

MASSIVE CAVERNOUS LYMPHANGIOMA OF THE BREAST AND THORACIC WALL: CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Lymphangiomas are benign lesions but are associated with high morbidity when they become very large, occur in critical locations, or when surgically removed, develop secondary wound infections. Almost all lesions require surgical treatment. Complete excision is curative; however, relapses must be anticipated with incomplete excision. We report the case of a patient with a long history of massive cavernous lymphangioma of the breast and thoracic wall extending into the axilla in whom complete excision was not possible.

Keywords: breast, cavernous lymphangioma, hemangioma, cystic lesions, vascular neoplasias, imaging, treatment

The differential diagnosis of cystic lesions of the breast encompasses simple cysts, post-operative seromas, haematomas, lymphoceles, galactoceles, abscesses, hemangiomas and, very rarely, lymphangiomas.

Lymphangioma

Cystic lymphangiomas or cystic hygromas are a congenital malformation of the lymphatic system. They result from sequestration of the lymphatic tissue, which no longer interacts normally with the surrounding lymphatic system (1). Histo-

logical examination reveals multiple protrusions lined with thin endothelium. They are generally located in the head, neck area (dorsally or laterally; 75%), the axilla (25%), mediastinum or, more rarely, in the retroperitoneum, abdominal organs, skeleton or scrotum. Cystic lymphangiomas are most commonly diagnosed in young children. 50–65% of lymphangiomas are present clinically in the newborn, and 90% are apparent by the age of 2 years (2). Lymphangiomas are generally cavernous lesions and are very rare in the breast, especially in adults. Secondary lymphangiomas after mastectomy and subsequent irradiation of the thoracic wall have been described (3) and only isolated cases have been reported in the literature (4-7).

Hemangioma

Rosen et al detected 1.2% benign hemangiomas in a series of 550 mastectomy patients (8). Benign vessel lesions of the breast are divided into 4 categories: perilobular hemangiomas, angiomatosis, venous hemangiomas and hemangiomas involving the subcutaneous breast tissue (9). Hemangiomas exhibit no arteriovenous shunts, endothelial proliferation or atypical characteristics. It is important to exclude low grade angiosarcoma (10). The vast majority of vascular neoplasms of the skin and soft-tissue layer are clinically benign tumors and



Fig. 1. Preoperative gross anisomastia and left macromastia.

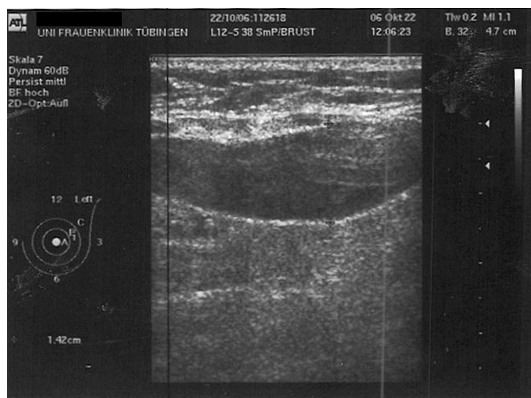


Fig. 2. Ultrasound of the outer quadrant of the left breast displaying multiple vessel conglomerates with a diameter of up to 2 cm with a smooth border.

mostly coincidental microscopic findings. Malignant vascular neoplasms, such as angiosarcoma, belong to the rare soft tissue sarcomas and comprise less than 1% of tumors in this class (11). Complete excision is recommended for all vascular lesions of the breast.

CASE HISTORY

We report the case of a 43-year old woman (grava 2, para 2) with a 10-year history of increasing pain and enlargement

of the left breast, mammograms, and two sample excisions. Histological examination showed a cavernous hemangioma. On presentation, the patient again complained of intense pain in the left breast with overheating, tenderness on palpation, and radiation into the left arm and back. On inspection, the left breast was almost 100% larger (*Fig. 1*). No pathological palpable lesions were found. The outer quadrant of the left breast felt edematous and soft. Enlarged lymph nodes up to 2 cm in diameter, tender on palpation, were found in the left axilla. Mammography (higher density in the left upper quadrant, BI-RADS-II), computed tomography (CT) of the chest, magnetic resonance imaging (MRI) of the breast, X-ray of the chest, upper abdominal ultrasound, and bone scintigraphy had been performed in another clinic. Lymphangioma of the left breast and thoracic wall with hemangioma-atosis regions were suspected at MRI and chest CT.

