Case Report

Rare case of cystic hygroma of upper extremity unusual site

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Received: 12 March 2015
Revised: 19 March 2015
Accepted: 20 April 2015

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ABSTRACT

Cystic hygroma is a cystic variety of lymphangiomas. Its common site is cervico facial region, followed by axilla, superior mediastinum, mesentry, and retroperitoneal region. Its occurrence in upper extremity is rarely reported.

Keywords: Cystic hygroma, Lymphangioma, Unusual site

INTRODUCTION

Lymphangioma is the congenital malformation of lymphatic system, resulting from failure of development of communications between lymphatics of various regions.¹ Eighty percent of cystic hygromas reside in cervico-facial region. Other sites are axilla, superior mediastinum mesentery, retroperitoneum, pelvis, and lower limbs. Upper extremity is a rare site where cystic hygromas can be located.²,³ Literature regarding this location of cystic hygroma is scarce. Only a few case reports have described its occurrence in the arm.

CASE REPORT

A 5 year old female child came to our OPD with complaints of swelling of left arm since birth. The swelling gradually increased in size and attained the present size. The swelling was not associated with any pain, fever. There was no history of trauma.

On examination

Single swelling of globular shape approximately 10x8 cm extending from the level of left axilla to elbow joint.

The swelling was non-tender and there were no signs of acute inflammation present. The swelling was palpated to have multiple cystic areas in it which were easily compressible and transilluminant. An X-ray of left upper limb was taken. USG of swelling showed cystic swelling with internal septations likely cystic hygroma. Our diagnosis after clinical examination was cystic hygroma supported by USG.

Figure 1: Clinical photo of swelling in surgery OPD.
DISCUSSION

Hemorrhage hygroma is from greek meaning moist or watery tumour. Cystic hygroma is the most frequent type of lymphangiomas resulting from congenital maldevelopment of lymphatic system.

During the course of embryogenesis of lymphatic system, six primary lymphatic sacs are formed; two jugular, two iliac, one retroperitoneal and one dorsal to the retroperitoneal sac, the cisterna chyli. Later on, numerous lymphatic channels from head, neck, limbs, and body wall join the lymphatic sacs. Any maldevelopment or event resulting in failed or insufficient communication between lymphatics of various organs results in the indexed entity.

Lymphangiomas may be classified as simple (capillary), cavernous and cystic. Simple lymphangiomas are composed of capillary-sized lymphatic channels with considerable connective tissue stroma. Cavernous lymphangiomas are composed of actively growing, dilated lymphatic channels in a lymphatic stroma.

Cysticlymphangiomas or cystic hygromas are single or multiple macrocystic lesions having scarce communication with normal lymphatic channels.6,7 Most of the cystic hygromas are evident clinically before the end of infancy; however, their nascent presentation in adults is also reported.4 Approximately 80% of all cystic hygromas involve the cervico-facial region. Other sites are oral cavity, axilla, mediastinum, abdominal cavity, retroperitoneum, scrotum and even skeleton. Their location in upper extremity is very rare and confined to a few case reports.5

Macroscopically, cystic hygromas are multiloculated, multilobular or macrocystic masses composed of manyindividual cysts. Microscopically, the cyst walls consist of a single layer of flattened epithelium.6 In the patient, the cystic hygroma of the limb was composed of cystic mass, easily delineated by inspection and palpation. The diagnosis of cystic hygroma in the patient was obvious at clinical examination. The cysts may contain milky, serous, serosangious or straw-coloured fluid. Cystic hygroma is usually a transluminant lesion; however, it may turn opaque when there is haemorrhage or infection in it.3,6,8 Cystic hygromas are benign lesions. However sometimes complications can arise. The reported complications are haemorrhage inside cysts, infection, spontaneous or traumatic rupture, nerve compression causing pain and paraesthesias, respiratory difficulty, dysphagia, and disfigurement.1

The management options are surgical excision or sclerosant therapy. The recurrence rates are high even after apparent complete excision of the lesion.3,6,9 We perform surgical excision for cystic hygroma in our institution. Most of the time, surgical excision therapy has proved a boon for the patients having cystic hygroma.

To conclude, upper extremity as location of cystic hygromas are a rarity and should be considered in the differentials of cystic swellings of limb.

Surgical excision is treatment of choice.
Funding: No funding sources  
Conflict of interest: None declared  
Ethical approval: Not required

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DOI: 10.5455/2349-2902.isj20150540  
Cite this article as: Rao DG, Srinivas T, Sreedevi, Satish. Rare case of cystic hygroma of upper extremity unusual site. Int Surg J 2015;2:308-10.