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A case study on the pitfalls in prenatal ultrasonic detection of butterfly vertebra

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ABSTRACT

Introduction: Butterfly vertebra is a rare congenital anomaly that is observed both in isolation and also as part of syndromic diseases. In prenatal ultrasonic scans the typical shapes of the two halves for butterfly vertebra are wedge-shaped or triangular. In the case we presented, the 3 dimensional computed tomography (3D CT) showed that the shape was unusual and rare. To improve the prenatal ultrasonic discriminability of this rare form of butterfly vertebra we used multi-directional ultrasonic images, corresponding to postpartum 3D CT images.

Case report: A 25-year-old woman was referred to our department for ultrasound screening. The routine fetal back spinal scan yielded findings indicative of an anomaly within the ninth thoracic vertebral body. The affected vertebra was examined by two-dimensional (2D) and three-dimensional (3D) ultrasound while the fetus was prone and supine. The focussed scanning of the fetal spine from the back, anterior and lateral approaches aided us to reach the final prenatal diagnosis of butterfly vertebra with asymmetric halves.The diagnosis of butterfly vertebra was confirmed by the radiologist with 3D CT after the woman chose to terminate the pregnancy due to multiple malformations. In 3D CT, the body of the ninth thoracic vertebra appeared to be two lateral halves of different sizes, and the bigger half was C-shaped. When prenatal ultrasonic images and postnatal CT images were compared, the echoic shape of the affected vertebra scanned from the front right side was very similar to the CT.

Conclusion: Due to the variable sizes and shapes of vertebrae affected in butterfly vertebra, prenatal diagnosis can be difficult using ultrasound. When the presence of fetal vertebral abnormalities is suspected, it is imperative for sonographers to adopt a comprehensive approach that extends beyond the conventional spinal examination performed solely from the dorsal aspect of the fetus. Instead, a thorough assessment should involve scanning the fetus from various angles, including anterior and lateral perspectives, in order to obtain a comprehensive and detailed evaluation of the identified vertebra.

1. Introduction

Butterfly vertebra also known as sagittal cleft vertebra is a rare congenital anomaly that is so named because the affected vertebra has the appearance of butterfly wings on x-rays. This anomaly is observed both in isolation and also as part of syndromic diseases, such

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as Alagille syndrome [1], Klippel–Feil syndrome [2], and Jarcho–Levin syndrome [3]. Katsuura et al. [4] reported that in the non-syndromic cases of butterfly vertebra, the most common complaint was lower back pain, observed in 30 % of the cases, followed by mid back pain, observed in 10 % of the cases. They also found that 90 % of the multiple butterfly vertebra cases were associated with a syndrome, while single butterfly vertebra was more likely to be an isolated defect. Butterfly vertebra can easily be missed or misclassified as other defects by prenatal ultrasonic examination because of its rarity. In prenatal ultrasonic scan, though the typical shapes of the two halves for butterfly vertebra are wedge-shaped or triangular, the affected vertebrae of butterfly vertebra can in fact have different sizes and shapes, which makes it challenging to diagnose this condition by ultrasound. In the case we presented, the shape was unusual and rare, which was proven by 3 dimensional computed tomography (3D CT). We provided multi-directional ultrasonic images corresponding to postpartum 3D CT images to improve the prenatal ultrasonic discriminability of this rare form of butterfly vertebra.

2. Case Report

A 25-year-old gravida 2 para 1 woman was examined by ultrasound at 22 weeks of gestation. The woman had no family history of any spinal abnormality. Her first baby was normal. During this pregnancy there was no vaginal bleeding, hypertension, diabetes mellitus, or any other abnormalities. In the non-invasive prenatal testing, low risk of trisomy 21, trisomy 18, and trisomy 13 was seen.

