Case Report

Localized primary breast amyloidosis and 1-year changes in imaging: A case report

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Abstract

Localized primary breast amyloidosis is a very rare benign disease characterized by abnormal protein deposition in the mammary glands. Amyloidosis may mimic the appearance of a number of pathologies, both benign and malignant. Clinically, the patient may present with a breast mass or simply with increased breast density and skin thickening. Herein, we report the case of a 45-year-old woman who presented with a breast mass and was ultimately diagnosed with primary breast amyloidosis, and the mass diagnosed with amyloidosis increased in size and there were a greater number of amorphous and irregular microcalcifications on mammography and ultrasound at the 1-year follow-up. To conclude, we presented changes in a case of localized primary breast amyloidosis on mammography and ultrasound images over a period of 1 year. The current standard of care of primary breast amyloidosis is surgical resection; however, the patient should be followed after surgery to monitor the possibility of recurrence of malignancy.

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Introduction

Amyloidosis is an uncommon disorder characterized by extracellular deposition of amorphous and insoluble proteins in an abnormal fibrillary configuration. It is divided into systemic amyloidosis, in which amyloid deposits form in organs throughout the body, and localized amyloidosis, which is limited to an individual organ [1]. Systemic amyloidosis causes a variety of symptoms, such as fatigue, weight loss, anemia, cardiovascular symptoms (congestive heart failure, arrhythmia), renal symptoms (nephrotic syndrome, kidney failure), gastrointestinal symptoms (malabsorption syndrome, macroglossia, hep-atomegaly, splenomegaly), neurologic symptoms (polyneuropathy, carpal tunnel syndrome, orthostatic hypotension, constipation, diarrhea, dysuria), and bleeding [1]. The examinations used to check for systemic amyloidosis include electrocardiography, echocardiography, blood analysis (renal dysfunction, M protein, free light chain, autoimmune antibody, and chronic inflammatory findings), urine analysis

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Primary breast amyloidosis typically presents with a painless and palpable mass [4]. Radiographic imaging reveals a wide variety of mammographic and ultrasound findings, including similar patterns of ductal carcinoma in situ and fibrocystic changes.

However, to the best of our knowledge, there have been no reports of localized primary breast amyloidosis and changes on mammography and ultrasound images over a period of 1 year, as detailed in this report.

Case report

A 45-year-old woman noticed a hard, nontender lump in the upper inner part of her left breast and visited the outpatient clinic for breast surgery. This mass had been growing slowly for 3 years. She had no family history of amyloidosis or breast cancer and no other past medical or surgical history.

A lump 2 cm in diameter was palpable on the upper inner part of the left breast. The mammogram showed a 3-cm mass with marginal microlobulation and a small round calcification in the upper inner left breast. Additionally, focal asymmetric density was observed on the upper outer side of the left breast (Fig. 1).

Ultrasonography showed an irregular 2.5-cm hypoechoic mass with small calcification in the upper inner left breast. No blood flow was observed entering the mass (Fig. 2A). Ultrasonography showed another indistinct 1.6-cm hypoechoic mass in the upper outer left breast. There was no microcalcification or blood flow in this mass (Fig. 2B).

The patient underwent ultrasound-guided core needle biopsy for the palpable mass in the upper inner side of the breast.

For pathologic diagnosis, hematoxylin and eosin staining showed pinkish amorphous nodular and peribasement membrane deposits with giant cells (Fig. 3A). The specimen was positively stained by Congo red and showed apple-green birefringence under polarizing light microscopy (Fig. 3B). Thus, the mass was diagnosed as an amyloid tumor. The patient had no symptoms suggestive of systemic amyloidosis. We considered

(Bence-Jones protein), nerve conduction test, bone marrow biopsy, and biopsy of sites suspected of amyloid deposition [1]. Despite the various morphologic tissue manifestations, staining for amyloid protein with Congo red reveals a characteristic apple-green birefringence under polarized light microscopy [1]. The precise etiology and pathogenesis of amyloidosis are unknown.

