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Cystic Lymphangioma of the Breast in a 4 Year Old: Case Report

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Abstract **Case Report**

Cystic lymphangioma, also known as cystic hygroma, is a rare congenital malformation of the lymphatic system characterized by congenital blockage of lymphatic channels or sequestered lymphatic sacs. They are frequently observed in childhood, yet their occurrence in the breast is exceedingly uncommon and is more frequently seen in adults, with only a small number of cases documented in children. We report a case of a 4-year-old female who presented with a large right breast lump which has progressively developed since birth and was diagnosed as mammary cystic lymphangioma after local excision. There was no recurrence of the lesion at the 1 year follow-up.

Keywords: Cystic lymphangioma of the breast, children, imaging, cystic hygroma.

INTRODUCTION

Lymphangiomas are rare congenital hemartomatous malformations of the lymphatic system, typically diagnosed by the age of 2 years in about 90% of cases [1]. While benign, these lesions can infiltrate surrounding tissues and are commonly found in the neck region and axilla. However, they rarely occur in the breast, especially in its cystic subtype, with only a few documented cases in medical literature, mostly in adults [2, 3]. Surgical excision is the preferred treatment for lymphangiomas [4, 5].

We present a case of cystic lymphangioma in the left breast of a 4-year-old boy and provide a brief review of the available literature on this topic.

CASE REPORT

A 4-year-old boy presented with a lump in his right breast that had been gradually increasing in size over the past 4 months and had been present since birth.

There was no history of fever, trauma, or previous surgery at the site. The lump was painless, without nipple discharge, and there were no significant findings in his family history or medication use.

Upon examination, a well-defined, soft, non-tender, cystic mass was detected in the upper outer quadrant of the right breast without glandular induration. Axillary lymph nodes were not palpable, and the opposite breast showed no abnormalities.

Ultrasound imaging revealed a multicystic mass separate from surrounding structures, measuring 43 x 54 x 55 mm (Figure 1).

The patient was then referred for a chest CT to rule out endothoracic involvement and revealed similar findings with thin walled multicystic breast lesion without solid enhancing components, wall calcification or adjacent infiltrative/inflammatory changes (Figure 2).

Fine-needle aspiration cytology yielded clear fluid with numerous lymphocytes but no atypical cells, leading to a provisional diagnosis of lymphangioma.

The patient subsequently underwent an MRI for scientific purposes which showed a multilocular intramammary cystic lesion with fluid signal, featuring thin septa with hyperintensity on T1 and T2 sequences (Figure 3).

We thought that this mass would be a cystic lymphangioma on the basis of the typical imaging findings and cytologic results. The mass was completely excised with safe margins, revealing multiple cystic spaces filled with clear fluid and lined by flat endothelial cells upon gross and microscopic examination. The final histological diagnosis confirmed cystic lymphangioma, and after 10 months of follow-up, there was no recurrence observed.

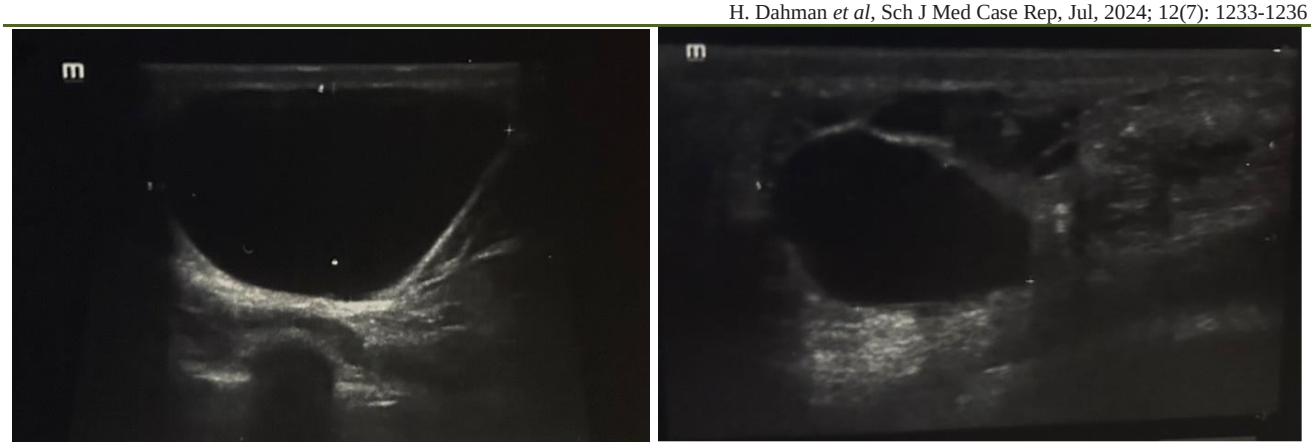

Figure 1: Ultrasound images showing a multi-loculated thin walled cystic mass with septations in the upper outer quadrant of the right breast

