

# Chapter 3

## Lesions of Vascular and Neural Origin

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## Chapter 3

# Lesions of Vascular and Neural Origin

### Learning Objectives:

1. To aid adequate imaging including doppler for excluding benign etiologies which may appear worrisome.
2. To discuss diagnostic pathway and further management of the affected patients.
3. To empower quick recapitulation of the topic content through a quiz-based approach.

## Introduction

This is the third part of the series that will strengthen your expertise in recognition and management of potential malignancy mimics.

This module illustrates a very unique collection from international sources and will familiarize you with the most challenging conditions of vascular and neural origin.

The main goal is to inspire you to turn your new knowledge into action.

## Vascular Lesions

Vascular lesions of breast can be classified into benign and malignant, but in our chapter we shall be emphasizing on the benign counterparts. They represent a very rare diagnostic dilemma in breast radiology and may mislead dedicated breast imagers regardless of experience.

We will discuss interesting cases of mammary angiomatosis, vascular malformation, and breast aneurysm or hemangioma that simulate cancer.

## Mammary Angiomatosis

This is one of the rarest vascular tumors in breast radiology. They clinically present as gradually progressive breast lump.

Appearances on sonography and mammography may be a matter of concern for primary malignancy.

Mammography may show well-defined homogeneous opacity with irregular margins and increased density (**Fig. 3.1a**, *arrows*).

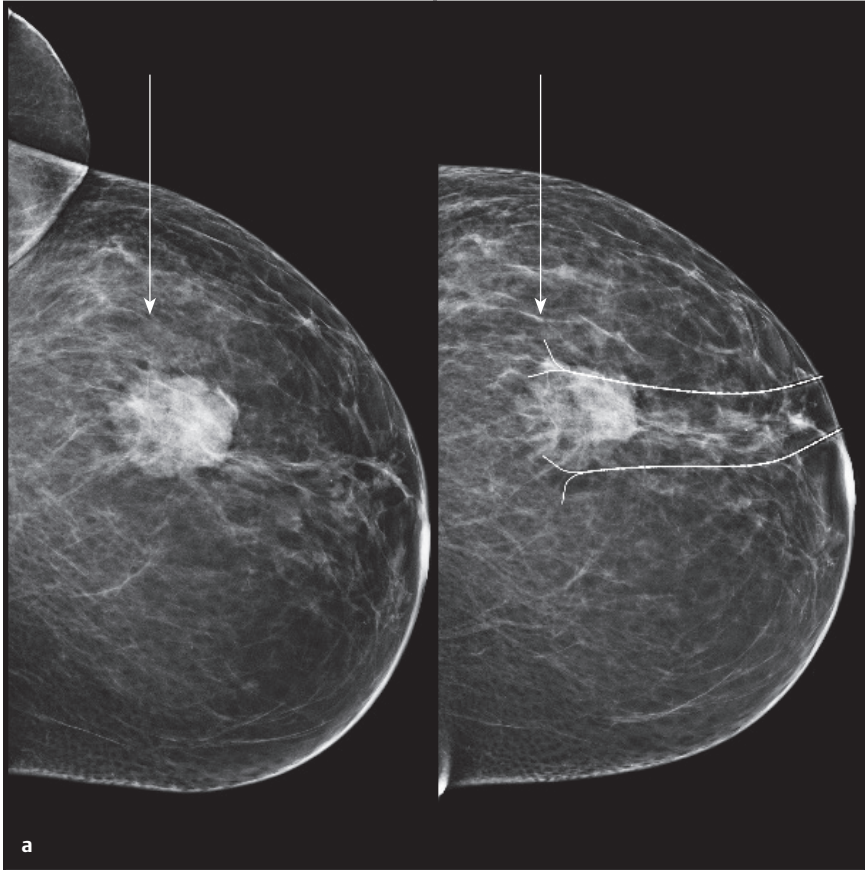
Sonographic features of a heterogeneously hyper to hypoechoic, often a diffuse mass with irregular margins, may raise concerns for malignancy such as lobular breast carcinoma (**Fig. 3.1b, c**). Physical examination can reveal nontender and palpable mass.

This unusual condition carries the risk of local recurrence. Thus, wide local excision with clear margins is required. Mastectomy can be considered in case of a very extensive disease.

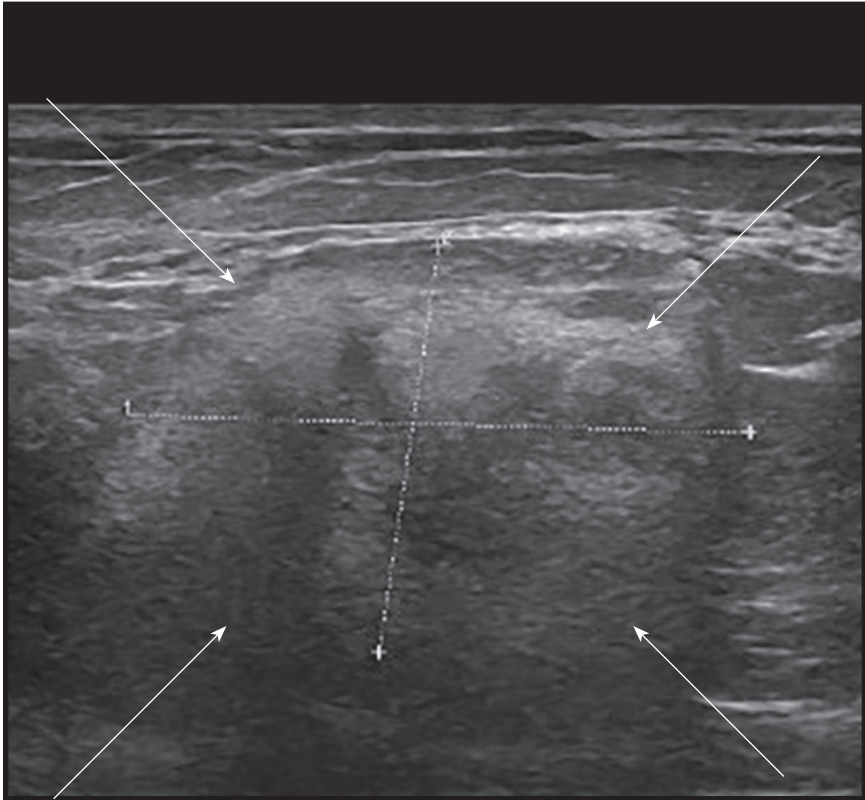
This is an entirely benign entity and prognosis is excellent. To the best of knowledge there is no proven association between mammary angiomatosis and von Hippel-Lindau disease or other phakomatoses.

Specimen histology usually show large irregular spaces lined by flattened endothelial cells lacking muscular wall. Sometimes, red blood cells can be seen in these spaces.

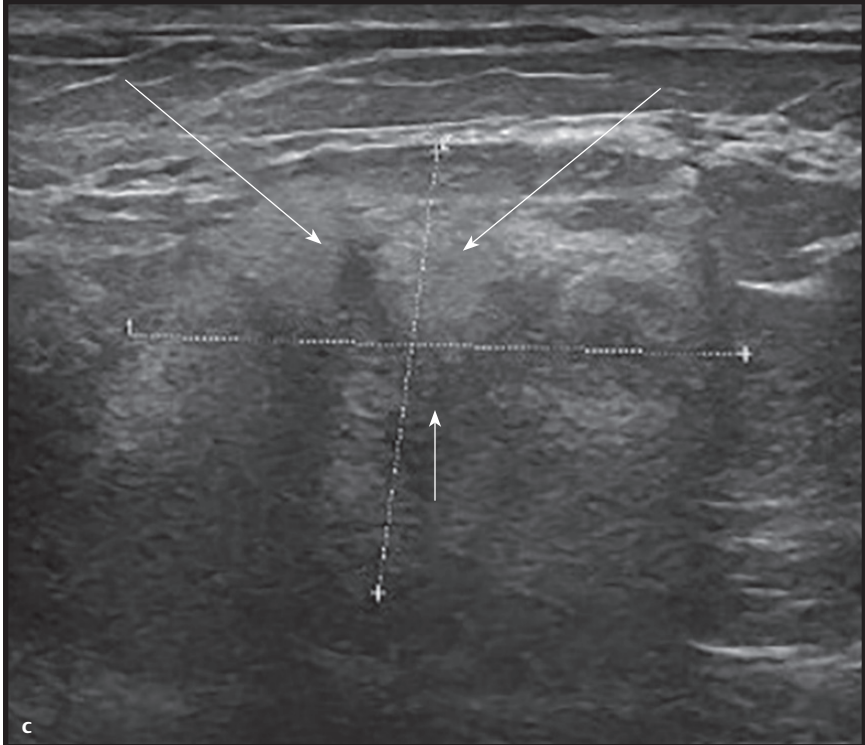
The main differential diagnosis includes angiosarcoma and carcinoma of the breast. Moreover, breast aneurysms, hemangiomas, or various vascular malformations may have misleading appearances on imaging (**Fig. 3.2** and **Fig. 3.3**).