Breast amyloidosis has rarely been reported in the literature; the first report was that of Fernandez and Hernandez in 1973 [2]. Breast amyloidosis can be part of systemic amyloidosis or it may be limited to the breast, which may be a cause of misdiagnosis [3].

Fig. 1 – A marginal microlobulated mass (size: 3 cm, arrowhead) with a small round calcification (white arrow) is observed in the upper inner left breast. Another focal asymmetric density (black arrow) is present in the upper outer left breast on initial mammography. The round radio-opaque density is a skin marker (Left: Craniocaudal view; Right: Mediolateral oblique view).

Fig. 2 – (A) A 2.5-cm irregular hypoechoic mass with a small calcification (arrow) is observed in the upper inner left breast. (B) Another indistinct hypoechoic mass (size: 1.6 cm) found in the outer left breast. There was no microcalcification in this mass.
further therapy to be unnecessary and annual follow-up was recommended.

At the 1-year follow-up, the mass diagnosed with amyloidosis increased in size (from 3.0 cm to 3.3 cm) and there were a greater number of amorphous and irregular microcalcifications on mammography and ultrasound (Fig. 4).

There was no change in shape or size of the other mass in the upper outer left breast on mammography and ultrasound.

To make a definite diagnosis, mass excision was performed. Histopathology examination revealed dystrophic calcifications and amorphous eosinophilic material deposition; the infiltration of periductal lymphocytes and multinucleated giant cells was revealed by hematoxylin and eosin staining. Spotty ossification was newly noted at this time (Fig. 5). The deposits showed apple-green birefringence on Congo red staining under polarized light microscopy. Immunohistochemistry for AA amyloid was negative. No malignancy such as breast carcinoma, lymphoma, or plasma cell myeloma was seen.

The patient is currently being followed-up at our institution and has shown no evidence of recurrence of breast mass at 1 year after surgery.

Discussion

Amyloidosis is the deposition of amyloid in the organs’ connective tissue framework. It is divided into systemic and localized forms according to the disease of the disease and can be classified as primary or secondary according to the etiology. Amyloidosis can also be classified as AA or AL based on the chemical composition. In clinical practice, the most commonly diagnosed form of amyloidosis is primary idiopathic amyloidosis (AL type) [5,6]. Breast amyloidosis typically appears as diffuse breast involvement as part of systemic amyloidosis with the primary AL rather than AA type [6]. A less common manifestation is isolated breast involvement that occurs as a localized form of the disease [5,6].

The clinical course tends to be benign, and patients typically complain of hard, nonpainful, and palpable masses in the affected breast [7].

Previous studies have reported that common mammographic findings of breast amyloidosis include a variety of solid shapes or multiple masses or nodules, with or without calcifications [8]. This was the first reported case of localized
primary breast amyloidosis with changes on mammography and ultrasound images over a period of 1 year.

The patient had no symptoms indicative of systemic amyloidosis; therefore, we considered examination for systemic amyloidosis to be unnecessary and diagnosed the patient with localized amyloidosis. If the patient develops systemic symptoms, she should be checked for systemic amyloidosis.

Since the amyloid breast tumor and breast cancer findings may be similar, pathologic diagnosis before surgery is important to avoid excessive invasiveness and unnecessary surgery. If deposits of nonstructural substances are observed by hematoxylin and eosin staining, Congo red staining should be added for confirmation.

In breast amyloidosis, the amyloid is histologically evident as depositions at the periductal, interstitial, or perivascular spaces with multiple multinucleated giant cells and calcifications. Amyloid fibrils have an affinity for calcium and deposition is found around the mammary ducts and blood vessels [5]; thus, this pathophysiology is strongly correlated with the mammography-evident branching or linear distribution of microcalcifications deposited in or around the vasculature or mammary ducts.

The current standard of care of primary breast amyloidosis is surgical resection; systemic therapy is not necessary. Localized primary breast amyloidosis has a good prognosis; however, the patient should be followed after surgery to monitor the possibility of recurrence of malignancy.

To summarize, we reported a case of localized primary breast amyloidosis with changes on mammography and ultrasound images over a period of 1 year.

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