscribed margins at mammography and at sonography usually are solid hypoechoic masses with indistinct margins.

7. LYMPHOMA. (Fig. 12)

Breast lymphomas represent less than 0.5% of the tumours of breast. Most are non-Hodgkin's type lymphoma.

Usually appear in women with an average age of 55 years.

Clinically, these tumors are characterised by rapid growth and therefore often manifest as palpable masses (1-7 cm).

At mammography, most frequent forms of presentation are well defined masses without calcifications and by ultrasound hypoechoic nodules.
There are lymph node metastasis in 30-50%.

8. **NEUROENDOCRINE CARCINOMA.** (Fig. 13)

Neuroendocrines tumors represent less than 2% of malignant breast cancer. The diagnosis must be made by histology and confirmed with Immunohistochemistry techniques. These are tumors of low biological aggression and they have not systemic symptoms due to the release of hormones as in other locations.

Often the neuroendocrines tumors are asymptomatics and they have good prognosis.

The treatment must be based on the clinical stage of the disease at the time of diagnosis, and in the majority of cases the treatment is conservative.

At mammography, the most frequent form of presentation is a hypodense nodule with irregular borders or density asymmetric and by ultrasound hypoechoic nodules with distal enhancement.

9. **METASTATIC BREAST.** (Fig.14 and 15)

They represent the 0,5-3% of all breast cancer.

Clinically metastatic are manifested as peripheral well delimited, solitary and rounded mass.

There are three types of metastases: hematogena, lymphatic and hematologic (leukemia and Lymphoma).

- **Hematogena:** Radiologicas features: Mammography: rounded, circumscribed, not spiculated nodule without calcifications. Ultrasound: hypoechoic rounded nodule. MR: rounded nodule with annular enhancement.

- **Lymphatic:** It is characterized by the appearance of thickening skin ("orange peel").

- **Hematologic:** It is characterized by the appearance of axillary lymph nodes.
Fig. 1: A 61 years old woman. Mammography: Nodule of 16 mms in upper outer quadrant of the left breast, shows morphology irregular and poorly defined margins (A and B). Ultrasound: Solid, irregular nodule with angulated edges and acoustics shadow(C). Pathological findings: Cells ovoid with cytoplasm eosinophilic, with granules PAS positive. S-100 Protein(D and E). GRANULAR CELL TUMOR.

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**Fig. 2:** A 75 years old woman. In PET zone of hypermetabolism (12 cms) in lower outer quadrant of the right breast suspicious of neoplasia. Mammography: Isodense lesion partially encapsulated (white arrows)(A). Ultrasound: Hypoechoic and hypervascular mass with indistinct borders, not easily seen(B). MR: Large mass hyperenhanced (C). Pathological findings: Shows characteristics of hibernoma (D). HIBERNOMA.
**Fig. 3:** A 41 years old woman. Asymptomatic with family history of breast cancer. Mammography: Nodule with well defined borders, located in deep layers of the union of outer quadrants of the right breast (white arrow) (A and B). Ultrasound: Hypogenic nodule with well defined margins (13 mm) (C). Pathological findings: Palisade cells and S-100 positive (D). SCHWANNOMA.

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**Fig. 4:** A 49 years old woman who presented a palpable nodule in the upper outer quadrant of the left breast. Mammography: Asymmetric density with pleomorphic microcalcifications (A). Ultrasound: Hypoechogetic nodule (16mm) with ill-defined margins (B). MR: Nonmass-like enhancement. The enhancement kinetics curves shows a rapid initial rise, followed by a drop-off with time (washout) in delayed phase (Type 3 curve)(C). Pathological findings: Large amount of cells lining tubules with loss of lobular architecture (D). TUBULAR CARCINOMA.

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**Fig. 5:** A 71 years old woman. Palpable nodule retroareolar of the right breast. Mammography: Relatively well-defined polilobular mass(A). Ultrasound: Mixed cystic and solid components, distal enhancement (B). Pathological findings: Abnormal cells that "float" in pools of mucin (C). MUCINOUS CARCINOMA.

