Huge cystic hepatic lymphangiomain a 2-months-old infant: A case report

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ABSTRACT

Hepatic lymphangioma is a rare benign lesion resulting from maldevelopment of the liver lymphatic vessels. It is usually associated with lymphangiomas of other viscera. Solitary lymphangioma of the liver is very rare. We report a rare case of solitary huge cystic hepatic lymphangioma in a two-months-old female infant. She presented with an abdominal mass. Imaging investigations showed a large, multiseptate cystic hepatic mass compressing the bladder but with no solid component. She had surgical resection of the hepatic mass. Histological examination revealed a cystic structure lined by epithelial cells and lymphocytes on the inner walls. The patient was last seen in the fourth-week postoperative. Subsequently, she was lost to follow up. A hepatic lymphangioma is a rare form of lymphatic malformation and its occurrence in a 2-months-old infant has never been reported.

Keywords: *Abdominal, Cystic, Hepatic lymphangioma, Mass.*

Lymphangiomas are benign lesions that result from maldevelopment of primitive lymphatic sacs. They are regarded as congenital malformation of the lymphatic system and account for 4% of all vascular tumors in children [1]. Lymphangiomas frequently occur in the neck and axilla where loose connective tissues allow the expansion of lymphatic channels. Hepatic lymphangiomas are uncommon [1] and intra-abdominal lymphangiomas account for less than 5% of all lymphangiomas [2]. Hepatic lymphangioma is usually associated with lymphangiomas of other viscera. In most cases reported, hepatic lymphangioma is a part of diffuse involvement of multiple organs including the liver, spleen, kidneys, skeleton, gastrointestinal tract, mesentery, mediastinum, lungs, pleura, pericardium and somatic soft tissues. Solitary hepatic lymphangioma is extremely rare [3]. Treatment of hepatic lymphangioma involves surgical resection due to symptoms or when discovered as an incidental finding [4]. A hepatic lymphangioma is a rare form of lymphatic malformation and its occurrence in a 2-months-old infant has never been reported. We report a case of solitary huge cystic hepatic lymphangioma in a 2-months-old female infant.

CASE REPORT

A 2-months-old female presented to our facility with a history of the right upper abdominal mass that was noticed one month prior

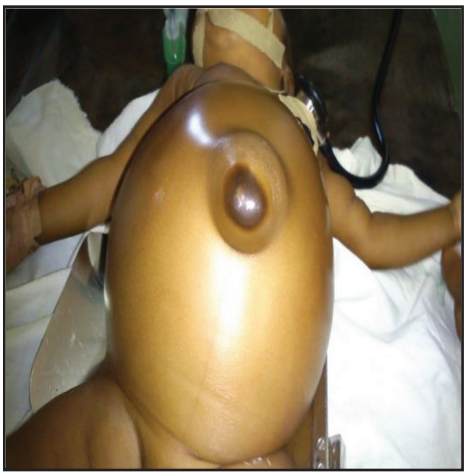

Figure 1: Preoperative picture of the index patient.

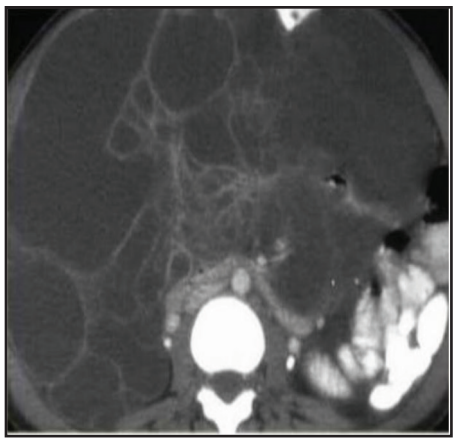

Figure 2: Abdominal Computed Tomography (CT) scan of the index patient.