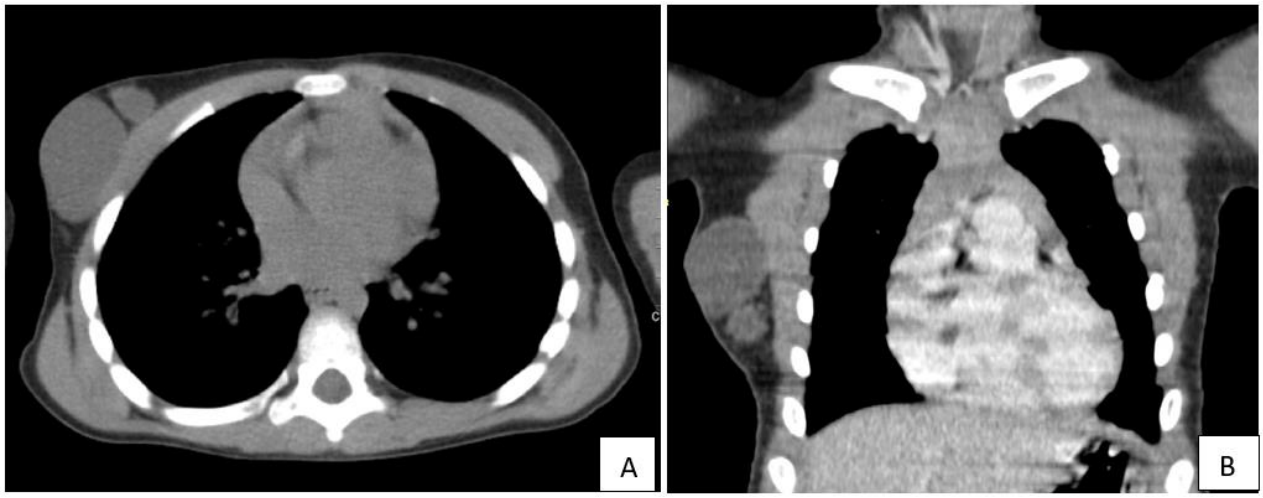

Figure 2: Axial (A) and coronal (B) without (A) and with contrast (B) CT images showing a non-enhanced multicystic mass in the upper outer quadrant of the right breast

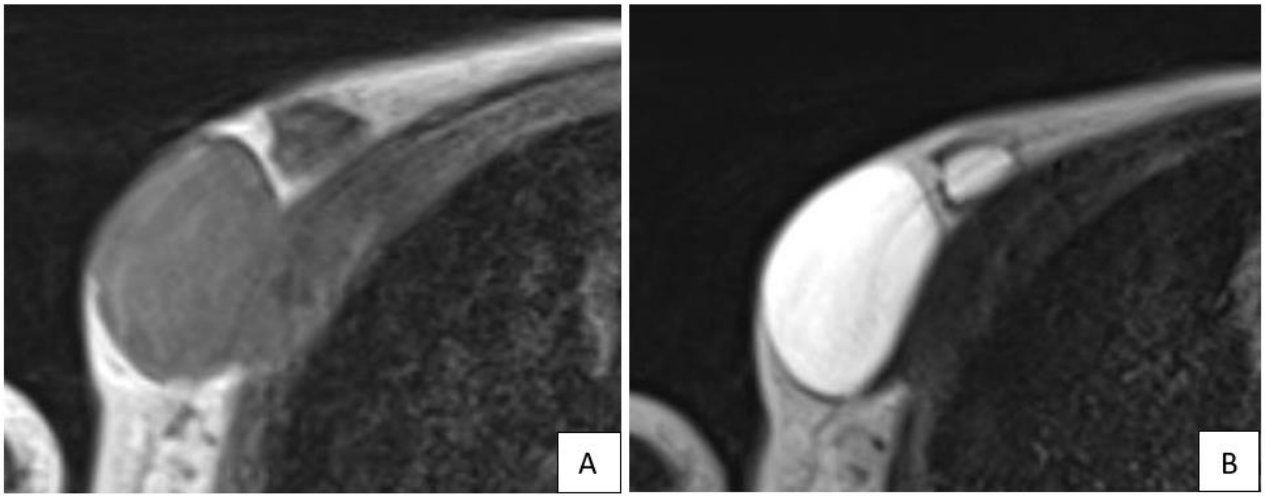

Figure 3: Axial T1(A) and T2 (B) weighted MR sequences showing multicystic breast lesion with fluid signal

