Prenatal ultrasonographic diagnosis of fetal neck teratoma

Anas Ismail *, A.M. Tabari

Radiology Department, Aminu Kano Teaching Hospital, Kano, Nigeria

Received 14 October 2011; accepted 2 November 2011
Available online 26 November 2011

KEYWORDS
Fetal;
Teratoma;
Perinatal;
Ultrasound

Abstract
Teratomas and other germ cell tumors are common pediatric neoplasms. They may have gonadal and extragonadal locations. Congenital teratomas of the neck are unusual. Imaging provides early diagnosis, which is crucial for planning the delivery, definitive treatment and that of associated complications. This report describes the prenatal sonographic features of fetal neck teratoma and its role in early diagnosis and treatment. A 30-year-old gravid woman presented with polyhydramnios in 31 weeks. Ultrasound scan revealed fetal complex neck mass of heterogenous texture and marked polyhydramnios. The mass was excised in the early neonatal period and teratoma was confirmed on histology.

© 2011 Egyptian Society of Radiology and Nuclear Medicine. Production and hosting by Elsevier B.V.
Open access under CC BY-NC-ND license.

1. Case report

A 30-year-old G2P1 woman presented 31 weeks after her last menstrual period, with increasing abdominal girth in excess of the gestational age. Her first pregnancy (five years earlier) was unbooked, which was delivered by spontaneous vaginal delivery with uncomplicated outcome. There was no history of fever, vaginal discharge or bleeding. Furthermore, there was no history of morphologic anomalies in previous delivery or in her family.

On examination, she was not pale, afebrile with mild pitting pedal edema to the ankle joint. Cardiovascular, respiratory and nervous system examinations were within normal limits. Abdominal examination showed gravid uterus with symphysio-fundal height of about 40 cm; with the fetus in longitudinal lie with cephalic presentation. The fetal heart beats were normal.

Laboratory results of full blood count, liver function test, retroviral screening and urine analysis were normal. Her blood group was A +ve while her hemoglobin genotype was AA.

Obstetric ultrasound scan revealed a single live intrauterine fetus in longitudinal lie with cephalic presentation. The fetal heart rate measured 126 beats per minutes and was regular. There is huge complex fetal anterior neck mass with heterogeneous echogenicity. The mass measured about 67 × 78 mm in widest dimensions and demonstrates multiple hyperechoic particles that cast distal acoustic shadow due to foci of calcifications. Additional sonolucent areas of cystic changes were also noted within the mass (Fig. 1). There was no demonstrable communication between this mass and the fetal cranial,
thoracic or abdominal cavities on ultrasound. Furthermore, no other fetal anomaly was shown; except for the persistent fetal neck extension as a result of the anterior neck mass. The fetal movements were also normal. However, there is associated marked polyhydramnios, showing tiny mobile echogenic specks (Fig. 2). The amniotic fluid index measured 36 cm based on four quadrants technique. The placenta has fundal location and appears normal.

Upon suspicion of fetal neck teratoma (FNT), she was referred to consultant obstetrician for further management and follow up scan was recommended.

She was counseled about the prognosis of her fetus and the need to have close monitoring in collaboration with pediatric surgeon. A week thereafter, she developed severe lower abdominal pain and drainage of clear fluid per vaginum. These necessitated consideration for emergency caesarian section in view of anticipated challenging vaginal delivery. On examination in the theater, the cervical os was fully dilated and she immediately expelled a live male baby weighing 2311.1 g by spontaneous vertex delivery with APGAR scores of 6 and 7 at the first and fifth minutes, respectively. The neonate also showed mild respiratory distress with central cyanosis. There

Fig. 1  Prenatal sonogram, showing the fetal neck lobulated mass (arrows).

Fig. 2  Sagittal view of the fetal neck (A), showing cystic and calcific foci. The sonogram of the lower uterine segment (B) shows marked increase in amniotic fluid volume.
is multi-lobulated cystic mass covering the antero-lateral aspect of the neck and supra clavicular region more on the left. The mass extending from the chin to the mid chest and maintaining the neck in a hyper-extension position.

The pulse rate was 102 beats per minute. The cardiac apex was located at fifth left intercostal space, mid-clavicular line. The heart sounds were first and second only, without murmurs. The respiratory rate was 54 cycles per minute with chest in-drawing and flaring of the ali nasi. Other system examinations were essentially normal.