**Fig. 3.1 (a)** Left-sided mammogram shows biopsy-proven mammary angiomatosis (arrows). Moreover, the image on the right shows wire localization prior to wide local excision. (Right image) Wire localization can be used to guide breast surgeons and helps with regard to adequate, complete removal of the mass. This presents as a lobulated and low-density mass. Physical examination revealed palpable and nontender abnormality.

**b**

**Fig. 3.1 (b)** The same patient as in **Fig. 3.1a**. Ultrasound revealed an ill-defined, heterogenous and irregular mass (*arrows*). Please see explanations on **Fig. 3.1c**.



**Fig. 3.1 (c)** The same patient as in **Fig. 3.1a, b**. Mammary angiomatosis on ultrasound. The presented mass is irregular in shape and heterogenous. It contains hyperechoic components peripherally (*long arrows*) and hypoechoic component centrally (*short arrow*).

## Thrombosed Pseudoaneurysm

We illustrate this interesting example of a thrombosed pseudoaneurysm (**Fig. 3.2a, b**).

They may present as mildly pulsatile, growing breast lump. Mammography can show smooth mixed-density oval to round opacity, often with adjacent overlap of vessels (**Fig. 3.2a**).

On sonography, a hypo to anechoic oval breast mass with distal acoustic enhancement was correlated. Color Doppler showed turbulent flow within the mass and a few twigs of radiating vessels from the mass. (**Fig. 3.2c**)

True aneurysms of breast are rarely encountered and need to be differentiated from breast carcinoma (**Fig. 3.2d, long arrow** and **Fig. 3.2e, short arrow**) or angiosarcoma.

## Mammary Hemangioma

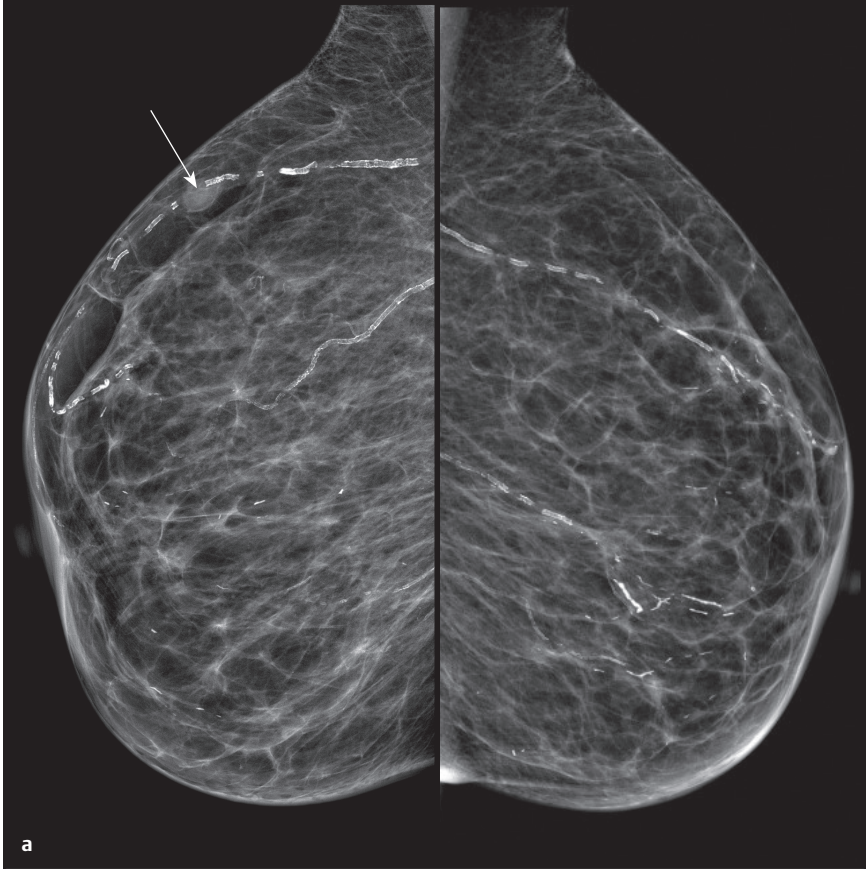
Breast hemangiomas are superficial oval or lobulated masses, often nonpalpable due to soft consistency, and can be broadly described as capillary or cavernous, depending on the dimensions of cystic spaces within the mass. Patients are asymptomatic and these lesions are often detected on screening mammograms.

On mammography, the density may be similar to the fibroglandular component (**Fig. 3.3a, b**). In addition, there may be variable degree of calcification.

On sonography, the masses are oval or lobulated and oriented parallel; however, the internal echotexture of the hemangioma can sometimes mimic a complex cystic malignancy (**Fig. 3.3c**).

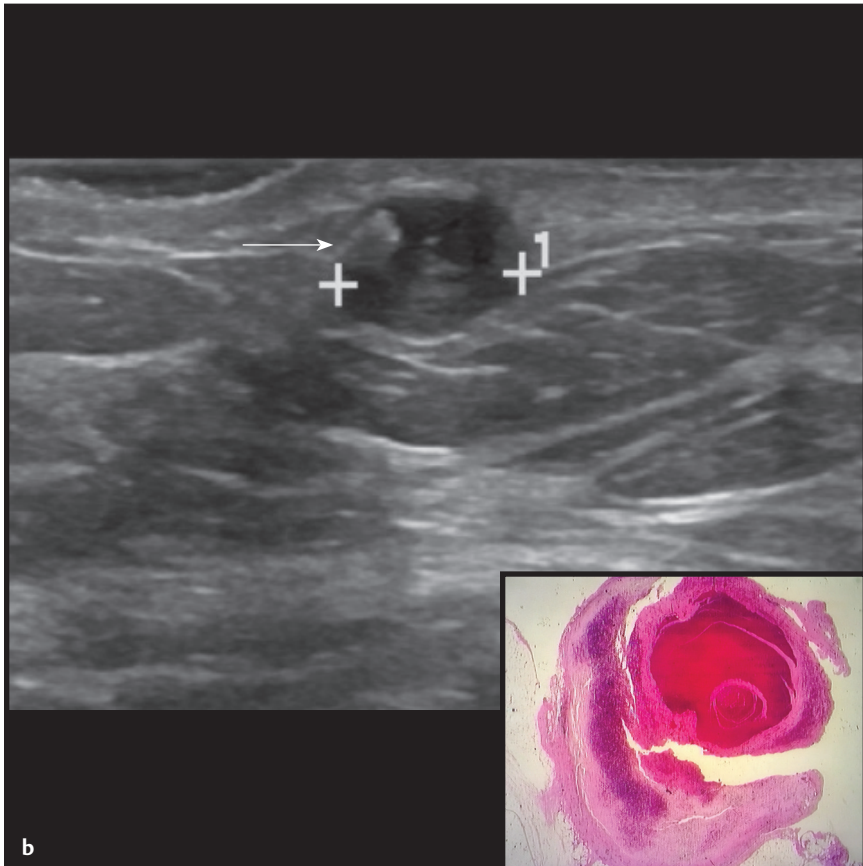
Color Doppler therefore becomes essential in the evaluation of cystic or pulsatile masses where intralesional flow can be detected and spectral Doppler can characterize the pattern of flow—arterial in cases of hemangioma (**Fig. 3.3d**).

These lesions are often biopsied in cases where malignancy is suspected but profuse bleeding postbiopsy may hint toward the diagnosis. Histology of the biopsy samples may show dilated tubular channels filled with red blood cells.



