

Cystic hygroma of the breast in a 5 year old boy presenting as a gynecomastia

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Abstract. – Cystic hygroma, also known as lymphangioma, are unusual congenital malformations of the lymphatic system. Cystic lymphangioma is a rare lesion in the breast of children. Only a few cases have been documented in literature. We report a rare case of a 5-year-old boy presented with a gradually enlarging, painless breast mass which was identified sonographically and histologically as a breast lymphangioma and treated by surgical excision. A local excision of the lump was performed and a diagnosis of cystic lymphangioma was made. Cyst was surgically removed, with preservation of normal tissue, and histopathologic findings showed a lymphangioma. The patient is well, after one year of follow-up, with no complaints or recurrence.

Key Words:

Breast, Cystic lymphangioma, Children.

Introduction

Gynecomastia, a benign enlargement of the male breast due to proliferation of the glandular component, is a common clinical condition. The condition is common in infancy and adolescence, as well as in middle-aged to older adult males. In contrast to gynecomastia in adolescent boys and men, prepubertal gynecomastia is rare and should always be considered pathological, prompting a search for a source of estrogen. Although hyperestrogenemia may be endogenous or exogenous in origin, most persons with prepubertal gynecomastia have normal serum concentrations of sex steroids, and an underlying cause is not identified¹.

Cystic hygromas are slow-growing benign tumours resulting from a developmental anomaly

of the lymphatic system². They can occur anywhere in the body, but 75% involve the posterior neck, 20% the axilla and 1% the mediastinum, groin, retroperitoneum and breast³. Lymphangioma of the breast is quite rare with only a few cases being reported in the literature. We present a case of lymphangioma in the breast of five years old boy.

Case Report

A boy who was 5 years old presented with a 6-month history of gynecomastia that had appeared gradually and painless swelling in his left breast. He was the second child of non-consanguineous parents. His medical history and family history were unremarkable. Mental development was normal. His family history was unremarkable.

On physical examination, the patient weighed 12.4 kg (above 25-50th percentile according to the Turkish population) and his height was 118.6 cm (25-50th percentile). The patient's body temperature, blood pressure, pulse rate and respiratory rate were 37.2°C, 120/80 mmHg, 80/minute and 20/minute, respectively. Physical examination revealed a well circumscribed, slightly mobile, and tender soft lump measuring about 6×4 cm, located in the left breast. His genitalia were prepubertal (Tanner stage 1). His general physical and systemic examinations were unremarkable. Laboratory examinations, including complete blood count, serum electrolytes, liver function tests, alkaline phosphatase levels and thyroid function tests, were all within normal limits. Chromosome analysis in cultured peripheral blood lymphocytes was also normal. Laboratory including endocrinological testing (FSH-ICMA 1.08 mIU/mL, LH-ICMA 0.06 mIU/mL, Estradiol 8 pg/mL) was normal for the age and prepubertal

stage, including levels of TSH and T₄. Values of tumor markers (AFP, β-hCG, CEA, and LDH) were also normal. Ultrasonography of the breast showed that a big cystic mass located in the left breast suggestive of lymphangioma.

The patient subsequently had excisional biopsy of the lump. Macroscopically, the lump measured 6×4 cm and weighed 20 g. Pathological gross findings were those of a very large multiseptate cystic lesion covered with serosa. Microscopy revealed that the cyst wall had bundles of smooth muscle with connective tissue. Gross and histopathological examination confirmed the diagnosis of cystic hygroma. The patient is followed up regularly and there is no evidence of recurrence 12 months after surgery.

Discussion

Cystic hygromas, more widely known as lymphangiomas, are rare, hamartomatous, congenital malformations of the lymphatic system with no risk of malignant transformation. They are classified as cavernous, lymphangioma sim-

plex, or cystic lymphangioma⁴. Cystic lymphangioma is a malformation of the lymphatic system. It can affect any site in the body but is seen more commonly in the head and neck region and the axilla. It is also reported to occur in the mediastinum, retroperitoneum, breast and other regions⁵. Cystic lymphangiomas most commonly affect children. About 90% of these lymphangiomas manifest before 2 years of age and are very rarely encountered in breast. Lymphangiomas in children are considered to arise from sequestered lymphatic sacs that fail to communicate with the draining lymphatic channels. This is a widely accepted theory⁶. However, abdominal trauma, inflammatory process, surgery or radiation therapy may lead to the secondary formation of such a tumour⁷. Typically, lymphangiomas are described as fairly well circumscribed and soft, as observed in our case. Imaging techniques such as ultrasonography, mammography, and magnetic resonance imaging may be used to assess, make a clinical diagnosis and for follow-up⁸. Ultrasound of the left breast in the patient showed a cystic mass in contrast to glandular tissue seen in gynecomastia. Differential diagnosis of cystic condition of the breast was made after the surgical excision.