During the scan, measurements of bi-parietal diameter, head circumference, abdominal circumference, and femur length corresponded to 22 weeks of gestation. Initially, with the fetus in a prone position, the sonographer conducted a standard spinal examination, scanning the dorsal aspect of the fetus. This initial assessment unveiled the presence of a deformity within the thoracic region. Then the sagittal, transversal, and coronal views of spine were obtained from the posterior aspect of the fetus by two-dimensional (2D) and three-dimensional (3D) ultrasound. The sagittal view detected two separate ossification centers in the anteroposterior direction at the level of the body of the ninth thoracic vertebra (T9) (Fig. 1a). In the transversal view, the T9 vertebral body (Fig. 1b) had a different appearance from that of the neighboring vertebral bodies (Fig. 1c). No scoliosis was detected in the 3D coronal view, and the T9 vertebral body looked like a single ossification center located on the right side, which appeared to be smaller than the neighboring bodies (Fig. 1d). The coronal cleft vertebra was interpreted as comprising two distinct ossification centers, discernible in both anterior and posterior orientations, as evident in the sagittal (depicted in Fig. 1a) and transverse sections (illustrated in Fig. 1b). The spinal column was subjected to 2D and 3D ultrasound scans from anterior and lateral orientations subsequent to positioning the fetus in a supine posture. It was observed that the vertebral characteristics exhibited a marked disparity when compared to the prone position. In the transverse view (as depicted in Fig. 1e) and the 3D coronal view (illustrated in Fig. 2a) when observed from the anterior aspect, a

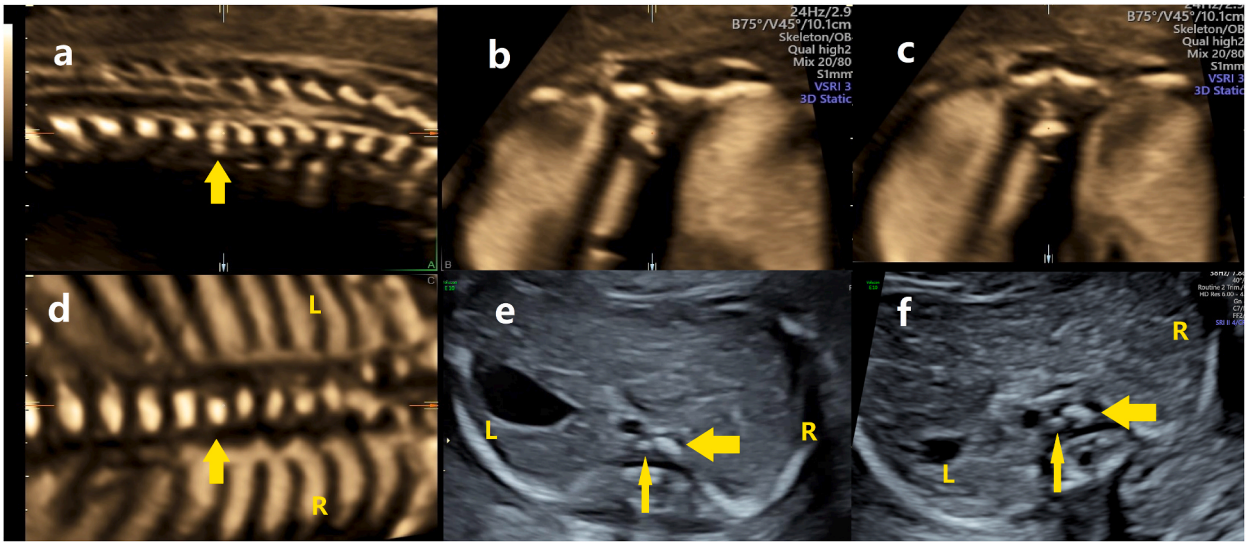

Fig. 1. a–d illustrates the findings derived from 3D volume data obtained while the fetus was in a prone position. (a) Sagittal view of the spine showed two separate ossification centers (arrow) in the anteroposterior direction at the level of the T9 vertebral body. (b) Transverse view of the affected body showed two separate ossification centers in the anteroposterior direction. (c) Transverse view of the normal neighboring body. (d) The 3D reconstructed coronal view. The T9 vertebral body (arrow) resembled a single ossification center located in the right and appeared to be smaller than the neighboring bodies. No scoliosis was detected. (e) Transverse views of the T9 vertebral body in a supine position, as observed from a direct anterior perspective, two distinct ossification centers are discernible. Notably, the left ossification center (indicated by the thin arrow) appears considerably smaller in size compared to the right ossification center (highlighted by the thick arrow). (f) Transverse views of the T9 vertebral body in a supine position, when observed from the right anterior aspect, also reveal the presence of two ossification centers. Notably, the left ossification center (indicated by the thin arrow) is noticeably smaller in size, whereas the right ossification center (highlighted by the thick arrow) exhibits a larger, C-shaped configuration.
Note: L, left side; R, right side.