**Fig. 3.2 (a)** An 81-year-old symptomatic female presented with a palpable mass within the upper outer quadrant of the right breast. Bilateral mammography showed a subcentimeter mass (*arrow*).

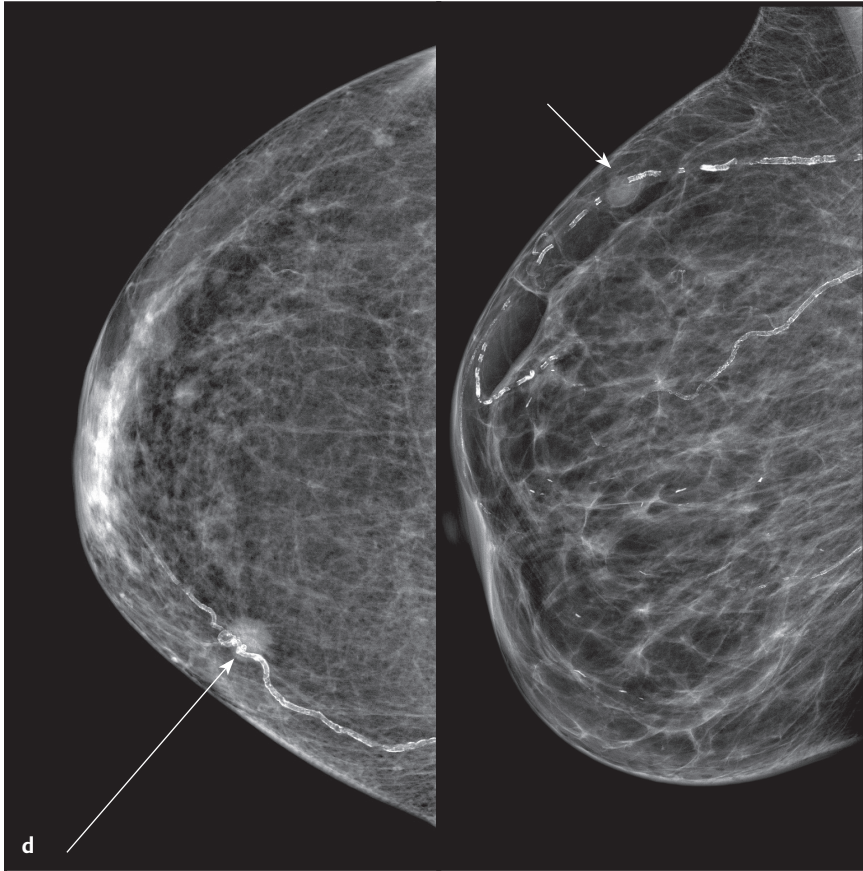


**b**

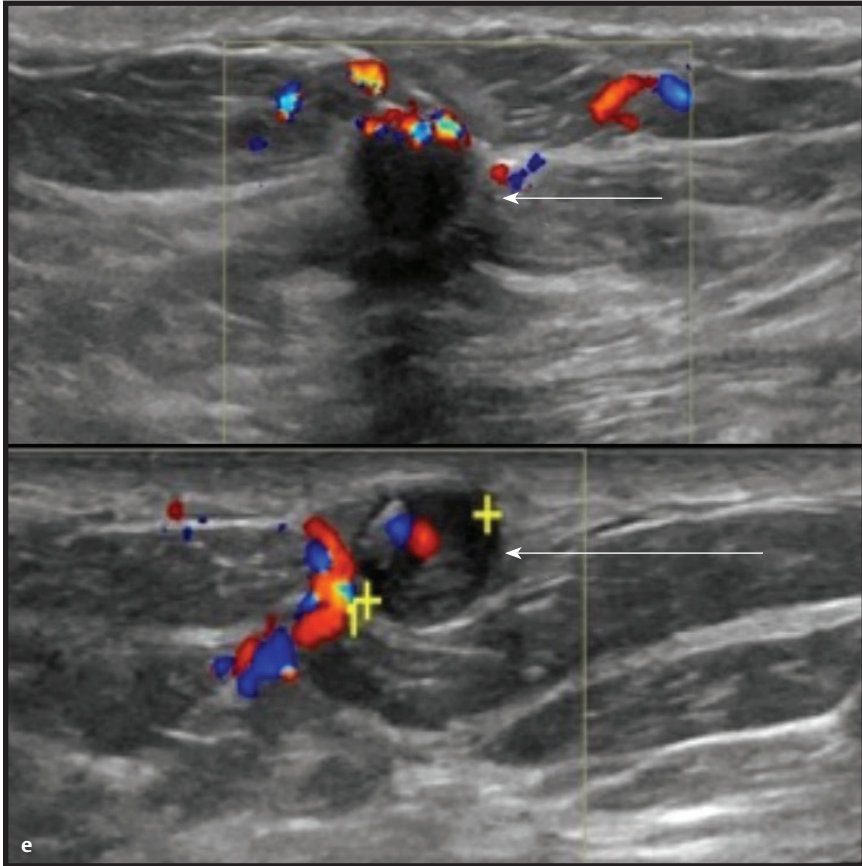
**Fig. 3.2 (b)** The same patient as on **Fig. 3.2a**. Ultrasound shows a hypoechoic lesion with hyperechoic intraluminal thrombus (*arrow*). Microphotograph confirms a benign vascular lesion (*inset*).



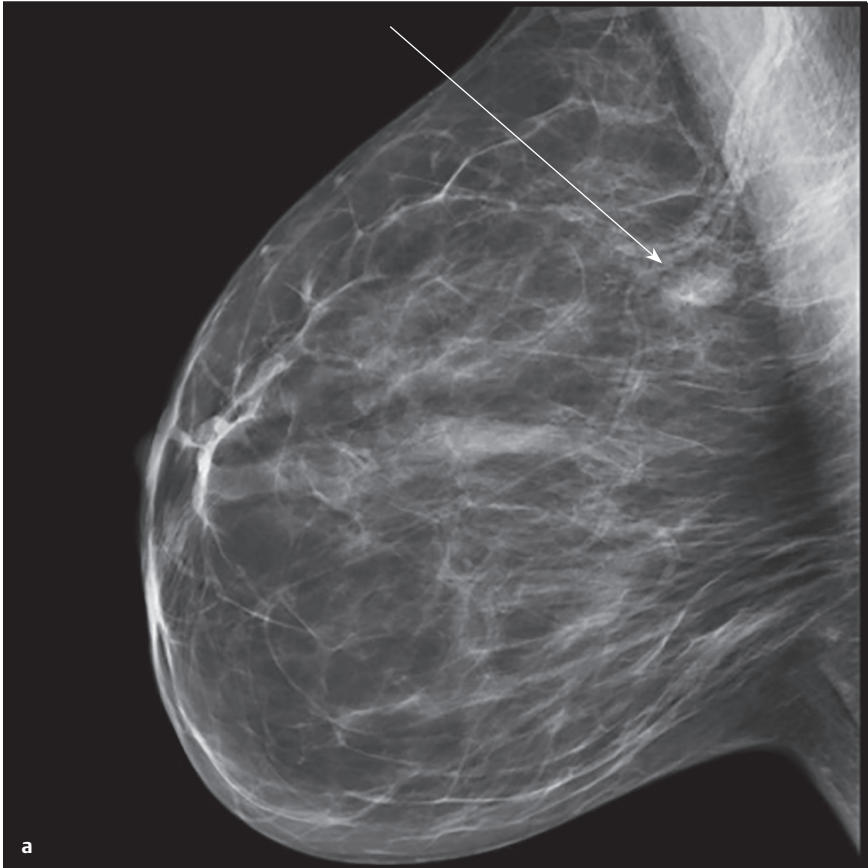
**Fig. 3.2 (c)** The same patient as in **Fig. 3.2a, b**. Color Doppler confirms a vascular lesion with distal acoustic enhancement (*arrow*) confirming the benign vascular origin of the lesion.



**Fig. 3.2 (d)** Comparison slide shows challenges of mammography. Invasive ductal carcinoma of no special type (*long arrow*) and a partially thrombosed aneurysm (*short arrow*). Carcinoma has slightly spiculated margins in contrast to a smoothly marginated aneurysm. However, please note that this is not obvious on mammography, and it is not possible to establish the diagnosis based on imaging alone.

