

Sonographic features of hemivertebra at 13 weeks' gestation

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Abstract

Hemivertebra is a rare congenital spinal disorder where only one side of the vertebral body develops, leading to deformation of the spine, such as scoliosis or kyphosis. Previous reports suggest that the diagnosis may be based on antenatal sonographic examination after 14 weeks. We present the sonographic features of a fetus with solitary hemivertebra at 13 weeks' gestation confirmed by postmortem babygram, magnetic resonance imaging (MRI) and pathological examination. It shows that the condition may manifest in the first trimester of pregnancy.

Key words: fetal abnormality, first trimester, hemivertebra, postmortem MRI, ultrasound.

Introduction

Hemivertebra (ICD 9 code 75614) is a rare congenital abnormality of the spine where only one side of the vertebral body develops, leading to deformation of the spine, such as scoliosis or kyphosis.¹ It has been reported to be associated with other skeletal anomalies of the spine, ribs, and limbs, cardiac and genitourinary systems (e.g. VACTERL syndrome).²

The incidence of hemivertebra is estimated at 5–10:10 000 births.³ A few reports have described the antenatal ultrasonographic findings. In these descriptions, the abnormalities were usually detected from 15 weeks onwards.^{4–7} The earliest sonographic diagnosis reported in the literature was at 14 weeks.^{6,8} We present the prenatal diagnosis of a fetus with hemivertebra at 13 weeks' gestation. Postmortem X-ray examination (babygram), and pathological findings confirm the sonographic diagnosis. It was demonstrated that the condition can be diagnosed by comprehensive ultrasonographic examination in the first trimester of pregnancy.

Postmortem MRI can provide additional information to autopsy for gross pathology of most organ systems. It is an especially valuable adjunct to autopsy for abortuses, which are sometimes too small for abnormalities to be appreciated in gross pathological examinations. Tissue autolysis is another genuine problem for postmortem examination of the central nervous system (CNS); this is especially true if autopsy is not immediately available.⁹ We also compare postmortem MRI with babygram to see if it can add useful information in this case.

Case Report

A 33-year-old Chinese woman had ultrasound examination at the first trimester of pregnancy for nuchal translucency measurement for aneuploidy screening. Her husband was Chinese. She had delivered one healthy normal baby. The couple's family histories were unremarkable. Abdominal sonography (Acuson Sequoia, 6 mHz, Mountain View, CA, USA) revealed a single fetus. The gestational age was compatible with

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Figure 1 Abdominal sonography showing hemivertebrae (arrow).

13 weeks' gestation based on crown-rump length of 67 mm by Hadlock's equation.¹⁰ Nuchal translucency measurement was within the 95 centile. A slight distortion of lumbar spine was demonstrated at the coronal plane. A small triangular bony structure was wedged against the normal vertebral bodies (Fig. 1). The initial diagnosis was single hemivertebra. No intracranial abnormalities were found. Because of the unsatisfactory position, the fetal facial profile could not be seen. The fetal heart, stomach, kidney and bladder appeared normal. The placental thickness and liquor volume were normal. Follow-up ultrasound examination at 16 weeks' gestation demonstrated the same vertebral anomaly. Left cleft lip and palate were also diagnosed. The fetal heart, stomach, intestine and urinary system were normal. Although no other abnormalities were found, it would be very difficult to conclude that the fetus was normal. The couple elected to terminate the pregnancy. This was performed by vaginal administration of prostaglandin. Postmortem examination revealed a female abortus with cleft lip and palate. In the postnatal examination, X-ray examination (babygram) and MRI (1.5 Tesla, Signa, GE Healthcare), a solitary hemivertebra was observed at 3rd lumbar segment of the spine with localized scoliosis (Figs 2,3). Thirteen pairs of ribs were identified bilaterally. All the limbs were normal. The placenta was of normal weight for gestational age. Postnatal examination, babygram and MRI confirmed the ultrasound diagnosis without additional abnormalities identified. Placental tissue for karyotyping study showed 46, XX. Pathological examination was consistent with the prenatal finding.