noteworthy observation was made regarding the T9 vertebral body. It became evident that the T9 vertebral body exhibited a bifurcated configuration, with the left half being noticeably diminished in size in comparison to the right half. This observation aligns with the characteristic features of butterfly vertebrae, where the two distinct ossification centers were found to be situated on the left and right sides, contributing to this anatomical variation. Moreover, in the transversal view from the front right side, the right half of the T9 vertebral body appeared to be C-shaped (Fig. 1f). The complete scanning of the fetal spine from back, front and lateral sides helped us to reach the final prenatal diagnosis of butterfly vertebra with asymmetric halves, which was not possible with the routine scanning from the back side only.

The woman chose to terminate the pregnancy as the fetus also suffered from tricuspid stenosis and severe right ventricle dysplasia. She refused to conduct a genetic examination. Fetal autopsy was permitted, and the cardiac defects were proven. We used 3D computed tomography (CT) to reconstruct the cadaveric spine, and the radiologist made a diagnosis of butterfly vertebra. In the 3D CT image, oriented in the anteroposterior direction, it was apparent that the T9 body comprised two lateral halves of different sizes (Fig. 2b). Furthermore, when viewed from the lateral perspective, the larger of these halves exhibited a C-shaped configuration (Fig. 2c). The echogenic morphology of the affected vertebra, as visualized in the transverse view from the right anterior perspective, closely mirrored the findings obtained from the CT scan.

3. Discussion

Butterfly vertebra, also known as sagittal cleft vertebra, is caused when a residual notochord produces a fusion defect in the two lateral chondrification centers of an embryonic vertebral body [5]. The typical prenatal ultrasound findings for butterfly vertebra are a vague or wedge-shaped vertebra on the sagittal section and two wedge-shaped or triangular halves with opposing tips on the coronal section [6]. In most cases of butterfly vertebra the two halves of the vertebra have been found to be equal in size, but they can also be asymmetric in some cases. Our case was just one of the few asymmetric cases. In the case described here, one half of the vertebra body was hypoplastic. When subjected to the standard fetal back scanning approach, whether utilizing 2D or 3D ultrasound modalities, the hypoplastic half of the vertebra proved challenging to discern clearly due to the presence of an acoustic shadow cast by adjacent fetal bones.

Moreover, in this case, the two halves of the affected vertebra were not typically wedge-shaped or triangular in ultrasonic images. A notable characteristic of the vertebral body was its C-shaped configuration. This particular morphology can be attributed to the diversity in shapes exhibited by sagittal clefts, with the shapes of the half-chondrification centers potentially varying accordingly. Merbs [7] classified sagittal clefts into three shapes based on the shape of the defect: linear, circular, and triangular. In our case the cleft was circular. In this exceptional case, the vertebrae exhibited an uncommon presentation, manifesting as two distinct ossification centers oriented in the anterior-posterior direction in the ultrasonic images acquired from the fetal dorsal aspect. We mistook it as a coronal cleft vertebra initially as coronal cleft vertebra results from the failure of fusion of ventral and dorsal ossification centers [8]. The echoes of the coronal cleft vertebra are discontinuous in the sagittal and transverse section by ultrasound, and are separated into two ossification centers in anteroposterior direction. Our misidentification was maybe because the scanning plane passed through the anterior and posterior branches of the C-shaped half, and the junction between these two branches were not scanned in the section or