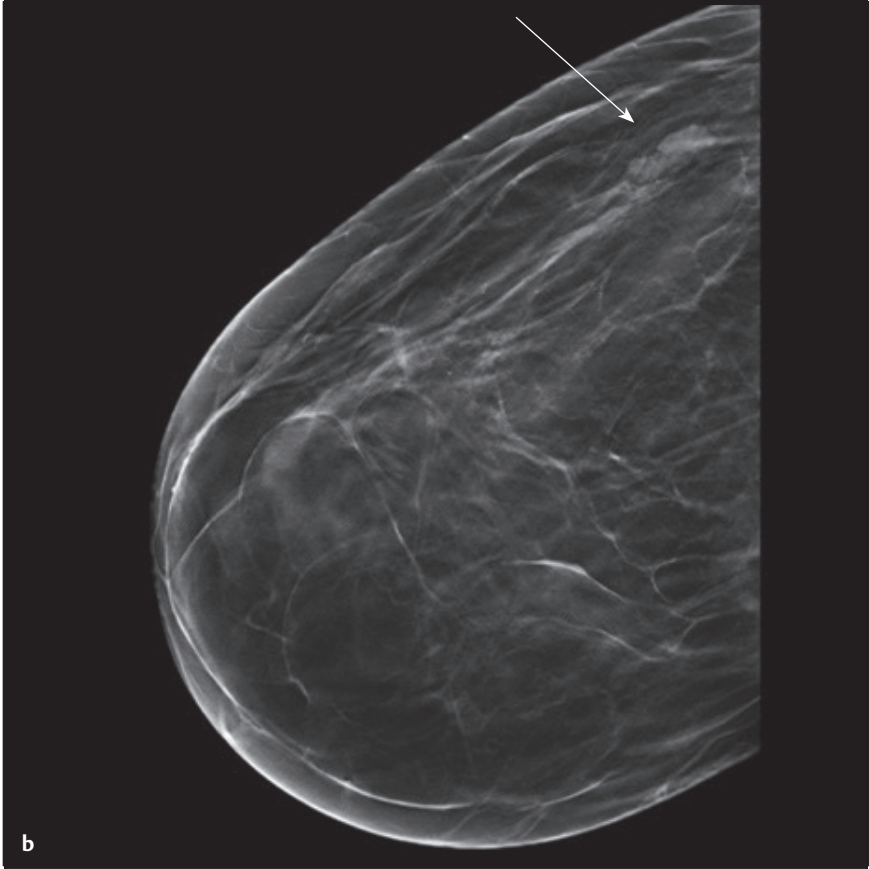


**Fig. 3.2 (e)** Comparison slide shows challenges on ultrasonography. The same patients as on **Fig. 3.2d**. The top image shows invasive ductal carcinoma of no special type (*short arrow*). The bottom image shows a thrombosed aneurysm (*long arrow*). Carcinoma demonstrates posterior acoustic shadowing which is absent in case of the presented benign aneurysm.

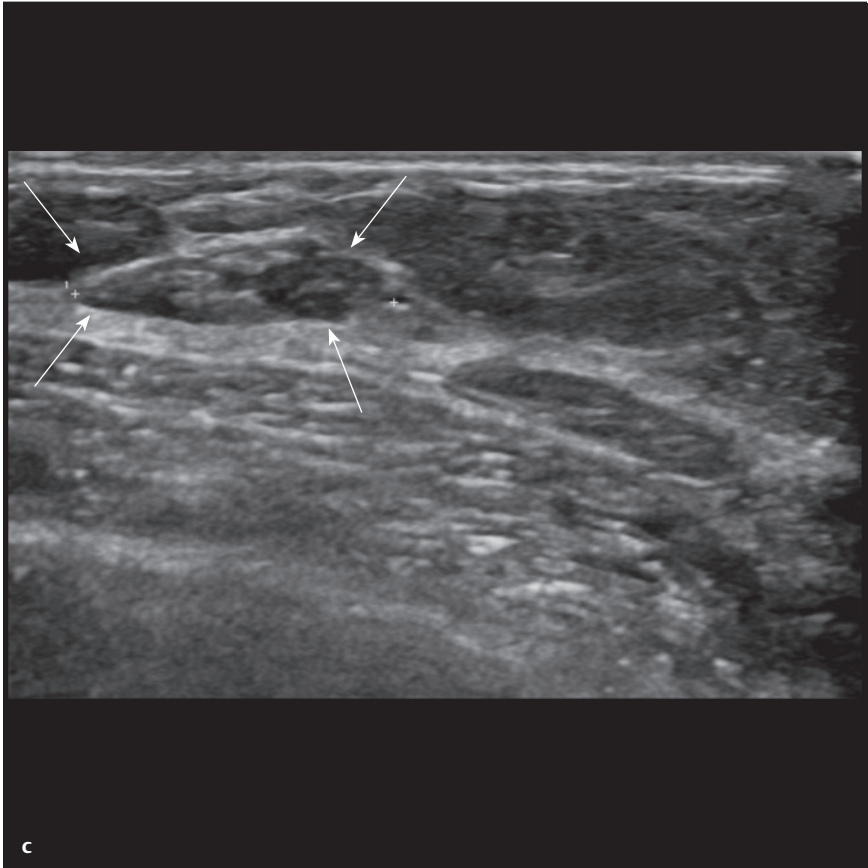


a

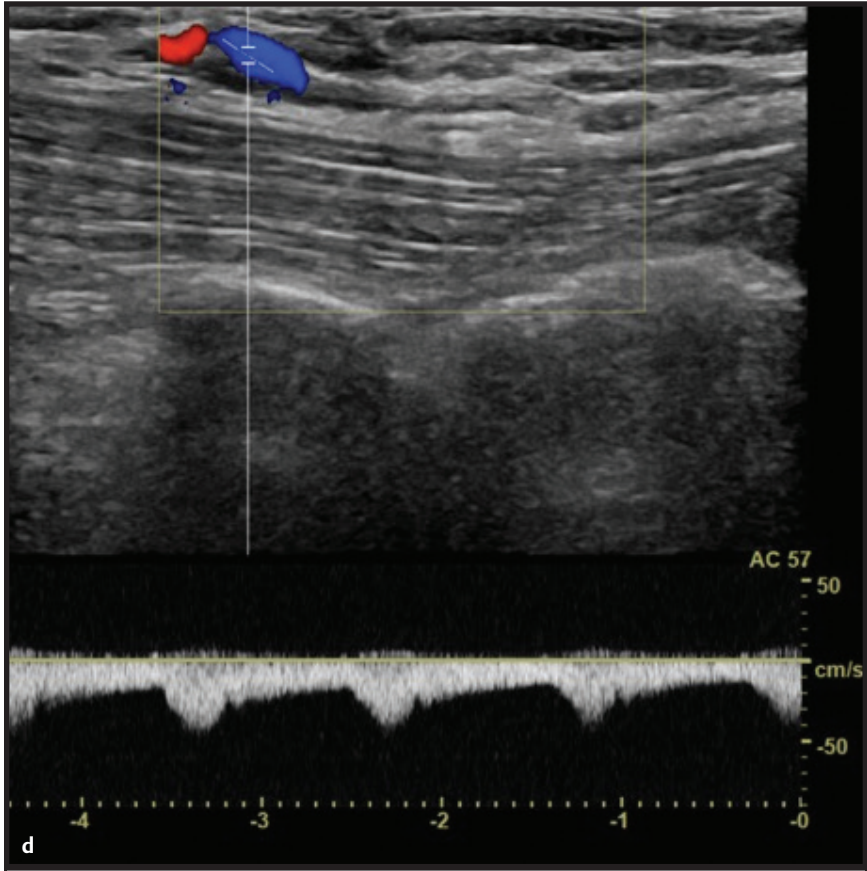
**Fig. 3.3 (a)** A 59-year old female was recalled from screening. Right MLO view showed an ill-defined and low-density mass (*arrow*). It was a painless, nonpalpable mass. Abbreviation: MLO, mediolateral oblique.



**Fig. 3.3 (b)** The same patient as in **Fig. 3.3a**. Right breast tomosynthesis showed a solitary low-density lesion within the upper outer quadrant (*long arrow*).



**Fig. 3.3 (c)** The same patient as in **Fig. 3.3a, b**. Right breast ultrasound revealed a benign vascular malformation. This demonstrated high flow on Doppler. However, the lesion comprised both arterial and compressible venous elements.