The neonatologists continued the assessment and resuscitation at the neonatal intensive care unit. The neck radiograph showed the mass to be of soft tissue density in the antero-lateral aspect of the neck with few specks of calcifications. There was associated persistent neck extension. However, there was no evidence of abnormal radiolucency or underlying bony destruction. Nevertheless, moderate tracheal compression was noticed (Fig. 3). From the above assessments, the clinical diagnosis of congenital FNT in a preterm, underweight neonate with severe respiratory distress was made.

The baby had remarkable symptomatic improvement in the neonatal intensive care unit and was taken to the operating room on his seventh day for a resection of the neck mass. On initial direct laryngoscopy and bronchoscopy, the upper airway appeared normal other than the extrinsic compression noted from the neck mass resulting in the abnormal position of the larynx and trachea. Via transverse skin incision, the mass was excised.

He had satisfactory recovery following the excision of the tumor but required ventilatory support for about two weeks and was then transitioned to a trach mask using room air. His initial feeding was by expressed breast milk via nasogastric tube two weeks following the surgery and was suckling breast by third week. He was subsequently discharged by the end of fourth week. He remained asymptomatic at four months of follow up.

Histology of the resected specimen showed numerous cysts with trabeculated and smooth walls, all containing yellow, clear fluid. Thick, spongy connective tissue septa separate the majority of these cysts. The mass also contained various types of tissues originating from ectoderm, mesoderm, and endoderm. Rudimentary lung tissue was also found with ciliated columnar epithelium.

2. Discussion

Teratoma is a tumor composed of ectoderm, mesoderm and endoderm; that resulted from abnormal development of pluripotent cells (1). The most common location of congenital teratomas is the sacro-coccygeal region; however, the head and neck are the least common sites (1–3). Furthermore, FNT are rare, accounting for less than 0.1% of all pediatric tumors (2). In a large series of 4257 childhood neoplasms, only four cases of FNT were recorded (2).

As demonstrated in this report, prenatal ultrasonography permits precise identification of FNT and differentiation from other differentials (2). Sonographic detection of a tumor in the fetal neck region also enables appropriate planning of challenging delivery and management of possible postnatal dyspnoea of the newborn (4–6).

In addition, FNT may be associated with polyhydraminos in 30% of cases which occurs due to esophageal obstruction; as shown in this case. Very large or giant FNT may cause fetal hydrops and still birth (1,5–8). In the neck, they most often occur on the anterolateral surface, extending midline from the thyroid gland. They may manifest clinically as severe respiratory distress and dysphagia due to tracheal and esophageal compression (3). As anticipated in this case, anatomical location of the mass may interfere with labor and vaginal delivery (7). Nevertheless, the fetus was delivered via spontaneous vaginal delivery.

The differential diagnoses of this patient include: encephalocele, lymphangioma/hygroma, teratoma, sarcoma, haemangioma, neuroblastoma and goiter (6,8). When a mass is seen in the back of the neck, it may be a fluid collection (abnormal nuchal translucency), a septated fluid collection (cystic hygroma) or a complex mass (neural tube defect, lymphangioma or other skin mass). When a mass originates in the mid-neck, is more likely to be FNT, lymphangioma, or thyroid goiter. These masses can result in hyperextension of the neck, impaired fetal swallowing and polyhydramnios, as well as mass effect on the airway (5–8).

On computed tomography and magnetic resonance imaging, teratoma can be suspected when a multi-loculated lesion with focal areas of low attenuation and high signal intensity (representing lipid) is seen, respectively (5). These modalities are desirable in planning a surgical treatment in extensive lesions (like this case) to prevent iatrogenic damage of the adjacent structures. However, they were not utilised in this case because of financial limitations and urgent need for intervention. This also underscores the relevance of ultrasound in resource-limited settings.

Management strategies for giant FNT include in utero resection, resection on placental support during an ex utero intrapartum treatment (EXIT) procedure, intubation and/or tracheostomy during EXIT, and postnatal resection (5). Early excision is necessary in view of remote possibility of malignant...
degeneration of these lesions (3,5). Although the surgical excision of the neck mass was successful in this case, operative mortality ranges from 9% to 15% (5).

3. Teaching point

Prenatal ultrasound scan provides early diagnosis, exclude differential diagnoses and fetal follow up. These are essential for intrauterine intervention, planned delivery or prompt postnatal treatment. This case shows the value of ultrasound in diagnosis and to describe the radiologic features of FNT.

References


