Case Reports

Cystic Breast Lymphangioma

S. N. Waqar, H. Khan*, S. F. Mekan, N. Kayani**
Departments of Biological and Biomedical Sciences, Surgery* and Pathology and Microbiology**, The Aga Khan University, Karachi.

Introduction

Cystic lymphangioma of the breast is a rare benign lymphatic tumor. Lymphangiomas consist of dilated lymph channels lined by endothelium. Possible etiological factors include blockage of the lymphatic channels with secondary dilatation, congenital weakness of the lymphatic wall or proliferation of lymphatic vessels. These lesions enlarge over time by the collection of fluid and the budding of pre-existing spaces. These cystic spaces communicate with large dermal lymphatics and subepidermal vesicles, but not with the regional lymphatic system.

The morphology of the lesions varies according to the character of the surrounding connective tissue. They may range from cysts in loose areolar tissue to superficial vesicles in dense fibrous tissue, features of both may coexist.

Superficially they appear as plaques of grouped, deep-seated, thick-walled vesicles with a warty appearance and which may contain hemorrhagic fluid. New crops can develop in areas of previously unaffected skin. Histopathologically, hyperplasia, hyperkeratosis, and papillomatosis of the epidermis overlying the vesicles are seen. These vesicles are painless in contrast to the painful vesicular eruption of herpes zoster, which follow a dermatomal distribution, having a short clinical course. Tzanck smear, direct fluorescent antibody, culture, and biopsy would be useful in the latter. We present a case of a 24-year-old woman with a congenital right breast mass diagnosed as a cystic breast lymphangioma. The development and clinical presentation of breast lymphangiomas is discussed with an emphasis on rational approach to diagnosis and treatment of these difficult lesions.

Case Report

A 24-year-old lactating Afghani woman presented with a 1 month history of high grade fever, chills and rigors
along with painful enlargement of a pre-existing right breast lump. The lump had been present since birth and had gradually enlarged over time.

On examination, the right breast and axillary tail were diffusely enlarged, appearing to be approximately three times the size of the other breast. On palpation the mass was firm, nodular and mobile with respect to underlying tissues but attached to overlying skin which was edematous (peau d’orange) with prominent veins. On the under surface of the breast were grouped thick-walled vesicles, not conforming to any dermatomal distribution. Some of these vesicles contained brownish hemorrhagic fluid. Preliminary diagnosis of a congenital breast mass with superimposed acute mastitis was made.

The complete blood count was within normal limits and serial blood cultures were negative. Ultrasound of the right breast showed heterogenous echotexture, with irregular, multiple cystic areas and increased soft tissue vascularity. No abscess collection was seen. MRI further revealed a mixed solid and cystic lesion infiltrating the breast parenchyma. Incisional biopsy revealed cystic lesions, lined by flat endothelium, with lymphocytes in the interstitium, consistent with the diagnosis of lymphangioma.

She was given analgesia, appropriate antibiotic therapy and lactation suppression. The mass was surgically resected after management of the acute mastitis.

The breast specimen of the patient measured 25x20x7cm. The skin surface had multiple nodular projections and the cut surface revealed irregular cystic areas with spongy appearance, involving the whole tissue. Skeletal muscle was attached to the deep margin. Microscopic examination revealed a neoplastic lesion composed of dilated lymphatic channels lined by a single layer of flattened cells. These were separated in areas by breast parenchyma. Lymphoid aggregates were seen.

Discussion

Cystic lymphangioma of the breast is a rare benign lymphatic tumor; only 10 cases have been reported in the past 30 years. Clinically, most are evident before the age of 2 years. Rarely, they may be detected later on in adolescence or adulthood on incidental radiological screening. In this case, lymphangioma was diagnosed on the basis of clinical, radiological and histopathologic evidence. Rare cases of breast lymphangiomas have also been discovered on routine screening mammography. Ultrasound is more useful in young women with dense breasts and differentiates solid from cystic masses. MRI demonstrates accurately the depth and extent of the tumor which would dictate therapy. However, the confirmatory diagnosis of lymphangioma is made on the basis of histopathology.

Lymphangiomas are treated for cosmetic and functional reasons. Failure to treat may result in secondary complications of infection and hemorrhage. Malignant degeneration into squamous cell carcinoma in a long-standing lesion, and lymphangiosarcoma at the site of irradiated lymphangioma circumscriptum have been reported.

Treatment options are many, with varying success and recurrence rates. The choice depends on depth and location of the lesion along with the patient's age and general health. Steroid injections and sclerosants are losing popularity owing to high rate of recurrence. In such cases, subsequent surgery becomes technically difficult owing to sclerosed tissue. Carbon dioxide laser yields functionally and cosmetically acceptable results by vaporizing some of the surface lymphatic vessels. Streptococcal lysin (OK-432) is a biological response modifier which induces local inflammatory cytokines that increase endothelial cell permeability, lymph drainage and flow leading to shrinkage of the cystic spaces. It can be used alone or to make the lesion more amenable to surgery. Wide surgical excision of the cysterns facilitated by MRI is the most effective treatment modality, which was also the treatment of choice in this patient. For poor surgical candidates, radiotherapy is the option considered.

References