**Fig. 3.3 (d)** The same patient as in Fig. 3.3a–c. The lesion demonstrated arterial flow on Doppler.



## Lesions of Neural Origin

Lesions of neural origin in the breast are very uncommon. We illustrate granular cell tumor of the breast, and a case of neurofibromatosis type 1 with associated cutaneous neurofibromas.

### Granular Cell Tumor (GCT)

It is a benign tumor with malignant clinical and imaging characteristics. Patients commonly present with rapidly growing lump which is often tender. This mass originates from Schwann cells and can present as a palpably firm mass on physical examination. Often associated coexistence of prominent axillary lymph nodes may raise the suspicion of locally advanced disease with metastasis. Enlarged lymph nodes can be usually due to inflammatory change or purely coincidental.

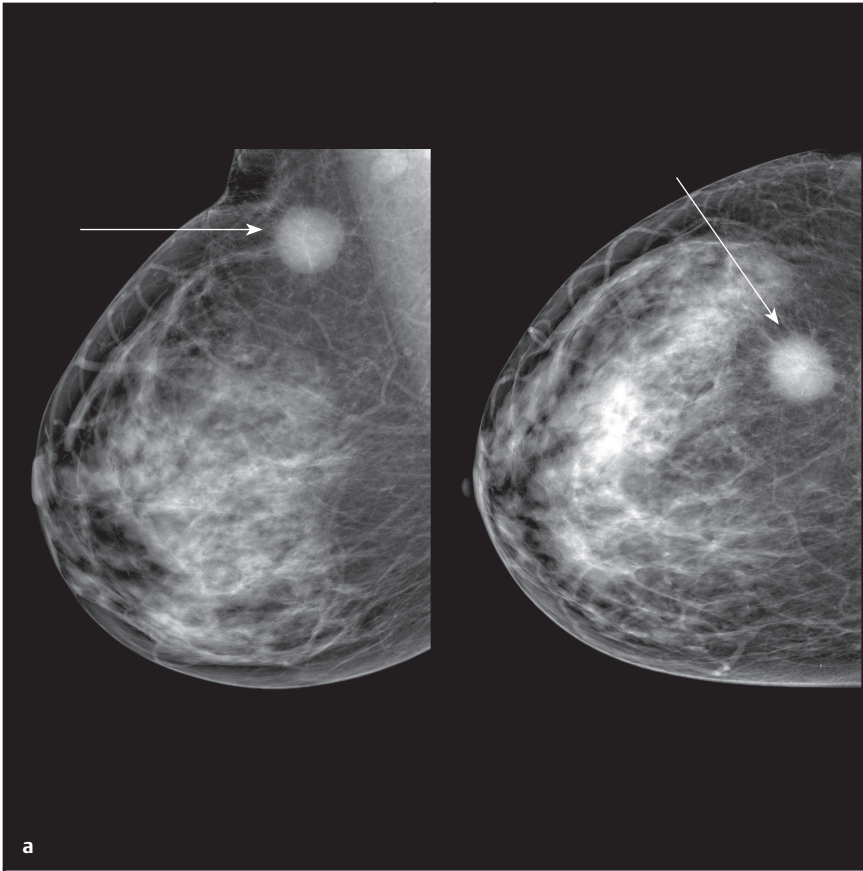
Mammography can show an indistinctly defined oval to round mass with irregular margins. Ultrasound shows hypo to anechoic mass with fuzzy margins.

Imaging features may be indistinguishable from those of a carcinoma and biopsy needs to be performed to exclude a malignant process (**Fig. 3.4a–c**). This entity is benign, but carries a risk of local recurrence, and wide local excision with clear margins is required.

Very rare and limited malignant presentations in the setting of presumed GCT transformations have also been reported.

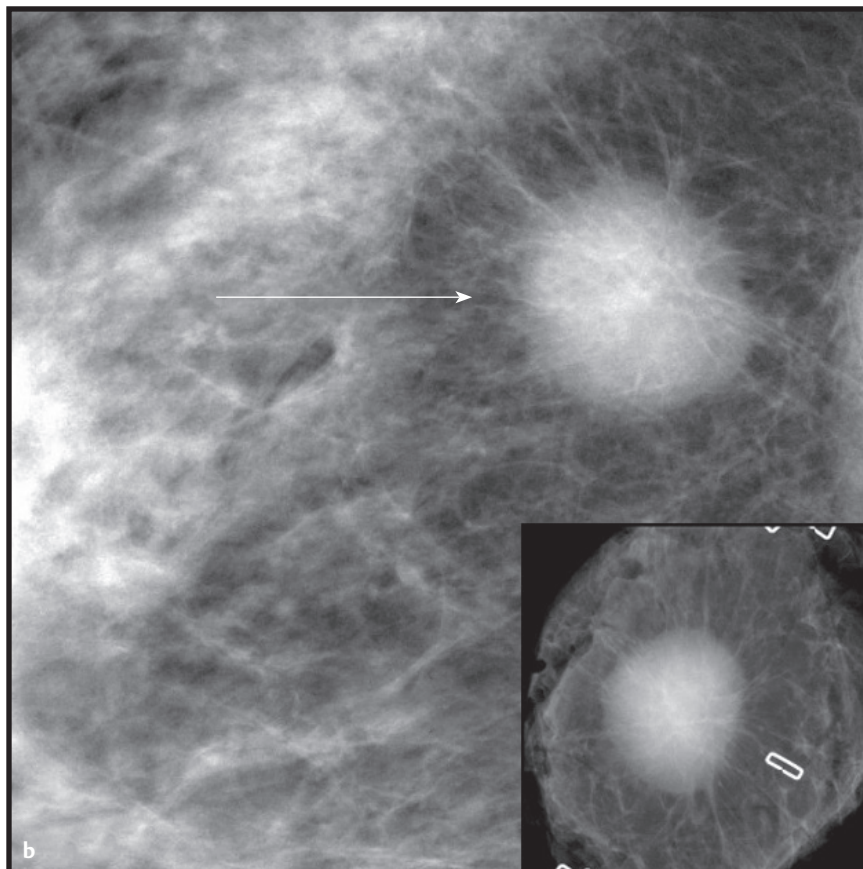
Histology confirms oval spindle-shaped cells and multinucleated giant cells. (**Fig. 3.4c**).

The cells are positive for CD 68 and S100, hence supporting the diagnosis.

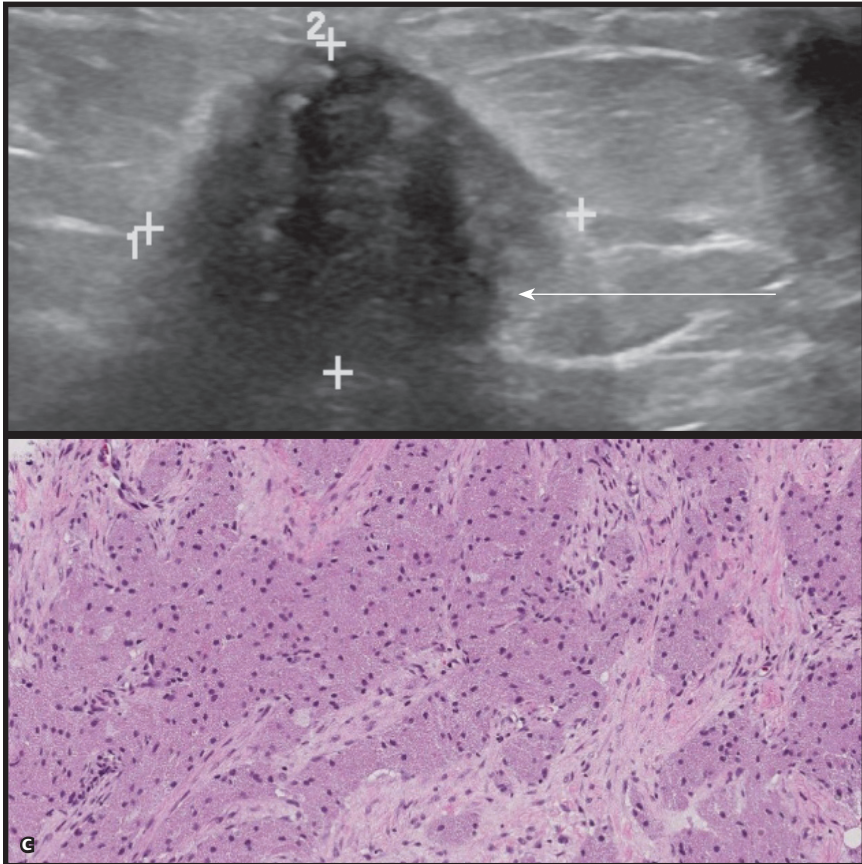


a

**Fig. 3.4 (a)** Right-sided mammogram, MLO view on the left, and CC view on the right. A 46-year old female presented with a palpably hard right breast mass. Mammography showed a well-defined mass within the upper outer quadrant of the right breast and close to the pectoralis muscle (arrows). Abbreviations: CC, craniocaudal; MLO, mediolateral oblique.



