Clinical case: Giant cystic lymphangioma of the breast. Report of a case with 20-year follow-up and review of the literature

ABSTRACT

Background: Lymphangiomas are congenital or acquired malformations (secondary to trauma, infection or neoplasm) in the mammary gland and are extremely rare. These lesions tend to infiltrate surrounding tissues and malignant degeneration is extremely rare. Lymphangiomas clinically manifest as benign, slow-growing masses that are diagnosed clinically and by imaging studies. Surgery with removal of the mass is performed for aesthetic reasons and to make a differential diagnosis with other common injuries.

Clinical case: We present the case of a 45-year-old female with progressive increase in the size of the left breast. She reported breast trauma concerns and there were no other symptoms. Histological diagnosis was giant cystic lymphangioma of the left mammary gland.

Discussion: Lymphangiomas are uncommon lesions and are extremely rare in the mammary gland. They may be locally aggressive and are benign where abnormal lymphatic tissue has some ability to proliferate and accumulate large amounts of liquid, representing a cystic appearance as presented in our case. Local surgical excision is the treatment.

Conclusion: This is the first case of giant breast cystic lymphangioma reported in Mexico, which corroborates the benign nature of the lesion.

Key words: Mammary gland, benign breast tumors, lymphangioma.
INTRODUCTION

Lymphangiomas are congenital or acquired malformations (secondary to trauma, infections or neoplasms) that occur as a tumor of varied aspect formed from embryonic cells of the ducts of the lymphatic system. In 20 to 40% of the cases they are associated with chromosomal abnormalities, various aneuploid cells or malformations. Lymphangiomas of the mammary gland are very rare, with only a few cases being reported in which the size of the lesions varies from 3 to 25 cm in diameter. They are mainly located in the upper outer quadrant of the breast gland. This distribution pattern is related to the lymphatic drainage of the breast, which is mainly towards the Spence’s tail and armpit. Lymphangiomas are composed of dilated lymphatic vessels lined by endothelium and >50% of these lesions exist from birth; 90% appear in the second year of life. These lesions do not expand very quickly but tend to infiltrate into surrounding tissues. Malignant degeneration is very rare. Lesion size increases after birth and invades adjacent structures usually with cavernous spaces lined with endothelial and lymphoid cells. Pathologically, they are lymph fluid-filled cavities of three types: capillary, cavernous and cystic (macrocystic with cysts >2 cm, otherwise microcystic). There are mixed forms and those associated with high-flow vascular lesions. They are divided into three histopathological subtypes: capillary—composed of fine capillary walls, cavernous—composed of dilated lymphatic compounds and cystic spaces—dilated cysts through an endothelium layer of variable size.

Clinically, lymphangioma manifests as a benign, white, slow-growing tumor, causing aesthetic or functional impairment. Its volume is increased due to infection or bleeding when coexisting with vascular lesions. It is diagnosed clinically and by imaging with ultrasound, computed tomography and magnetic resonance. To define the lesion and its extent, fine needle aspiration biopsy (FNAB) should be carried out to distinguish this from any cysts, lipomas or other vascular malformations.

Treatment is not entirely standardized and depends on size and location. Small, non-growing lesions that do not affect organ function are observed. With larger lesions, surgical removal is indicated. One recurrence during pregnancy and lactation has been reported. Surgery is carried out for cosmetic reasons and for differential diagnosis with other more common lesions. The surgical procedure involves tumor removal and other therapeutic options include radiation and sclerotherapy, but these are ineffective.

