CASE REPORT

In Utero 3D Sonographic Depiction of Cervical Cutaneous Myxoma

Chih-Yao Chen 1,2, Yi-Cheng Wu 1,2, Peng-Hui Wang 1,2, Hsing-I Wang 1,2,3*, An-Hung Yang 2,4, Kuan-Chong Chao 1,2

1 Department of Obstetrics and Gynecology, 2 School of Medicine, National Yang-Ming University, 3 Taipei Mackay Memorial Hospital, and 4 Department of Pathology, Taipei Veterans General Hospital, Taipei, Taiwan

Received 11 May, 2011; accepted 30 May, 2011

KEY WORDS

cervical cutaneous myxoma, cystic neck mass, fetal anomaly, three-dimensional ultrasound

Introduction

Myxoma is a rare, locally infiltrative, benign, connective tissue tumor. It is a mesenchymal tumor of fibroblast origin that produces an excess of mucopolysaccharide and is incapable of producing mature collagen. Histologically, this tumor resembles a umbilical cord, and it can occur in a variety of locations, including the heart, bones, skin, subcutaneous and aponeurotic tissue, genital urinary tract, and skeletal muscle [1,2]. Herein, we report on a case of extracardiac myxoma, which arose from the fetal head and neck. We have also included the three-dimensional (3D) sonographic image.

Case report

A woman aged 34 years old, gravida 2, para 1, was referred to our hospital because of a detected fetal neck cystic mass and the absence of a fetal heart beat when she underwent ultrasonography at 18 weeks of gestation. This woman and her husband were nonconsanguineous. She denied any medical disease except galactorrhea and she had been given bromocriptin to treat it before her pregnancy. An initial ultrasound examination (Kretz Technik, Zipf, Austria) at 20 weeks of gestation revealed the biparietal diameter (BPD) of 33.6 mm (<10th percentile), the occipital frontal diameter (OFD) of 45.0 mm, the head circumference (HC) of 127.3 mm (<3rd percentile), the femur length (FL) of 20.1 mm (<50th percentile), the abdominal circumference (AC) of 162.1 mm (>97th percentile), the humeral length (HL) of 23.8 mm (<50th percentile), the ulna length (UL)
of 21.0 mm (<50th percentile), the tibia length (TL) of 20.7 mm (<50th percentile), and the fibula length (FiL) of 18.9 mm (<50th percentile). A male baby without a fetal heartbeat was noticed. A postcystic neck mass measured approximately $52 \times 54 \times 50$ mm, with a thick wall, a well-defined border, a blurred septation, a hypoechoic density, and an absence of a vascularity; (C and D) a 3D sonographic examination showed a neck cystic mass without vessels.

Fig. 2  A 200-g male baby who died but was delivered after a vaginal induction.

Fig. 3  The postmortem pathology revealed myxoma, which was composed of markedly hypocellularity and a slightly basophilic proteoglycan matrix with a myxoid background. Near avascular but no pleomorphism are noted.
approximately 52 × 54 × 50 mm (Fig. 1A). This mass was thick-walled, had a well-defined border, two major lobulations, and hypoechoic echogenicity with internal multilinear hyperechoic components interspersed within the cystic space (Fig. 1B). No calcification was seen in this mass, and there was no blood flow was detected within this lesion by using color Doppler ultrasound. The trachea and esophagus were patent, and the stomach was visible. No other fetal anomalies were detected. Under the impression of cystic hygroma, amniocentesis was performed and the result disclosed a normal 46, XY karyotype. A further 3D sonographic examination showed a neck cystic mass, which extended from the posterior occipital to the bilateral shoulder area. (Fig. 1C and D). After a detailed discussion with the couple, a deceased 200-g male baby was delivered after vaginal induction due to a missed abortion (Fig. 2). The postmortem pathology revealed myxoma, which was composed of a markedly hypocellularity and myxoid background, nearly avascular, and no pleomorphisms were noted (Fig. 3).

Discussion

Myxoma was first discovered by Virchow in 1871, and he described tumors that histologically resembled the mucinous tissue of the umbilical cord [3]. Myxoma is diagnosed as a true mesenchymal neoplasm that consists exclusively of undifferentiated stellate cells in a loose mucoid stroma that do not metastasize [4].

Fetal nuchal masses are rare, and numerous types of cystic neck masses are needed to make a differential diagnosis, including cystic hygroma, cervical teratoma, nuchal edema, encephalocele or other neural-tube defects and the twin sac of a blighted ovum. Lymphangioma and teratoma are two of the most common causes of fetal cervical masses [5,6]. It is not difficult to differentiate cervical teratoma from myxoma because calcification is commonly identified in teratoma. However, there was barely a vascular structure in the cystic myxoma in our case for which we could use to exclude the probability of lymphangioma.

The sonographic characteristics of cystic hygroma include fluid-filled cystic spaces divided by fine septae, which are commonly observed in the nuchal region and anterior and posterior triangles of the neck. This septum is dense, a so-called nuchal ligament, and it always located at the postmidline and extends from the fetal neck across the full width of the hygroma. Cystic hygroma are typically multiloculated cystic masses with poorly defined borders. In our presenting case, there are three major key points to be differentiated from our case and cystic hygroma (Table 1). Firstly, the cystic wall in our case is thicker than that seen in cystic hygroma. Secondly, coarse septa are more of a hypoechoic density than fine septa, which are observed in cystic hygroma. And, thirdly, the background within this cystic mass is of an inhomogeneous low-level echo, which is different from the anechoic density seen in cystic hygroma.

<table>
<thead>
<tr>
<th>Sonogram index</th>
<th>Cystic hygroma</th>
<th>Cervical teratoma</th>
<th>Lymphangioma</th>
<th>Cystic myxoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wall thickness</td>
<td>Thin</td>
<td>Thick</td>
<td>Thin</td>
<td>Thick</td>
</tr>
<tr>
<td>Border</td>
<td>Poor-defined</td>
<td>Well-defined</td>
<td>Well-defined</td>
<td>Well-defined</td>
</tr>
<tr>
<td>Septation</td>
<td>Multiple, thin, or fine</td>
<td>Coarse</td>
<td>Multiple</td>
<td>Blurred</td>
</tr>
<tr>
<td>Echogenicity</td>
<td>Hypoechoic, heteroechoic</td>
<td>Coarse</td>
<td>Hypoechoic, heteroechoic</td>
<td>Hypoechoic</td>
</tr>
<tr>
<td>Calcification</td>
<td>Absent</td>
<td>Multiple</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Lobulation</td>
<td>Multiple</td>
<td>Increased</td>
<td>Multiple</td>
<td>Multiple</td>
</tr>
<tr>
<td>Vascularity</td>
<td>Absent</td>
<td>增大</td>
<td>Absent</td>
<td>Absent</td>
</tr>
</tbody>
</table>

References