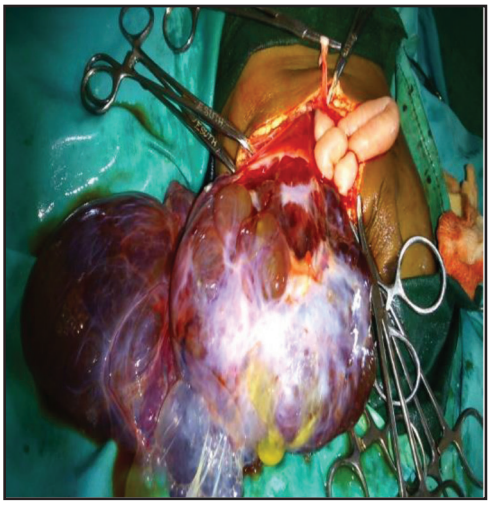

Figure 3: Intra-operative picture of the patient showing the liver mass.

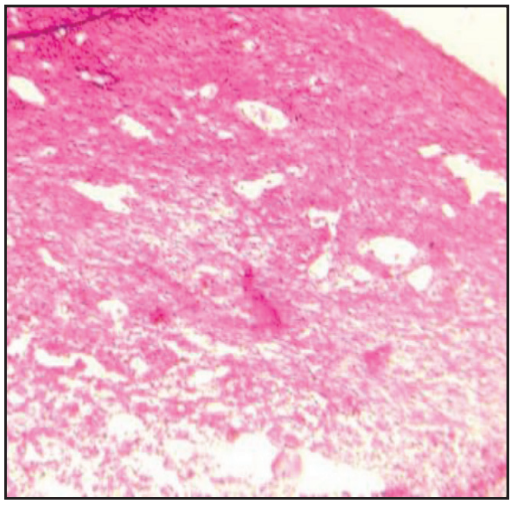

Figure 4: Photomicrograph of hepatic lymphangioma (H & E, X10 magnification).

to presentation. This mass was located in the right upper quadrant and progressively increased in size. There was no jaundice and no fever. On examination, the vitals were stable. Local examination showed a firm, non-tender mass in the right hypochondriac region that extended towards the right iliac fossa and measured about 20cm by 10 cm (Fig. 1).

Laboratory test showed normal full blood count results. Serum bilirubin, alanine transaminase, aspartate transaminase and alkaline phosphatase results were also normal. Abdominal ultrasonography showed a huge complex intra-abdominal cystic mass with multiple septations. The mass occupied almost the entire abdomen and displaced the bowel to the periphery. Further evaluation with computed tomography scan revealed large multiseptate cystic intra-abdominal mass compressing the bladder but with no solid component (Fig. 2). Based on the above-mentioned findings, a differential diagnosis of the large mesenteric cyst, intra-peritoneal lymphangioma and duplication cyst of the gastrointestinal tract were made.

Parents were counseled and the patient was optimized for laparotomy. The intraoperative finding was multiseptate cystic mass attached to the left lobe of the liver measuring 25cm by 12 cm by 7cm (Fig. 3). The cystic hepatic mass was resected and specimen sent for histology. Histopathologic study of the specimen showed large, thin-walled lymphatic channels in loose connective tissue (Fig. 4). These histological findings were compatible with a diagnosis of lymphangioma. The patient was last seen at the out-patient clinic on the fourth week postoperative but she was subsequently lost to follow-up. At the fourth week postoperative, she developed hypertrophied scar and there was no evidence of recurrence.

DISCUSSION

Lymphangiomas are congenital malformation of the lymphatic system composed of lymph containing endothelial lined spaces that vary in size from small capillary channels to large cystic spaces [5]. Lymphangioma could be classified into macrocytic (>1 cm), microcytic (<1 cm), or mixed [5]. It may also be classified as capillary, cavernous, or cystic lymphangioma [5].