**Fig. 3.4 (b)** The same patient as in **Fig. 3.4a**. Right-sided magnification view shows a dense mass with subtle spiculations (*arrow*). Please note wide, local excision specimen with the mass in the central part (*inset*).



**Fig. 3.4 (c)** The same patient as seen in **Fig. 3.4a, b**. Ultrasound showed a hypoechoic and irregular mass (*arrow*). There was associated posterior acoustic shadowing. No skin thickening was observed. Microphotograph shows features of a GCT with granular cytoplasm, small central round nuclei, and small nucleoli. The cells were positive for CD 68 and S100 which supported the diagnosis. There was no evidence of malignancy or atypia such as necrosis, pleomorphism, or more mitotic figures. Abbreviation: GCT, granular cell tumor.

## Cutaneous Neurofibromas

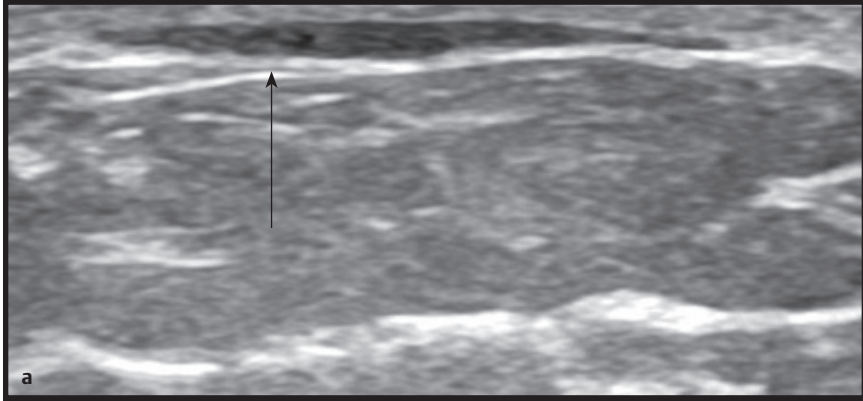
These disorders most commonly present as multiple epidermal nodules.

These can present as palpable breast lumps on physical examination and raise our suspicion of malignancy. Moreover, it is suggested that patients with Von Recklinghausen disease have increased risk of developing a breast cancer.

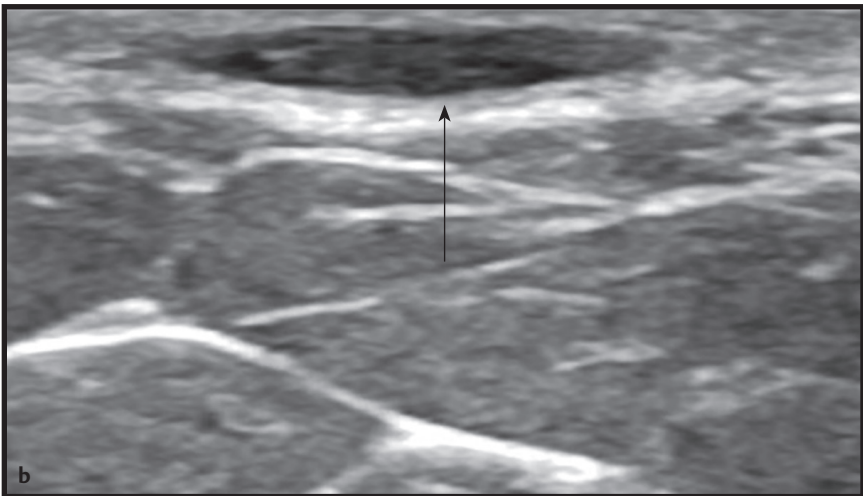
We illustrate high-resolution ultrasound imaging of a male patient with NF1 who presented with a palpable right breast lump in **Fig. 3.5a, b**.

Neurofibromas are round to oval and well-defined hypoechoic lesions with benign features on sonography (**Fig. 3.5a, b**).

Cutaneous neurofibromas are benign tumors and do not require any surgical treatment, unless they grow large or cause cosmetic problems.



**Fig. 3.5 (a)** A 27-year-old male patient with a history of neurofibromatosis type 1 was referred to the symptomatic breast unit with a palpable lump within the upper inner right breast. Ultrasound revealed multiple well-defined, flat, and hypoechoic cutaneous lesions in both breasts (*arrow*). Imaging features were entirely benign in keeping with cutaneous neurofibromas. There were no associated suspicious features. Changes in the setting of the patient's main disease were more prominent on the right side.



**Fig. 3.5 (b)** The same patient as in **Fig. 3.5a**. There were similar benign-appearing cutaneous lesions within the contralateral left breast (*arrow*). These represented multiple neurofibromas in the setting of the patient's known Von Recklinghausen disease.

### Points to Ponder

*Lesions of vascular and neural origin can mimic carcinoma.*

*However, these are very “rare zebras” in breast radiology and it is essential to exclude malignancy at first instance.*

*“When you hear hoof beats, think of horses not zebras.”*

*—Dr. Theodore Woodward*

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- Wegner U, Juette A, Ryan AG, et al. Granular cell tumor (GCT) of the breast: case report of uncommon benign neural tumor with malignant imaging characteristics. *J Med Clin Res & Rev.* 2019;3(1):1–3

## Summary MCQs

### ■ Questions

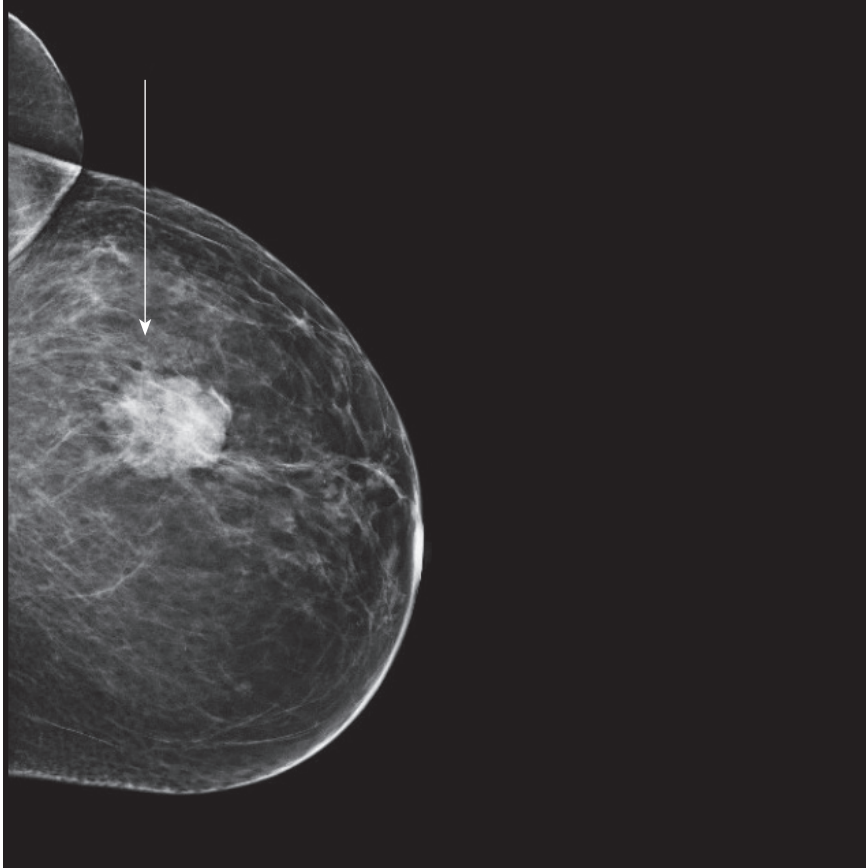
Please choose true or false. More than one answer option may be correct.