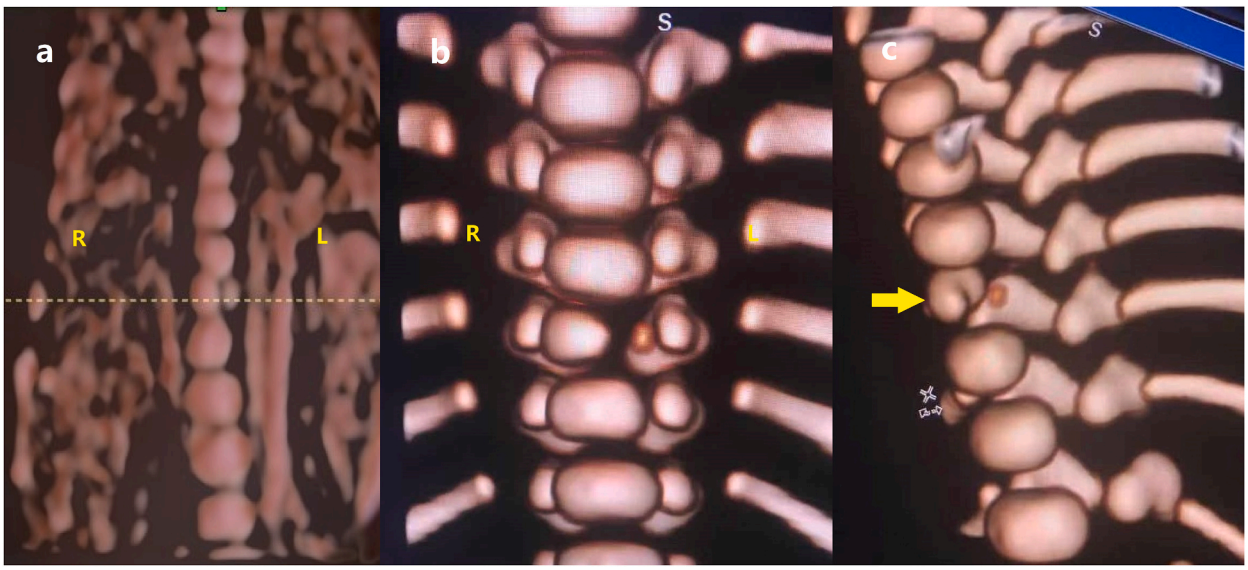

Fig. 2. (a) Reconstructed coronal view obtained via 3D ultrasound while the fetus was in a supine position. Two ossification centers of the T9 vertebral body are visualized and the left one appears to be smaller than the right one. (b) 3D CT coronal view. Two ossification centers of the T9 vertebral body are shown. (c) The 3D CT side view. The right half of the affected body was C-shaped (arrow).
Note: L, left side; R, right side; S, superior.

was hidden by the acoustic shadow of adjacent fetal bones.

This particular case involved the acquisition of 2D and 3D images, capturing the unique presentation of a butterfly vertebra from multiple vantage points. Furthermore, the customary practice of scanning the fetal spine from the dorsal perspective was employed as part of the evaluation process. Consequently, this endeavor significantly enriched the prenatal ultrasound image repository dedicated to butterfly vertebrae. In this particular case, it is noteworthy to acknowledge a limitation in our findings. Specifically, we were able to closely approximate the actual vertebral morphology, as depicted by the 3D CT scan, solely when the affected vertebra was examined from the right anterior aspect using 2D ultrasound. We still are unsure about how to select a more proper 2D scanning direction and obtain more accurate 3D ultrasonic images. Radiology can be an important option in cases of uncertainty, to clarify the diagnosis and get more information whether prenatally or postnatally.

In conclusion, the prenatal diagnosis of this condition by ultrasound can be challenging as the affected vertebra can be of different shapes and sizes in cases of butterfly vertebra. Hence, in cases where there is suspicion of fetal vertebral abnormalities, it is imperative for sonographers to refrain from solely relying on the customary spinal examination performed from the dorsal aspect of the fetus. Restricting the assessment to a partial view may lead to inaccurate conclusions. To ensure a comprehensive evaluation, sonographers should adopt a multi-directional approach, encompassing imaging from the anterior and lateral aspects of the fetus, thereby obtaining a holistic perspective of the identified vertebra.

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Consent to participate

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Additional information

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CRediT authorship contribution statement

Jia-Qi Hu: Conceptualization, Data curation, Writing – original draft. **Yu-Guo Zhang:** Conceptualization, Formal analysis, Writing – review & editing. **Wei Feng:** Data curation, Writing – original draft. **Hua Shi:** Formal analysis, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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