Case Report

Case report of a type II single umbilical artery seen in the first trimester anomaly scan

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ABSTRACT

Umbilical arteries normally originate from a pair of allantoic arteries. Obliteration or non-formation of any of the two umbilical arteries gives rise to a single umbilical artery (SUA). 0.5–1% of fetuses show the presence of a SUA. A rarer abnormality is when a SUA is found originating from the abdominal aorta, representing the persistent vitelline artery. Such an anomaly is termed Type II SUA. Here, we present a case of Type II SUA in a 30-year-old first trimester gravid female. Obstetric ultrasound done in the first trimester showed non-visualization of the umbilical arteries adjacent to the urinary bladder. Instead, an aberrant, large vessel was seen tracking anteriorly from the aorta towards the umbilical cord. The aorta distal to the origin of this aberrant vessel appeared hypoplastic.

Key words: First trimester, Single umbilical artery, Type II single umbilical artery, Ultrasound, Umbilical artery anomalies

In a developing fetus, two umbilical arteries are noted. When one of these umbilical arteries fails to develop, the fetus has a single umbilical artery (SUA). As per the review of the literature, four types of single umbilical arteries were hypothesized. Our case is of type II SUA, which has a reported incidence of 1.5% of all SUAs [1]. In a few case reports of Type II SUA published in literature [2-6], rarely normal fetal development was seen. Most of these cases were detected in the second trimester and had an unfavorable prognosis due to multiple fetal developmental anomalies. Ours is one of the first cases of early detection of Type II SUA in the first trimester of pregnancy.

CASE REPORT

A 30-year-old gravid female of 6–7-week gestation was referred to our Department of Ultrasound in September 2021, for a routine obstetric scan. This was her second pregnancy (G2P0A1). All her scans were performed on a GE Voluson E10 ultrasound machine. The patient had a history of a spontaneous missed abortion in July 2021 (crown-rump length corresponding to 7 weeks 2 days – no chromosomal analysis done on the abortus). In her subsequent pregnancy, she presented at 6-week 2-day gestation, according to the menstrual age. Ultrasound revealed a single live embryo of 7-week 1-day gestation (according to the crown-rump length). The rest of the scan was unremarkable.

After 2 weeks, at 9-week gestation, the patient presented in the emergency with vaginal bleeding. On ultrasound, few clots were noted within the cervical canal. The rest of the scan was unremarkable.

In the first-trimester anomaly scan (13 weeks of pregnancy) done on November 03, 2021, the following ultrasound findings were noted: Non-visualization of the umbilical arteries adjacent to the urinary bladder (Fig. 1). Instead, an aberrant, large vessel was seen tracking anteriorly towards the umbilical cord (Figs. 2a, b and 3b) with the arterial flow on color Doppler (Fig. 4). This vessel was seen arising from the abdominal aorta (as seen in the sagittal section) at the level of the superior mesenteric artery (SMA). The aorta distal to the origin of this aberrant vessel appeared hypoplastic (Fig. 3a). A cross-section of the umbilical cord showed the presence of a two-vessel cord (one artery and one vein). On ultrasound examination, no other gross structural anomaly was seen in the fetus. The patient underwent a Dual Marker Test, which revealed a low risk for Trisomy 21, 18 and 13.

Another obstetric ultrasound examination was performed at 16–17 weeks of gestation on November 24, 2021. The findings listed above were documented and confirmed again (Figs. 5a, b and 6).

The patient presented on December 17, 2021 (at 18–19 weeks of gestation) with abdominal pain and per vaginal bleeding. An emergency obstetric ultrasound revealed no cardiac activity.