**Question 1:** Which of the following statements regarding the presented cases are correct?

- Mammary angiomatosis and granular cell tumor (GCT) carry the risk of local recurrence.
- Lesions of vascular and neural origin may mimic malignancy on imaging.
- GCT is a vascular lesion.
- Mammary angiomatosis originates from Schwann cells.
- Cutaneous neurofibromas have benign sonographic features.

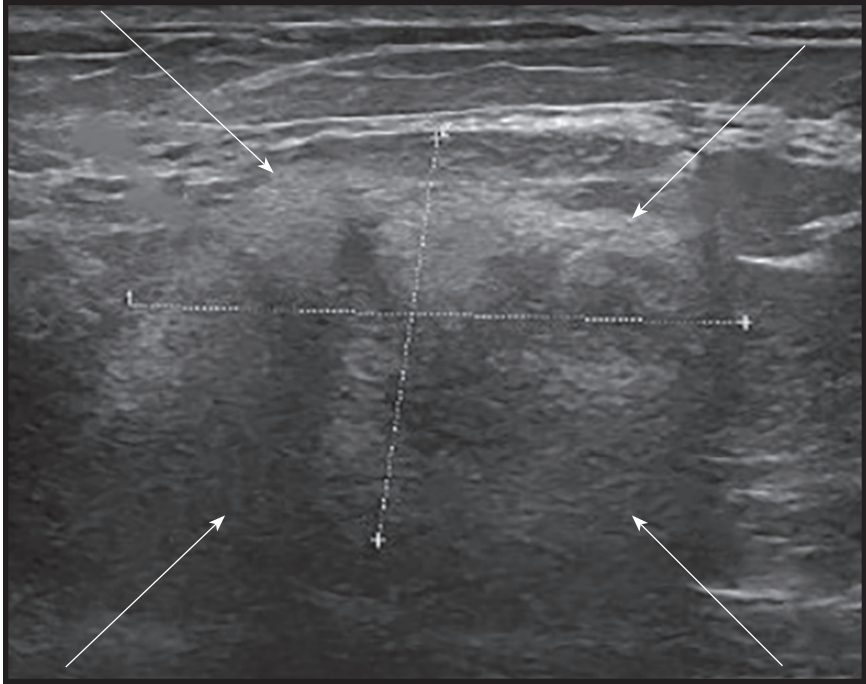


**Question 2:** Mammary angiomas are shown in the following figure. How would you describe the mammographic features?



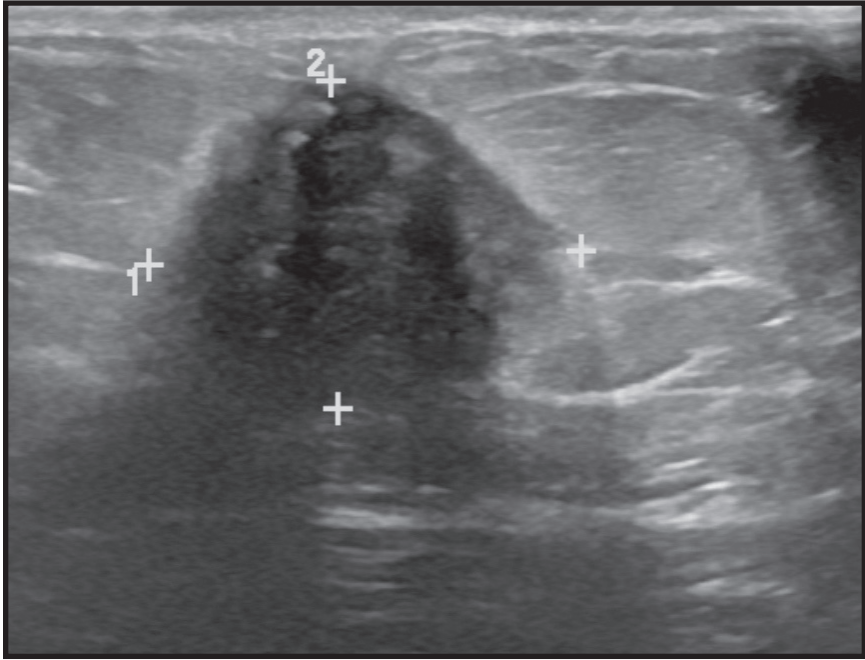
- a) Oval lesion with extensive halo sign.
- b) Architectural distortion.
- c) No significant findings identified.
- d) Microcalcifications.
- e) Irregular mass.

**Question 3:** Angiomatosis of the breast is shown in the following figure. How would you describe the sonographic features?



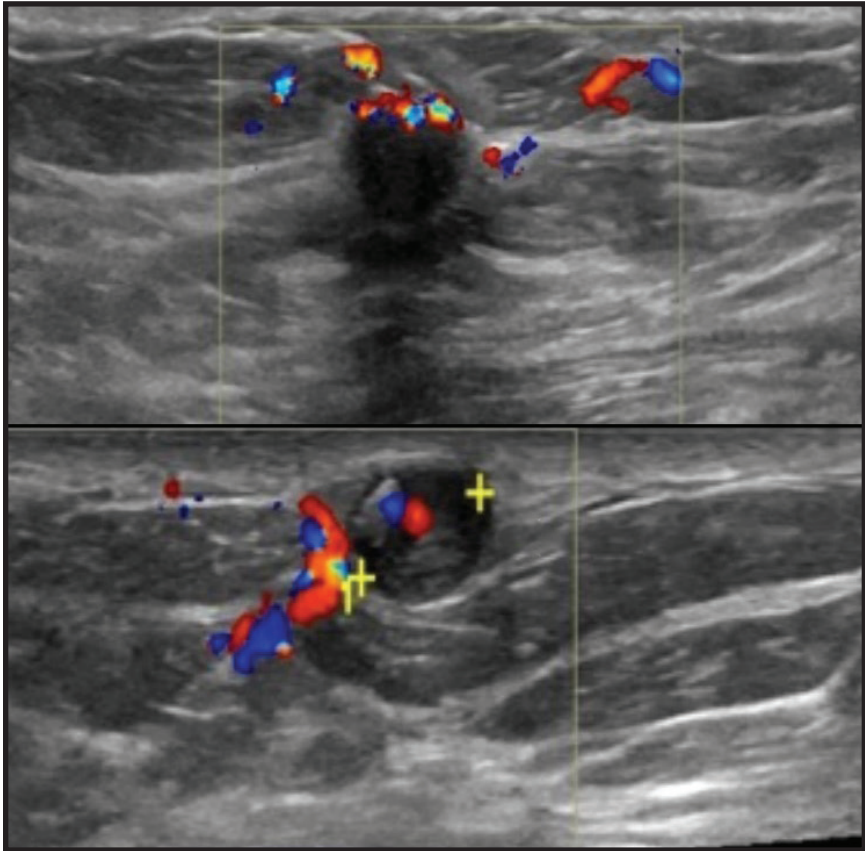
- a) Anechoic round and sharply defined lesion.
- b) Hyperechoic round subcutaneous lesion.
- c) An ovoid well-defined hypoechoic lesion.
- d) An irregular heterogenous lesion.
- e) An ovoid heterogeneous lesion.

**Question 4:** Ultrasound image of a symptomatic female patient with a palpable breast mass is shown in the following figure. What would be the next diagnostic step?



- a) No further management required.
- b) Ultrasound-guided core needle biopsy.
- c) Breast MRI.
- d) Stereotactic biopsy.
- e) Aspiration.

**Question 5:** Sonography images of a carcinoma and thrombosed aneurysm are shown in the following figure. Which image belongs to the patient with an aneurysm and why?