Ultrasound (US) of the breast showed a partially involuted breast with easily assessable breast tissue. Both outer quadrants of the left breast up to the axilla were interspersed with multiple echo-poor cavernous structures with diameters of up to 2 cm. The areas were slightly perfused and collapsed under compression with the scanner head (*Fig. 2*). The total extent was estimated to be 15 x 10 cm. Malignancy of the lesion or axillary lymph nodes was not suspected.

Angiography with preparation for embolization showed a lesion of the left thoracic wall without increased arterial perfusion, indicating that the vascular pathology existed on the venous side and consequently made embolization impossible.

Primary irradiation of the region affected was discussed with radio-oncologists but was rejected (irradiation is not an accepted treatment for lymphangiomas).

The patient had been under severe, longstanding psychological stress from her condition. We therefore decided to operatively remove the lesion as a last resort. The patient



Fig. 3. Operative site displaying extent of lymphangioma into the pectoral muscle and subcutis.

was informed of the possibility of ablation in the case of uncertain findings or complications with hemostasis and was prepared for transfusion of packed red cells.

The neoplastic growth had a minimum diameter of 10 cm and was not completely resectable, despite mastectomy. The tumor extended into the pectoral muscle and extended up to the subcutis (*Fig. 3*). Excision in the region of the axilla was complicated by the lesion penetrating the complete subcutaneous area up to the axillary vein. At the junction of the axillary vein, the thoracodorsal vascular bundle had a diameter of approximately 3 x 4 cm. Three Redon drains were put in place. Despite an estimated intraoperative blood loss of more than one liter, the patient did not require administration of packed red cells. The patient had no prior history of heavy intraoperative bleeding, and this blood loss may have been due to shunting of blood into lymphatics (see Pathology below). Symptomatic pain was significantly reduced postoperatively. Secondary reconstructive surgery is not an option for the patient at present and cannot be recommended due to the unclear risk of relapse.

Pathology

Frozen sections confirmed the diagnosis of a cavernous angioma. The level I axillary lymph nodes removed as a precaution showed no signs of malignancy. Histology revealed diffuse tumorous angiomatosis with conglomeration of dilated capillary and cavernous vessels with partly calcified thrombi of variable age (these are frequently present in phleboangioma and can be diagnosed with direct X-ray). The new vessels reached the axillary and dorsal resection margins. The flat cells showed a strong positive immunoreaction with antibody D2-40, identifying them as lymphatic endothelium, and the endothelium stained negatively for the CD 34 endothelial marker for blood vessels (*Figs. 4-6*), indicating that the vessel conglomerations were cavernous lymphatic vessels secondarily filled with blood.

DISCUSSION

Lymphangiomas and hemangiomas are benign lesions, and almost all require surgical treatment when localized in the breast. They are associated with high morbidity because they can become very large, occur in critical locations, and, when surgically removed, there is an increased probability of a secondary wound infection. Pathohistological differentiation between lymphangioma and hemangioma was difficult in our patient, and we made a primary misdiagnosis of hemangioma. Immunohistochemical investigations were required to distinguish between hemangioma and lymphangioma. Correct diagnosis is achieved using the D2-40 antibody or other markers, such as podoplanin, which reliably mark lymphatic endothelium (12). It is important to exclude angiosarcoma, especially with low malignant potential.