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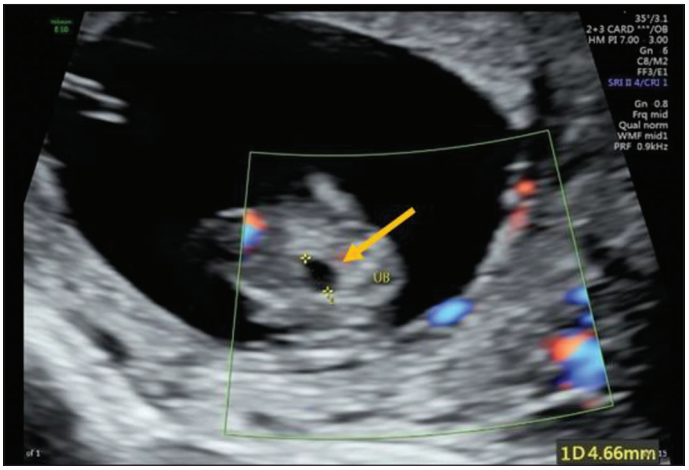

Figure 1: Images of first trimester anomaly Scan (13 weeks). Ultrasound with color Doppler in axial view at the level of the urinary bladder. Arrow showing non-visualization of the umbilical arteries on either side of the urinary bladder

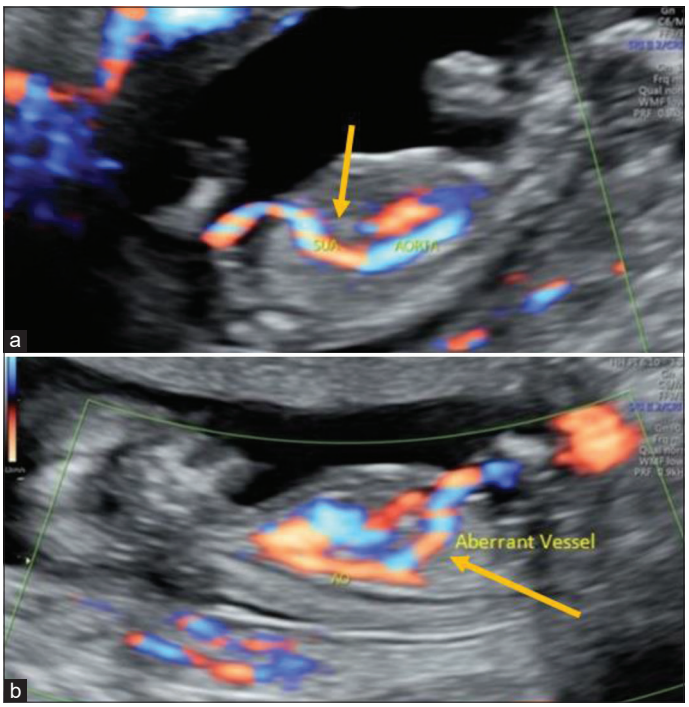

Figure 2: (a and b) ultrasound with color Doppler in sagittal view. Arrows showing an aberrant, large vessel arising from the aorta tracking anteriorly towards the umbilical cord. Beyond its origin, the aorta was hypoplastic

Significant scalp and body wall edema were seen. A diagnosis of intrauterine fetal demise was made (Fig. 7a-c).

The patient was referred back to the obstetrician and, by induction of labor, the abortus was delivered on the same day. Karyotyping examination of the abortus did not reveal any abnormality.

DISCUSSION

Usually, two arteries and one vein are seen in the human umbilical cord. In 0.5–1% of cases, one of the two umbilical arteries may

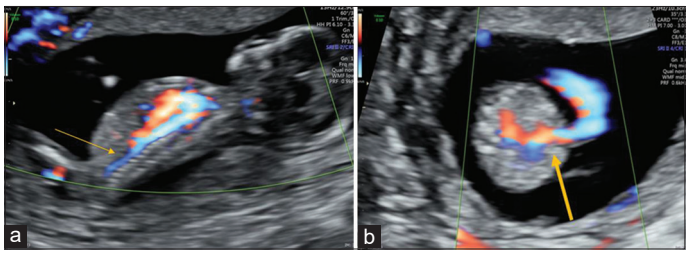

Figure 3: (a) ultrasound color Doppler in sagittal view. Arrow showing hypoplastic distal aorta; (b) ultrasound colour Doppler in axial view. Arrow showing single umbilical artery in a cranial location

