CASE REPORT

Mesenteric Lymphangioma In A Neonate With Spontaneous Regression: Case Report

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Abstract: Lymphangiomas are benign congenital hamartomas of the lymphatic system. They are preferentially located in the head, neck, and axilla in children. The incidence is estimated to be 1:20,000 in children. Abdominal cystic lymphangiomas are more frequent in boys. They account for 5-6% of all benign tumors in children and only 10% occurring in the abdomen. They are usually discovered incidentally. After delivery, imaging studies including abdominal ultrasonography and computed tomography revealing a diagnosis of mesenteric lymphangioma in the right upper quadrant. They revealed a well-defined cystic mass which was hypotattenuating, mutli-loculated, deformable, multiseptated cystic and solid mass, measuring 6.35×5cm. The lymphangioma has regressed remarkably without any intervention. Based in this case, close observation and tight clinical follow up of such cases can be recommended.

Keywords: congenital hamartomas, cystic mass, mesenteric lymphangioma.

Introduction

Lymphangiomas are rare benign lymphatic hamartomas. The incidence is estimated to be around 1:100,000 and 1:20,000 in adults and in children, respectively [1]. They have a propensity to be located in the head, neck, and axilla in children [2]. Lymphangiomas account for about 5%-6% of all benign tumours in paediatric population [3]. They are usually discovered incidentally during examination for an unrelated abdominal illness. Abdominal ultrasound is the diagnostic modality of choice. Computerized topography (CT) and Magnetic resonance imaging (MRI) are also useful. Diagnosis is confirmed by histopathology [4, 5]. The treatment of choice mainly involve complete surgical excision.
In this case report, we present a female neonate who was diagnosed antenatally to have cystic mass. Diagnosis confirmed by postnatally with ultrasonography and CT to be mesenteric lymphangioma. Thereafter, lymphangioma has regressed remarkably without any intervention.

Case report

A 23-year-old Saudi woman, gravida 1, para 0, had abnormal sonographic findings at 35 weeks’ gestation. The fetus was initially found to have a pelvic cystic mass by routine antenatal ultrasonography in a primary care center of which it was initially thought to be a giant ovarian cyst measuring 4.81x5.37cm (Figure 1). Patient was referred to a secondary hospital and has been re-evaluated. Provisional diagnosis was make to be teratoma. The dimensions of the cystic mass was measured to be 5.4x5.8cm. The patient had an uneventful pregnancy with an increasing in size cystic mass (Table 1) and therefore it was decided for induction of delivery at 39 weeks gestation, as it was afraid the mass could further expand. The pregnancy was complicated by generalized pruritus and polyhydrominos.

A 2.7 kg full term female infant was delivered by induction at a tertiary hospital due to the increasing in size of the neonatal cystic mass. Apgar score were 4 and 8 at 1 minutes and 5 minutes, respectively. At birth, CT of the neonatal abdomen revealed a multi-loculated, deformable, multiseptated cystic and solid mass in the right upper quadrant of the abdomen. It was well defined hypoattenuating mesenteric cystic mass with thin mutable internal septation measured 6.35x5cm. The giant mass situated below the right lobe of the neonatal liver (Table 1). The mass was clearly separate from adjacent abdominal organs which apparently did not communicate or show peristalsis. No evidence of calcification was found, signs of fat attenuation or signs of internal hemorrhage. There is a significant displacement of the internal organs, namely the bowel loops and urinary bladder, to the let aspect of the abdomen. There was normal liver, kidneys, and pancreas. The Liver, kidneys, and the pancreas were normal. There was no retro-crural, para-aortic or pelvic lymphadenopathy. There was no ascites, fluid collection or free air. All of the abovementioned radiological findings suggest mesenteric lymphangioma. Neoplastic markers (AFP, CEA, CA 19-9, CA 15-3, CA125, TPS) were negative.

Surgery was deferred at the time because of the parental choice and was postponed for the following week. After one week, abdominal ultrasonography was performed showing a decrease in the cystic mass to be 3.8x3.5cm in size. The dimension of the mesenteric cyst had halved. Thus, parents decided not to perform the operation.

One week after birth, abdominal ultrasound imaging showed 1.35x2.10cm abdominal mass (Figure 2) and patient still was asymptomatic. One month follow up, abdominal ultrasound imaging showed 1.35x2.10cm abdominal mass (Figure 3) and patient was asymptomatic. Four months later, ultrasound showed a 2.20x0.99cm abdominal mass (Figure 4). After 7 months of follow up, ultrasonography showed no abdominal abnormalities, however, two small pelvic cysts were discovered that were not apparent previously (Figure 5). The patient was asymptomatic. After 1 year of follow up, abdominal ultrasound showed no abnormalities. The patient remained asymptomatic. After 4 years of follow up, there is nothing abnormally detected and patient is symptom free, see Table 1.
Discussion

Abdominal cystic lymphangiomas are very uncommon [6, 7]. Lymphangiomas are more frequent in boys (5:2) with a mean age at presentation of 2 years [6]. Generally, they are common in children, 40% usually present by the age of 1 year and 80% by the age of 5 years. Fifty percent of cases involve the head and neck, with only 10% occurring in internal organs [8]. Mesenteric lymphangioma of the small bowel has been described in less than 1% of lymphangiomas [2]. Sixty percent of these masses are present at birth.