After two previous local excisions, this third, extensive intervention eased our patient's pain considerably. We found that exact determination of the extent of the lesion with ultrasound, MRI and angiography was necessary to plan the correct surgical approach. Angiography is not always

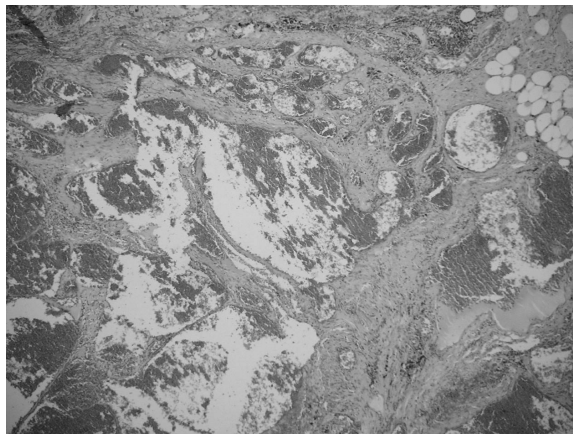


Fig. 4. H&E staining. Benign vascular lesion of the breast with diffuse proliferation of cavernous dilated vessels of varying size, empty or secondarily filled with blood. Vessels sometimes surrounded with a supporting muscular coat and focal lymphoid cells. Immunohistochemical findings in Figs 5 and 6.

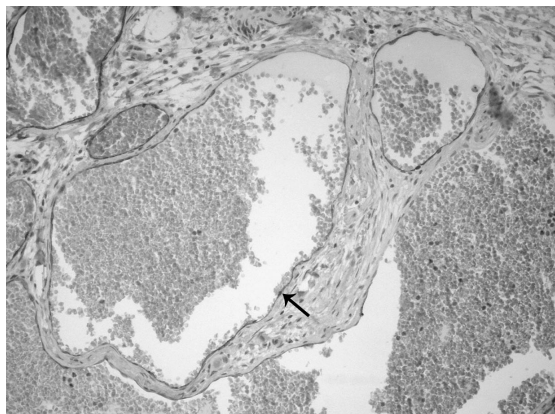


Fig. 5. The flat cells showing a strong positive immunoreaction with antibody D2-40, identifying them as lymphatic endothelium (arrow).

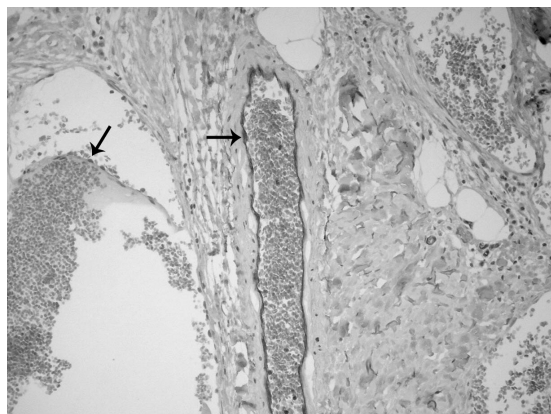


Fig. 6. Immunohistochemistry using an antibody for CD 34 displays positivity for the endothelium in the blood vessels (center arrow), but no staining of lymphatic vessel endothelium (left arrow).

necessary. Our patient is still stable 23 months after surgery. Reconstruction is not being considered at present as a result of the unclear risk of the resected lesion in the axillary area (Fig. 7) and the patient's wishes.

Further Treatment Options

Cavernous hemangiomas respond well to radiotherapy (10–30 Gy, in divided doses spread over 3 weeks), and this may improve the operability of extensive lesions but does carry the risk of secondary tumor induction,

especially in young patients. Very little has been published on the treatment of breast lymphangioma. Atypical vascular lesions have been described in the skin and mammary gland tissue of patients after surgical treatment and radiotherapy of breast cancer.

Sclerosis of such lesions has been reported but was not regarded as an effective alternative to surgical treatment in our patient (13). Because only complete excision is curative, we must still anticipate a relapse in our patient because of the incomplete axillary excision of the lesion. For future



Fig. 7. The healed operative site. The black markings indicate areas with further sonographically visible vessel anomalies.

relapse or progression, irradiation of the axillary region or oral prednisolone, as described by Meunier (6), may be an option.

CONCLUSION

Lymphangioma or hemangioma of the breast must be considered in adult women with progressively multiplying cyst formation. The early recognition of these rare diseases that often develop slowly over years, as seen in our patient, is of great importance, because complete surgical excision of small lesions and consequent cure is possible.

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