DISCUSSION

Cystic lymphangiomas, often referred to as cystic hygromas, are relatively uncommon anomalies that result from congenital hamartomatous malformations in the lymphatic system [6, 3]. These malformations arise due to either the failure of lymphatic connections with the venous system or the sequestration of small lymphatic tissue unable to communicate with the main lymphatic system [2]. Factors contributing to their development include lymphatic channel obstruction leading to secondary dilatation, inherent weakness in

lymphatic walls, and increased lymphatic vessel proliferation [1]. There is some debate regarding the nature of cystic lymphangiomas [6], with some considering them as developmental abnormalities in the lymphatic system and thus classified as hamartomatous lesions, while others view them as potentially aggressive neoplasms. However, due to the lack of documented malignant transformations, they are generally regarded as benign tumors [6, 4].

More than 90% of cystic lymphangiomas present in childhood and are diagnosed by the age of 2 years. Seventy percent of lymphangiomas occur in the neck region; 20% in the axilla; and the remaining 10% in abdominal organs, the retroperitoneum, skeleton, scrotum [7, 1]. Breast involvement is particularly rare, with only a few documented cases in medical literature [8]. The majority of reported cases in the breast are found in the upper outer quadrant, which can be attributed to the drainage patterns of the breast's lymphatics toward the tail and axillary regions [9, 10]. Patients with lymphangioma typically notice a rapid increase in the size of a mass, which is otherwise asymptomatic and lacks noticeable local changes. The most common sign is a painless mass that grows gradually. This rapid growth can be due to factors like cyst hemorrhage, inflammation, or trauma. Upon examination, the mass is well-defined, smooth, mobile, and non-tender [11, 12, 1].

Lymphangiomas are divided into three subtypes: simple lymphangioma, cavernous lymphangioma, and cystic lymphangioma. The distinguishing feature of cystic lymphangioma is the presence of large cyst-like spaces filled with clear lymph fluid, lined by flat endothelial cells. These spaces may contain proteinaceous fluid, lymphocytes, and occasionally red blood cells [1]. Differential diagnosis of cystic lymphangioma includes simple cysts, post-surgical fluid collections, hematomas, abscesses, and hemangiomas. Simple cysts are often bilateral and can occur in various breast regions, characterized by turbid yellow or greenish fluid without lining cells. Hemangiomas typically exhibit thick-walled blood vessels with numerous red blood cells [3, 7].

The standard imaging modalities for diagnosing and monitoring mammary cystic lymphangiomas include typically ultrasound and MRI [13, 2]. The mammographic imaging characteristics of cystic lymphangioma in the breast are generally nonspecific. Additionally, these lesions are often located high in the axilla, making imaging challenging [14]. It is particularly advisable to avoid radiation in pediatric patients whenever possible [6]. Ultrasonography typically reveals a multiloculated, hypoechoic cystic mass with linear septa containing solid components from the cyst walls. Larger lesions may lack well-defined margins and could extend into surrounding tissues.

While the features on ultrasonography can vary, it remains a cost-effective and easily accessible imaging tool without the risks associated with radiation exposure [15]. Magnetic resonance imaging (MRI) is preferred for diagnosing and assessing the extent of cystic lymphangioma in the breast. On MRI, these lesions usually appear as septate masses with low T1- and high T2-weighted signal intensities, showing variable enhancement primarily from septa [16].

Surgical excision is the primary treatment option for cystic lymphangioma in the breast or other body parts. Due to their aggressive nature and tendency to infiltrate nearby tissues, achieving clear surgical margins can be challenging, especially with larger lesions that may have unclear boundaries [10] . Magnetic resonance imaging (MRI) is beneficial in assessing the extent of excision needed [5].

However, complete excision may not always be feasible, particularly when vital structures are nearby or preserving breast tissue is a concern, such as in young females. In such cases, sclerotherapy can play a crucial role as an adjunct to surgery. Sclerosants like Ethibloc and OK-432 help make these lesions more manageable for surgery by reducing fibrosis and facilitating dissection. OK-432, in particular, has shown effectiveness in managing multicystic lesions with septae and those involving multiple anatomical spaces. Other treatment options like drainage, irradiation, and cryotherapy have been explored but are generally less effective and may pose risks like hemorrhage and infection, making them more suitable for patients who are not good candidates for surgery [1, 4, 5].

Incomplete excision carries a high risk of rapid recurrence, as seen in our case where cystic lesions recurred around the previous excision site at 11 months postoperatively.

CONCLUSION

Cystic lymphangioma of the breast is exceptionally uncommon, especially among children. The diagnosis is frequently based on ultrasonography findings. Magnetic resonance imaging should allow a better anatomic correlation. These lesions are typically benign but can present a locally aggressive behavior, therefore complete surgical removal is the treatment of choice.

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