These lymphatic tumors are divided into: 1) simple, with capillary lymphatic channels; 2) cavernous, with dilated lymphatics and the presence of capsule; and 3) macrocystic malformations, that is "cystic hygroma". This results from an embryological failure of the lymphatic system: lack of communication between small bowel lymphatic tissue and the main lymphatic vessels during fetal development result in blind cystic lymphatic spaces lined by endothelial layers. In the abdomen, lymphangiomas occur most commonly in the mesentery, followed by the omentum, mesocolon, and retroperitoneum. The etiology is unclear, but they are considered primarily to congenital in origin [9].

The main differential diagnoses are as follows: hemangioma, branchial cysts, lipomas, dermoid cysts, pancreatic tumours, teratomas, leiomyosarcomas, neurofibromas liposarcomas and rhabdiossarcoma [10]. Usually, they are discovered incidentally during examination for an unrelated abdominal illness. Abdominal ultrasound is the diagnostic procedure of choice. CT and MRI are also useful. Diagnosis is confirmed by histopathology examination.

The definitive treatment is complete surgical excision. Aspiration and injection of sclerosant agents may be recommended for emergency decompression, but as definitive therapy they have a high recurrence rate. Surgical excision has been proved to be superior to mother therapeutic interventions [3, 10].

There is a 10% recurrence rate. To prevent recurrence, complete excision with or without intestinal resection is mandatory. Some studies, however, recommend conservative management in asymptomatic patients, based on a 10% spontaneous regression rate reported in these patients [10]. One could argue whether the spontaneous regression rate is higher than the reported rate. Therefore, as it goes with our case that there is a dramatic regression within the first month. Moreover, the recurrence rate is around 10% and in our case it regressed spontaneously without recurrence for a follow-up period of 4 years. Thus, it could be recommended that conservative management in asymptomatic patients of weekly radiological checks could be done before going for surgical resection in cases of congenital lymphangioma.
Figures and tables

**Figure 1**: shows the antenatal screening at primary healthcare center revealing the ultrasound finding of cystic mass in the abdominopelvic regio.

**Figure 2**: shows the ultrasound finding of cystic lymphangioma at week 1.
Figure 3: shows the ultrasound finding of cystic lymphangioma at week 4.

Figure 4: shows the ultrasound finding of cystic lymphangioma at 4 months.
Figure 5: shows the ultrasound finding of cystic lymphangioma at 7 months.

Table 1: shows the follow-up period and related clinical and radiological findings.

<table>
<thead>
<tr>
<th>Follow up</th>
<th>Clinical examination</th>
<th>Imaging modality</th>
<th>Imaging findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antenatal screening</td>
<td>Uncomplicated pregnancy of mother</td>
<td>U/S</td>
<td>Abdominopelvic cystic mass of 4.81x5.37 cm</td>
</tr>
<tr>
<td>0 week</td>
<td>Palpable abdominal mass with deep abdominal palpation in right upper quadrant</td>
<td>CT</td>
<td>6.35x5.0 cm</td>
</tr>
<tr>
<td>1 week</td>
<td>Palpable abdominal mass with deep abdominal palpation in right upper quadrant</td>
<td>U/S</td>
<td>3.8x3.9 cm</td>
</tr>
<tr>
<td>1 month</td>
<td>Partially palpable abdominal mass with deep abdominal palpation in right upper</td>
<td>U/S</td>
<td>1.35x2.10 cm</td>
</tr>
<tr>
<td>quadrant</td>
<td>4 months</td>
<td>7 months</td>
<td>12 months</td>
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<td>----------</td>
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<tr>
<td></td>
<td>Nothing abnormal detected (NAD)</td>
<td>NAD</td>
<td>U/S</td>
</tr>
<tr>
<td></td>
<td>U/S</td>
<td>Two cysts: 1- 0.7x0.8 cm right</td>
<td>2- 0.7x1.5 cm left</td>
</tr>
</tbody>
</table>

Abbreviation: CT for abdominal computerized topography, U/S for abdominal ultrasonography.

Conclusion

Mesenteric lymphangioma in children is a rare entity. It is usually discovered accidentally. Useful diagnostic modalities are abdominal ultrasound and CT or MRI. Definitive diagnosis is confirmed by histopathology. Complete surgical resection is the ideal choice of treatment. There is, however, a chance of spontaneous regression for mesenteric lymphangioma. Therefore, close observation of such patients with tight clinical follow-up check can be recommended.

Competing interests

The authors declare that there is no competing interest.

Authors’ contributions

Mohammed Alhuwaykim has contributed to the writing the manuscript. Hussain Binamir and Ayman Alhwaykem have contributed in writing and revising the introduction and discussion. All the authors have read and approved the final version of the manuscript.
References