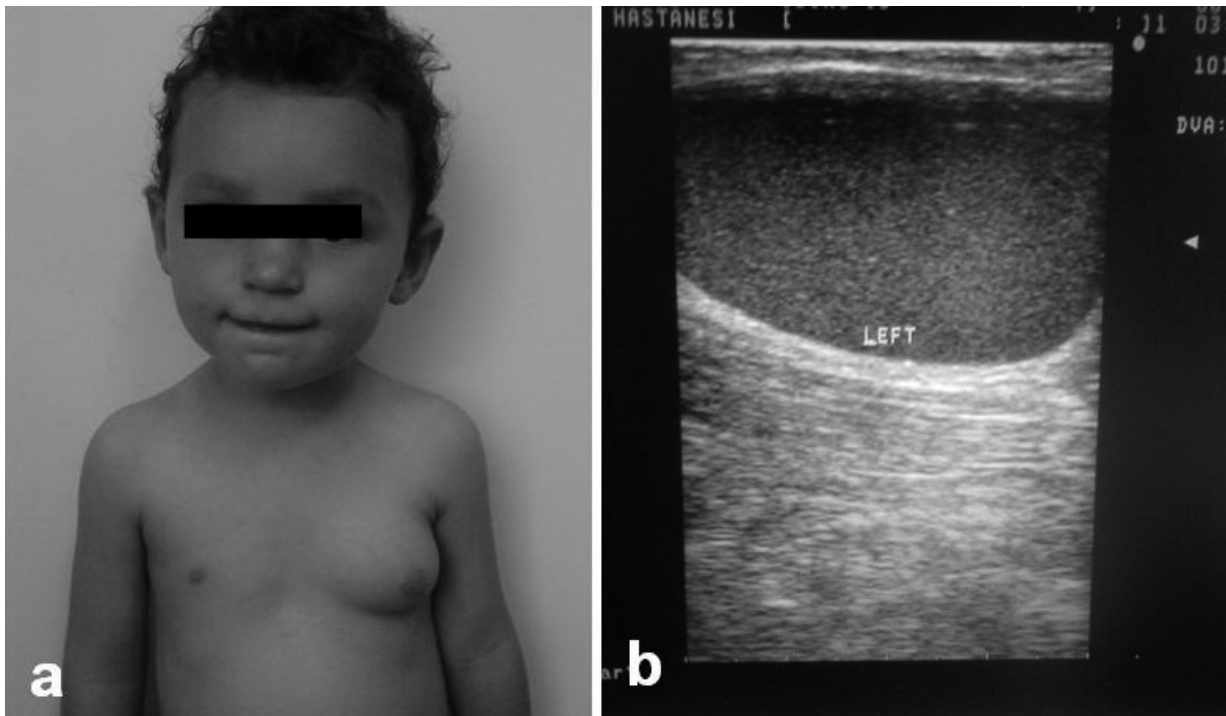


Figure 1. *A*, The patient with left breast mass. *B*, Ultrasonographic examination showed a large unilocular cystic mass with a thin wall and clear content.

Differential diagnoses of the breast cystic lymphangioma include gynecomastia, neoplasms, chronic diseases, trauma, drugs and endocrine disorders. Asymmetric gynecomastia is common, and unilateral gynecomastia may actually represent a stage in the development of bilateral diseases. Although other disorders such as neurofibromas, lymphangiomas, hematomas, lipomas, and dermoid cysts may lead to unilateral, often eccentric breast enlargement. Fine-needle aspiration biopsy may be very helpful in discriminating between gynecomastia and cancer, although a surgical biopsy should be performed if these procedures do not clearly indicate a benign process⁹. The patient had normal hormone profile and tumor markers. The diagnosis of cystic hygroma was made by a histological examination. There was no recurrence during one year at follow-up.

Surgical excision is the first choice of the treatment in cases of breast cystic lymphangioma¹⁰. Complete tumor resection has satisfactory results although few cases of local recurrence have been described, especially after partial resection. We were able to achieve total excision of the cyst. Although other types of adjuvant treatment, such as radiotherapy or injection of sclerosing agents (OK-432 or bleomycin), have been proposed, they are still controversial⁸. In conclusion; the diagnosis of cystic hygroma should be considered in children who present with a breast mass.

References

- 1) CARLSON HE. Gynecomastia. *N Engl J Med* 1980; 303: 795-799.
- 2) WEISS SW, GOLDBLUM JR. Tumour of Lymph vessels. In *Enzinger and Weiss's Soft tissue tumours*, 4th edition. St Louis: Mosby, 2001; pp. 955-998.
- 3) TORCASIO A, VENEROSO S, AMABILE MI, BIFFONI M, MARTINO G, MONTI M, TINTISONA O, PASTA V. Cystic hygroma of the breast: a rare lesion. *Tumori* 2006; 92: 347-350.
- 4) WAQAR SN, KHAN H, MEKAN SF, KAYANI N, RAJA AJ. Cystic breast lymphangioma. *J Pak Med Assoc* 2004; 54: 531-533.
- 5) YILDIRIM E, DURAL K, KAPLAN T, SAKINCI U. Cystic lymphangioma: report of two atypical cases. *Interact Cardiovasc Thorac Surg* 2004; 3: 63-65.
- 6) SEKI H, UEDA T, KASUYA T, KOTANAGI H, TAMURA T. Lymphangioma of the jejunum and mesentery presenting with acute abdomen in an adult. *J Gastroenterol* 1998; 33: 107-111.
- 7) BOSSERT T, GUMMERT JF, MOHR FW. Giant cystic lymphangioma of the mediastinum. *Eur J Cardiothorac Surg* 2002; 21: 340.
- 8) GUINIER D, DENUE PO, MANTION GA. Intra-abdominal cystic lymphangioma. *Am J Surg* 2006; 191: 706-707.
- 9) GUPTA RK, NARAN S, SIMPSON J. The role of fine needle aspiration cytology (FNAC) in the diagnosis of breast masses in males. *Eur J Surg Oncol* 1988; 14: 317-320.
- 10) TSUKADA H, TAKAORI K, ISHIGURO S, TSUDA T, OTA S, YAMAMOTO T. Giant cystic lymphangioma of the small bowel mesentery: report of a case. *Surg Today* 2002; 32: 734-737.