There are several hypotheses on the etiology of lymphangioma. Some authorities regard lymphangiomas as true neoplasms that are capable of local aggressive behavior, but overall they are benign [6]. However, others believe that lymphangiomas are hamartomatous malformations that result from the failure of the lymphatic system to communicate with the venous system [6]. The third hypothesis postulates that lymphangiomas represent sequestrated lymphatic tissue that fails to communicate normally with the lymphatic system. These abnormal rests of lymphatic tissue possess some capacity to proliferate and accumulate a vast quantity of fluid, accounting for their cystic appearance [6].

Classically, lymphangioma presents as a soft tissue mass in the neck or axilla of a neonate or infants. Occasionally, lymphangioma may be found in the thoracic, abdominal or pelvic cavities. Our patient had an abdominal (hepatic) lymphangioma.Lymphangioma can be a diagnostic dilemma due to various clinical and imaging features [7]. Hepatic lymphangioma, a malformation of the liver lymphatic system, is an unusual entity that may remain asymptomatic until it presents with abdominal pain or a palpable mass. The index case had a palpable abdominal mass but there was no pain. Any part of the liver could be involved with lymphangioma including the quadrate lobe [8]. Abdominal distension (due to the mass) and pain are the most commonly experienced symptoms in hepatic lymphangioma [9].

Preoperatively, imaging investigations help in the assessment of hepatic lymphangioma. Ultrasound scan is the initial mode of investigation and Makni et al. [7] reported that ultrasound scan has 100% sensitivity when used in evaluating hepatic lymphangioma with experienced hands. Ultrasound is essential for monitoring the size and extent, preceding the excision [10]. Computed tomography (CT) scan is useful in determining the content of the mass, relationship to major vessels, surrounding structures and this is essential in surgical planning [9]. Magnetic resonance imaging (MRI) is an essential modality for assessing

the extent and nature of the lesion especially in solid organs like the liver [10]. Abdominal X-ray has no role in investigating hepatic lymphangioma [11]. MRI was not done in the index patient. A combination of ultrasound scan and CT scan give as much information as that provided by MRI [12]. Our patient had an ultrasound scan and computed tomography scan.

Histological examination confirms the diagnosis of lymphangioma. This shows as numerous spaces, lined by monolayer endothelial cells, containing amorphous eosinophilic fluid and lymphocyte [6,13]. Occasionally, the channel may contain blood due to secondary haemorrhage (due to trauma), resulting in misdiagnosis of the lesion as a haemangioma. But the presence of large collections of lymphoid cells in the stroma tilts the diagnosis in favour of lymphangioma [6]. Immunohistochemistry of the specimen show lymphangioma AS thin-walled vessels vessels lined by lymphatic endothelial cells that are immunohistochemically positive for endothelial markers D2-40 and lymphatic vessel endothelial receptor 1[14].

Hepatic lymphangioma is successively treated by surgical excision and surgical excision is known to be the gold standard for treatment of all types of lymphangioma [12]. However, there have been cases of recurrence with patients who have undergone incomplete excision [12]. Mendez Gallart et al reported a recurrence rate of 10% when the entire cyst wall of the lymphangioma was not resected [11]. Haemorrhage, hepatic damage, hypertrophied scar and wound infection are possible postoperative complications following surgical excision of hepatic lymphangioma.

Our patient developed hypertrophied scar postoperatively which was noticed at the fourth week before she was lost to follow-up. Some success has been recorded with the use of sclerosing agent such as bleomycin [15]. Propranolol has also been tried with some good results. Propranolol works by downregulation of the Raf mitogen-activated protein kinase signaling pathway, with reduced expression of vascular endothelial growth factor (VEGF) which reduces the growth of the lymphangioma [15] but propranolol was not used in the index patient.

CONCLUSION

A hepatic lymphangioma is a rare form of lymphangioma. There is very limited regional data regarding lymphangioma in Africa. To the best of our knowledge, this is the first case of hepatic lymphangioma to be documented in a 2-months-old infant.

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