CLINICAL CASE

We present the case of a 45-year-old female patient whose evolution began in January 1992, 3 years prior to her medical consultation. There was an increase in the size of the left breast, which became progressively more severe 6 months before admission. She reported having experienced trauma to the breast without reporting the date. She had no other symptoms. Family history was clinically nonsignificant. Past medical history included viral hepatitis with no complications. The patient was treated for renal tuberculosis and she is under medical control for hypertension. She had an abdominal hysterectomy due to leiomyomas at the age of 43. Physical examination revealed significant breast asymmetry with increased tension of the left breast three times greater than the contralateral breast (Figure 1). There is no palpable tumor, and only the visible venous network in the mammary skin is observed. Regional axillary nodes were not palpable. The remainder of the examination was without significant findings. Clinical diagnosis was a large breast cyst. Ultrasound reported a multilobulated lesion and mammography reported only increase in size and extremely dense breasts. Based on these findings, FNAB was performed in the affected breast, and initially 1200 cc of serosanguine-
ous material was removed (Figure 2). Cytology was negative and inflammatory and FNAB was negative. On another occasion, 500 cc of the same liquid was aspirated, and a cystic nodule of 20 × 20 mm was palpated and not attached to deeper layers. Based on the above, we performed a surgical excisional biopsy. Diagnosis was breast cancer clinical stage T2,N0M0. A multilobulated septate surgical specimen was removed and sent to pathology. Macroscopic report of the tumor was 18 × 10 cm in diameter, 600 g in weight, and was soft and nodular with multiple septated cysts. Serial sections of the tumor surfaces were grayish-white with dark brown multifocal areas (Figure 3). The microscopic appearance of the tumor was composed of numerous cavernous cysts with dilated cysts lined by flattened endothelial cells with eosinophilic bright lymphatic fluid. Dilated lymphatic channels in the stroma were fibrous prominent. There was no stromal infiltration by lymphocytes. Around the dilated lymphatic channels, areas of adipose tissue and normal lobules of the mammary gland were observed. Surgical margins were free of tumor. Definitive histological diagnosis was giant cystic lymphangioma of the left mammary gland (Figure 4).

The patient has remained under clinical management for the last 20 years. She has been treated during this time due to ovarian cancer and lipoma of right shoulder. The patient is alive without recurrence or tumor activity (Figure 5).

**DISCUSSION**

Lymphangiomas are infrequent and extremely rare lesions in the mammary gland, with a locally aggressive behavior but are benign. Some
authors consider lymphangiomas to be hamartomatous malformations arising from the inability of the lymphatic system to communicate with the venous system. Another point of view is that lymphangiomas represent sequestered lymphatic tissue that cannot normally communicate with the lymphatic system. According to the latter two scenarios, abnormal lymph tissue has some ability to proliferate and accumulate large amounts of fluid, which represents its cystic appearance, as presented in our case.

Lymphangiomas are classified into three types: capillary, cavernous and cystic. However, the distinction between cavernous lymphangioma and cystic lymphangioma is arbitrary. Both can develop in the same lesion, indicating that the lymphangioma cyst is simply a cavernous lymphangioma of long duration in which the cavernous spaces are large as in the case presented.

Most lymphangiomas are located in the upper outer quadrant of the breast related to the drainage pathways of the breast’s lymphatic system. They are described as well-defined and smooth lesions as observed in our patient. Imaging studies with ultrasound, magnetic resonance and mammography support the clinical diagnosis and its surveillance, but the final diagnosis was based on clinical, imaging and histopathological findings.

Histologically, numerous small spaces containing amorphous eosinophilic liquid and some

**Figure 3.** Surgical specimen of 18 × 10 cm in diameter with a weight of 600 g, soft consistency with multiple cysts.

**Figure 4.** Frontal view of the appearance of the mammary gland at 3 months after surgery.

**Figure 5.** Frontal view of the appearance of the mammary glands at 20 years of monitoring.
lymphocytes were identified and are surrounded by a monolayer of decreased endothelial cells. Some lymph nodes may contain blood due to secondary hemorrhage as in the case reported here and may be misdiagnosed as a cavernous hemangioma. However, large collections of lymphoid cells in the stroma, sometimes with formation of lymphoid follicles and the relatively greater irregularity of the cavernous spaces, confirmed the diagnosis of lymphangioma. Some vascular endothelial markers such as factor VIII and the associated antigen to CD31 may be positive in the lymphatic endothelium but do not distinguish a hemangioma from lymphangiomas. Laminin can be expressed in the basal lamina of the lymphatic vessels but is better expressed in the blood vessels and makes a clear distinction between the two structures. The differential diagnosis is with lymphangiomatosis, which only occurs in women and is located in lymphatic vessels of the mediastinum, retroperitoneum and lung parenchyma. It is characterized by smooth muscle proliferation different from that of the affected lymph nodes. Smooth muscle cells express melanocytic markers. Another differential diagnosis is with lymphangiectasia, which appears in post-surgical patients or with radiation therapy for cancer patients and in morbidly obese persons secondary to the weight of the great folds of fat that cause lymphatic obstruction. Local surgical excision is the treatment indicated, similar to that of other parts of the body as reported in other case reports as well as in our case reported.

In conclusion, we have presented the first documented case in Mexico of a giant cystic breast lymphangioma reported in a 45-year-old patient with longer follow-up and evolution of the disease reported in the literature, which confirms the benign nature of the lesion.

REFERENCES