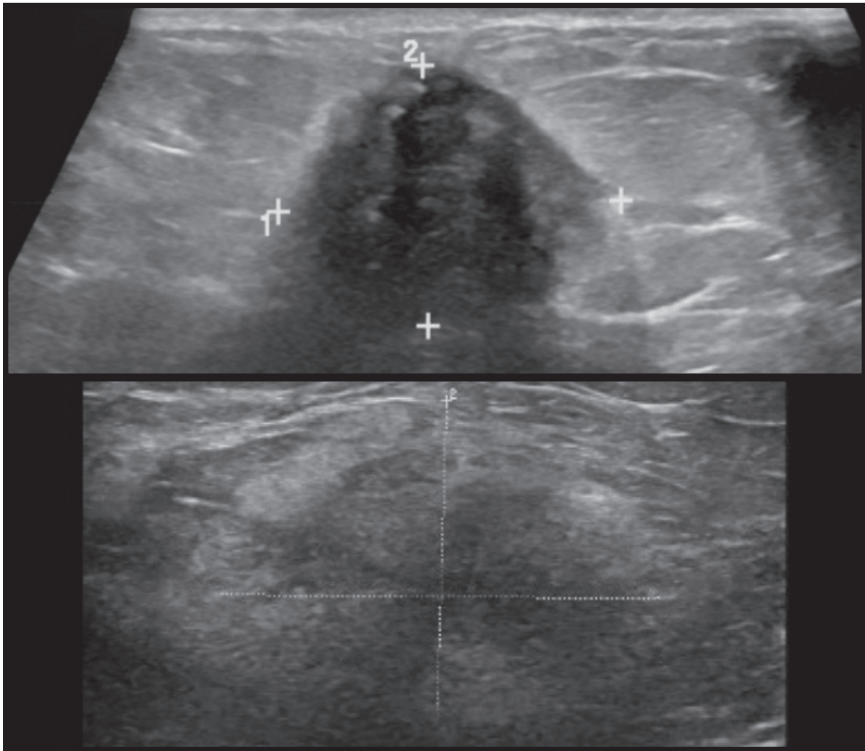


- Top image could represent a benign vascular lesion as it shows typical benign appearances on ultrasound.
- Top image could represent an aneurysm as the mass is taller than wide and irregular in shape, with associated posterior acoustic shadowing.
- Bottom image does not belong to the patient due to its suspicious sonographic appearance.
- Bottom image is related to the patient with an aneurysm as it shows a well-defined, oval mass with intralesional Doppler flow.
- None of the above is correct.

## ■ Answers

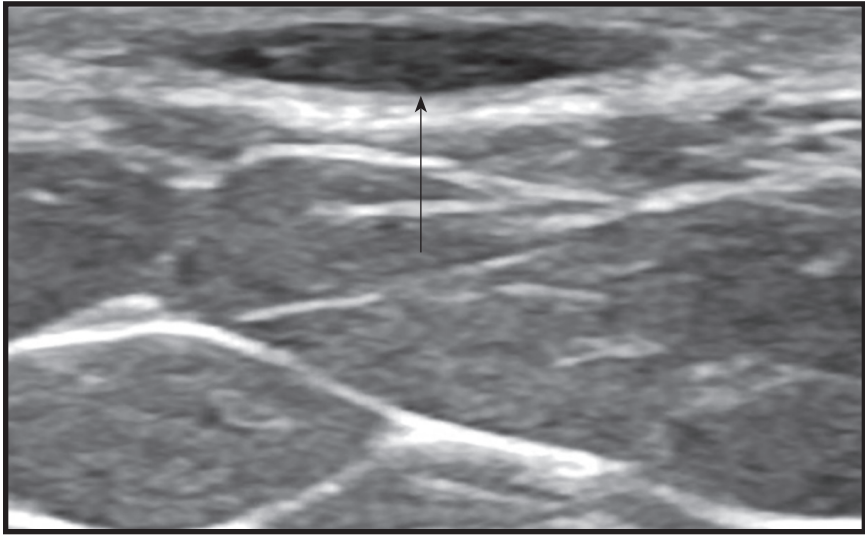
### Answer 1:

- a) (T) Both benign tumors carry the risk of local recurrence, thus wide local excision with clear margins is recommended.
- b) (T) Lesions of vascular and neural origin may mimic malignancy on imaging as shown in the images below:



- c) (F) GCT is a benign neural tumor which originates from the Schwann cells.
- d) (F) Mammary angiomatosis does not originate from the Schwann cells as angiomatosis of the breast is a benign vascular tumor.

- e) (T) Sonographic appearances of cutaneous neurofibromas are benign. Cutaneous neurofibromas presents as round to oval, well-defined, flat and hypochoic cutaneous lesions, as shown in the below image (*arrow*).



## References

- Kopans DB. Breast imaging. 3rd ed. Hagerstown, MD: Lippincott Williams & Wilkins; 2007  
Tabar L, Dean PB. Teaching Atlas of Mammography. 4th ed. Thieme; 2011

### Answer 2:

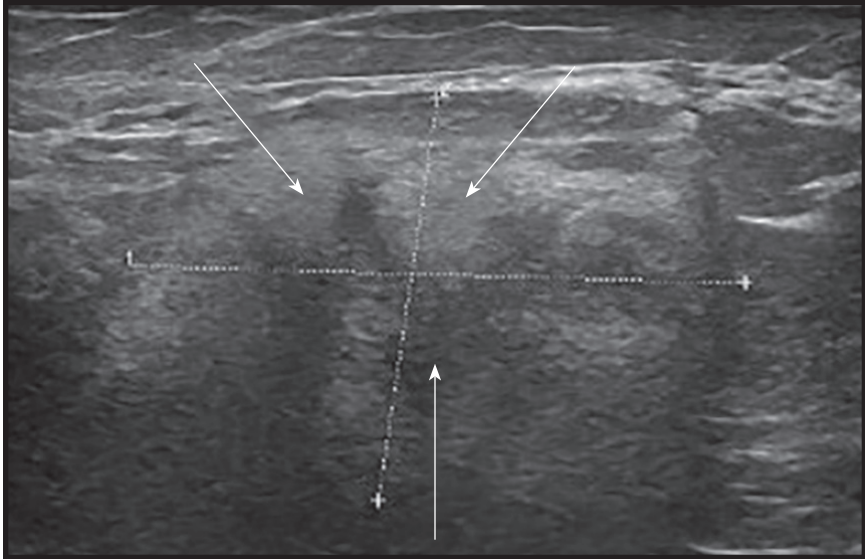
- a) (F) No air outlining or halo sign seen.  
b) (F) No radiating structure with spicules is visualized.  
c) (F) Irregular mass identified.  
d) (F) No calcification is identified.  
e) (T) This mass is irregular in shape.

## Reference

- Tabar L, Dean PB. Teaching Atlas of Mammography. 4th ed. Thieme; 2011

**Answer 3:**

- a) (F) These are features of a simple cyst.
- b) (F) These are features of a typical lipoma.
- c) (F) These are classical features of a fibroadenoma.
- d) (T) An irregular and heterogeneous lesion is identified. See the following figure:



- e) (F) An irregular mass is identified which is not ovoid in shape.

**References**

- Kopans DB. Breast imaging. 3rd ed. Hagerstown, MD: Lippincott Williams & Wilkins; 2007
- Tabar L, Dean PB. Teaching Atlas of Mammography. 4th ed. Thieme; 2011

**Answer 4:**

- a) (F) Biopsy is required to exclude malignancy.
- b) (T) Ultrasound-guided core needle biopsy is the preferred next diagnostic step as microscopic verification is needed to establish the final diagnosis.
- c) (F) A solitary, solid mass on ultrasound is not an indication for MRI.
- d) (F) This could be an option with regard to mammographic abnormality.
- e) (F) This is a solid mass. No drainable collection identified.

## References

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Shah BA, Mandawa SR. Breast Imaging A Core Review. 2nd ed. Philadelphia, PA: Wolters Kluwer; 2017

### *Answer 5:*

- a) (F) Top image represents malignancy as the lesion is irregular in shape and demonstrates posterior acoustic shadowing.
- b) (F) The lesion on the top image demonstrates malignant sonographic characteristics: posterior acoustic shadowing and irregular margins.
- c) (F) Bottom image does belong to the patient with an aneurysm as the lesion appears more benign and has no posterior acoustic shadowing.
- d) (T) This appears benign on imaging and shows well-circumscribed oval lesion without posterior shadowing. Intralesional flow on Doppler application correlates with the final diagnosis.
- e) (F) Option d is correct.

## Reference

Kopans DB. Breast imaging. 3rd ed. Hagerstown, MD: Lippincott Williams & Wilkins; 2007