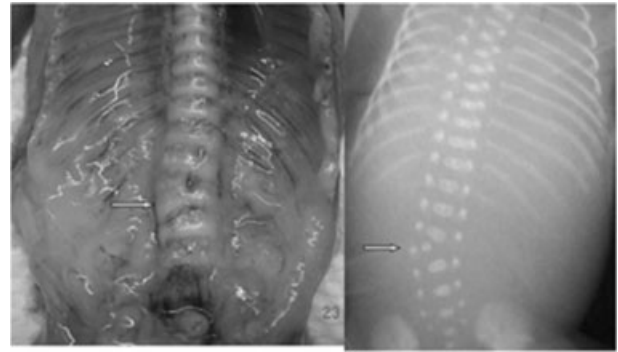


Figure 2 Lumbar hemivertebrae (arrow) was demonstrated by the postmortem photograph (left side) and babygram (right side).

Discussion

Hemivertebra is one of the most common vertebral anomalies. It results from failure of a vertebra to form on one side, resulting in a laterally based wedge vertebra with half a vertebral body, a single pedicle, and hemilamina.^{1,11} It has been suggested that the anomaly occurs at an early stage of membranous vertebral element formation, and once a membranous vertebral body is formed, the process of chondrification and ossification occurs in this outline.¹² At 6 weeks' gestation, two lateral chondrification centers arise in the developing vertebral bodies. These chondrification centers then unite by 7–8 weeks to form the primary ossification center of the vertebral body.¹¹ Because the ossification centers of vertebra can be initially visualized by ultrasound examination after 12 weeks' gestation, the defect in ossification can be visible on early scans.⁵

The cause of hemivertebra is unknown. The distribution pattern of the anomaly does not implicate a specific environmental or genetic factor.⁶ The association with chromosomal abnormalities is small.⁵ Hemivertebra should be differentiated from the other vertebral abnormalities (wedge vertebra, butterfly vertebra, bloc vertebra, bar vertebra or any combination) that cause congenital scoliosis, open neural-tube defects, and diastematomyelia.² Because hemivertebra might be associated with other anomalies that might place a great burden on the future outcome,^{13–15} the diagnosis of such a skeletal spinal deformity calls for a comprehensive anatomic scan and a detailed echocardiographic examination.⁶ The most common associated anomalies involve other skeletal anomalies of the spine, ribs, and limbs. Cardiac and genitourinary tract anomalies are the more common



Figure 3 Postmortem MRI T2-weighted coronal image, showed lumbar hemivertebra (arrow) and normal kidneys.

extra-musculoskeletal anomalies with hemivertebra. Anomalies of the CNS and gastrointestinal tract have also been reported. Hemivertebra may be part of a syndrome including Jarcho-Levin, Klippel-Fiel, and VACTERL syndrome.^{14,15} When involving the thoracic spine, there is usually a missing rib in the affected side, presented as an unequal number of ribs. The association of hemivertebra with cleft lip and cleft palate is rare. In addition, this fetus had 13 pairs of ribs. We do not know whether this represents a previously unreported syndrome or association.

Advances in ultrasonography allow prenatal description of many rare syndromes and malformations in the first trimester.¹⁶ We have shown here that the prenatal diagnosis of hemivertebra is feasible in the first trimester of pregnancy. The prognosis of hemivertebra is related to the site of the affected vertebra, the number of affected vertebrae, and the associated anomalies.² In addition, recent corrective surgery of congenital scoliosis caused by hemivertebra has much improved the prognosis. If no other major fetal malformations are detected, the prognosis of isolated hemivertebra is good and expectant management of pregnancy can be adopted.⁸ An early diagnosis can give parents valuable information that helps them to decide the fate of pregnancy.

The advent of fetal MRI has revolutionized the ability to visualize fetal anatomy and pathology. It can add information in cases in which sonography is insufficient to provide an adequate diagnosis. The usefulness of *in utero* MRI in detecting some CNS abnormalities has been reported. Fetal MRI has been shown to contribute in the evaluation of neck, chest, abdominal and pelvic masses. It is also helpful in the assessment of normal lung volume and documentation of liver position in fetuses with congenital diaphragmatic hernia,¹⁷ and in fetal skeletal assessment.¹⁸

Postmortem MRI can provide similar information to autopsy for gross pathology of most organ systems. In this case, postmortem MRI was performed not only for confirmation of the vertebral abnormality, which is well demonstrated by a babygram. Postmortem MRI mainly serves to identify associated CNS anomalies, which are difficult to assess during autopsy because of the very small size of the abortus, neurological immaturity and tissue autolysis.^{9,19} Other soft tissue abnormalities associated with VACTERL syndrome, such as renal anomalies can also be well demonstrated on MRI. These would help to focus the subsequent autopsy examination.

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