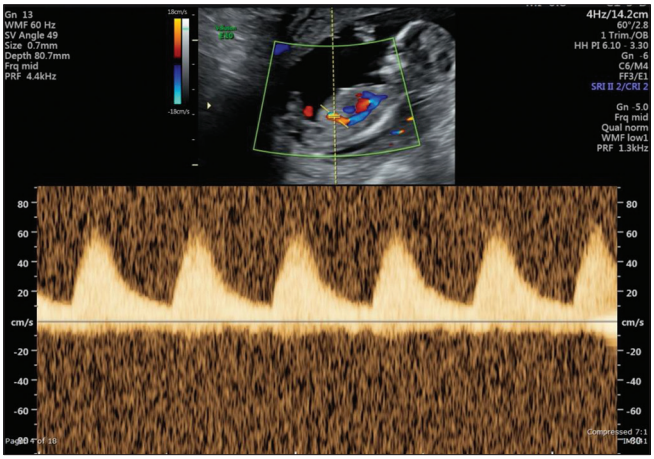

Figure 4: Ultrasound with spectral Doppler in sagittal view. A large single umbilical artery was seen continuing in an anterior direction toward the umbilical cord, showing arterial waveform on spectral Doppler

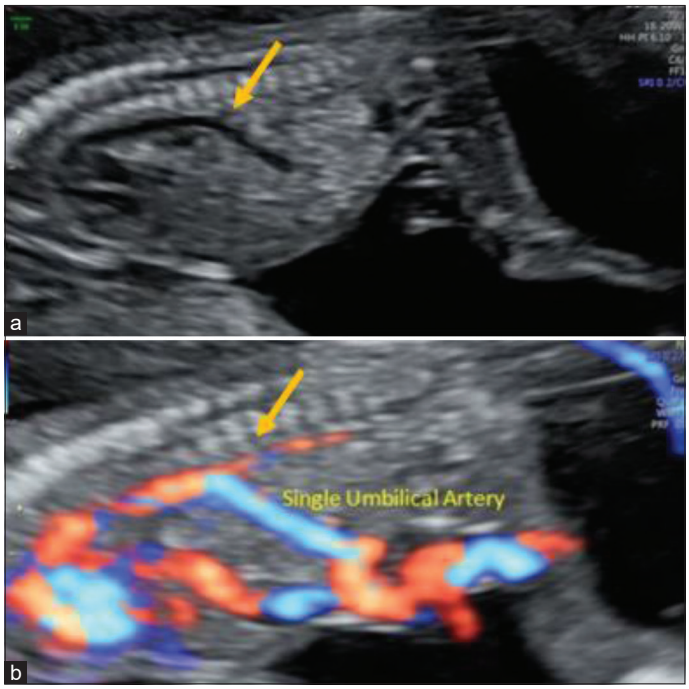

Figure 5: (a and b) images of second trimester scan (16–17 weeks). On follow-up, ultrasound done at 16 weeks, on B mode and color Doppler imaging in sagittal view. Arrows showing an abrupt narrowing of the aorta in the region of the mid abdomen distal to the origin of the umbilical artery

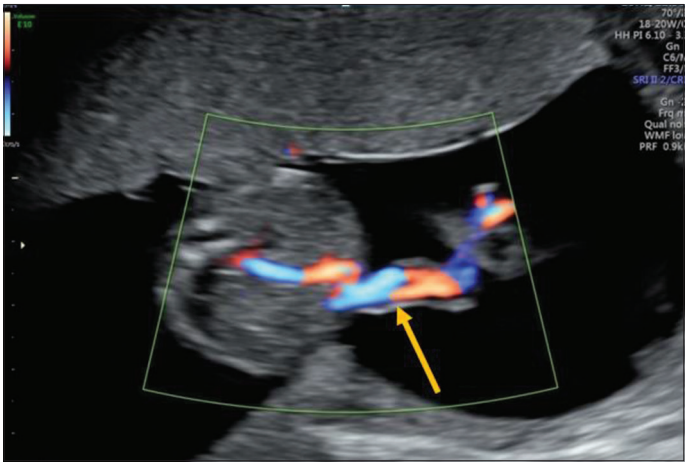

Figure 6: Ultrasound color Doppler in axial view. On follow up, ultrasound done at 16 weeks, arrow showing single umbilical artery in a cranial location