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**Fig. 6:** A 70 years old woman with palpable nodule in upper outer quadrant of left breast. Mammography: Nodule with well defined borders (A). Ultrasound: Hypoechoic nodule (22 mm) with distal enhancement (B). Pathological findings: Poorly differentiated cells with scant stroma and prominent lymphoid infiltration (C). MEDULLARY CARCINOMA.

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A 47 years old woman with palpable nodule in left breast. Mammography: Density asymmetric with irregular borders in upper outer quadrant (white arrow)(A). Ultrasound: Heterogeneous nodule (12mm) with irregular margins (B). MR: Nodule with intense and early enhancement (C). Pathological findings: Typical "signet-ring" cells (D). SIGNET RING CELL CARCINOMA.

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Fig. 8: A 61 years old woman with palpable nodule periareolar in the right breast. Mammography: small nodule with partially spiculated margins and few microcalcifications (A). Ultrasound: nodule with irregular margins and posterior acoustic shadow (B). Pathological findings: pseudopapillary and tubuloalveolar structures floating in empty, clear spaces lined by delicate strands of stroma.(C). MICROPAPILLARY CARCINOMA.
**Fig. 9:** A 51 years old woman, presenting secretion through the right nipple. Mammography: asymmetric density of irregular and ill-defined margins. (A). Ultrasound: hypodense lesion with irregular borders and posterior acoustic shadow. (B). MR with gadolinium: right breast: early and intense ring-shaped contrast enhancement. (C). Pathological findings: areas with formation of osteoid substance. Positive for vimentine. (D). OSTEOSARCOMA.

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**Fig. 10:** A 34 years old woman with palpable mass at left breast. Mammography: left breast : large mass that occupies almost the entire breast. (A) Ultrasound: heterogeneous mass of imprecise margins. (B) The result of the biopsy was carcinosarcoma (not shown). CARCINOSARCOMA.

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Fig. 11: A 53 years old woman with palpable nodule at right breast. Mammography: dense nodule with well-defined edges located in the upper outer quadrants of the right breast. (A). Ultrasound: solid, hypoechoic and well defined mass.(B). Pathological findings: positive for smooth muscle actin. (C). LEIOMIOSARCOMA.

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**Fig. 12:** A 76 year old woman with hypermetabolic mass in the breast righ in CT. Mammography: Hyperdense mass (6 cm) in lower inner quadrant with ill-defined margins (A). Ultrasound: Large partially lobulated irregular margins nodule with intraparenchymal projections (B). Pathological findings: CD20 positive. Marginal zone lymphoma (C).

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**Fig. 13:** A 49 Year old woman with palpable nodule in inner upper quadrant of the left breast. Mammography: Density asymmetric with microcalcifications (white arrow)(A). Ultrasound: Multiple nodules with irregular and poorly defined borders and microcalcifications (B). MR(C): Left breast: Multifocal carcinoma. Right breast: Nodule suspicion of malignancy. Pathological findings (D): Left breast: Neuroendocrine carcinoma. Right breast: Tubular carcinoma.

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Fig. 14: 40 years old woman with melanoma of choroidal and palpable nodules in both breasts. Mammography: Several nodules in both breasts (white arrows)(A). Ultrasound: Complex cysts of different sizes in both breasts(B). MR: Nodules with annular enhancement and The enhancement kinetics curves shows a rapid initial rise, followed by a drop-off whit time (washout) in delayed phase (Type 3 curve) (C). The result of the biopsy was METASTATIC MELANOMA(not shown).

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**Fig. 15:** 60 years old women with ovarian cancer and nodule palpable in the right breast. Mammography: Nodule (5 mm) in the lower outer quadrant of the right breast (white arrow)(A). Ultrasound: Cyst complicated (B). Pathological findings: Cells WT1 and CK7 positive and markers of breast negatives (C). OVARIAN CARCINOMA METASTASES.

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Findings and procedure details

A retrospective study of a period of five years, we found 91 cases of rare breast lesions.

In all cases, core biopsy and ultrasound guidance or stereotactic vacuum gun was performed, and fine needle aspiration of suspicious nodes.
Conclusion

Knowledge of the radiological characteristics of the rare breast lesions, helps the radiologist to make a correct diagnosis and therapeutic management, in order to avoid unnecessary aggressive treatments although definitive diagnosis requires histological performing analysis.
References