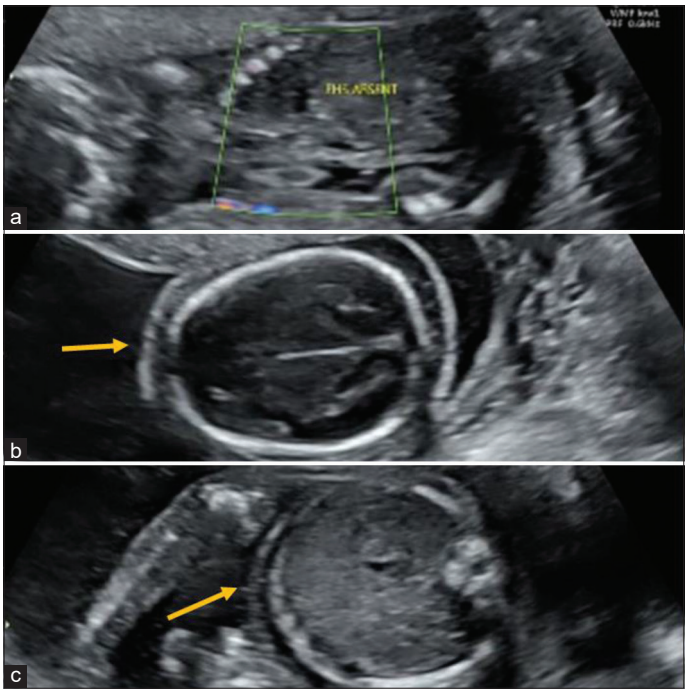

Figure 7: Images of second-trimester scan (18–19 weeks). Ultrasound and color Doppler. Arrows showing absent fetal cardiac activity, scalp edema, and abdominal wall edema – intrauterine fetal demise

Table 1: Differential diagnosis of single umbilical artery [9]

Single umbilical artery type I	Single umbilical artery type II
Most common – 98%	Less common – 1–1.5%
Origin of single umbilical artery is from left or right iliac artery	Origin of single umbilical artery is from aorta/superior mesenteric artery
Usually associated with anomalies of the central nervous system, acardia, lower genitourinary tract	Usually associated with anomalies such as caudal regression, anal agenesis, and sirenomelia

not be present, leading to SUA syndrome. Multiple theories and hypotheses have been proposed for SUA, it has been accepted that SUA syndrome is due to atrophy or atresia of one of the normally existing umbilical arteries [7]. As per a study done by Blackburn and Cooley, related to the developmental etiology, four types of SUA have been hypothesized [1]. In the case of Type II SUA syndrome, the umbilical cord has two vessels – one artery and one vein. The umbilical artery is of vitelline origin rather than the usual allantoic origin. Rather than arising from the common iliac artery, in Type II SUA syndrome, the umbilical artery is seen arising from the SMA. Type II SUA is associated with multiple fetal malformations and abnormalities, including sirenomelia, caudal regression, and anal agenesis. Very rarely, normal fetal development has been reported in Type II SUA [1]. The classification listing the types of SUA is theoretical and of "academic interest." More than 98% of the reported cases are Type I SUA. Only a small fraction of 1.5% of cases is Type II. For all practical purposes, Type III or IV SUA syndrome is essentially not known [1].

As per a study done by Nalluri *et al*., a persistent vitelline artery is seen tracking along an abnormal course upward and backward between the coils of the intestine (instead of the usual course of downward and backward toward the urinary bladder). Doppler ultrasound of the umbilical arteries is a diagnostic tool for persistent vitelline arteries. The prognosis is poor as multiple malformations are seen in the caudal body. Whenever there is a vitelline origin of umbilical arteries rather than the allantoic origin, all the blood from the abdominal aorta reaches the placenta through this SUA, resulting in a vascular steal phenomenon. Due to this steal phenomenon, the amount of blood circulating through the distal aorta is minuscule, resulting in caudal regression. Thus, further evaluation for caudal regression on an antenatal scan becomes essential if the presence of a vitelline origin of an umbilical artery is noted [2].

As per Ronni and Yaron [3] and Stevenson and Hall [4], normal fetal development was rarely observed in a fetus with Type II SUA. As reported by Loh *et al*. and Prust and Abouatme, if a fetus with Type II SUA is born, the neonate can suffer from acute intestinal obstruction, intra-abdominal hemorrhage, or recurrent intestinal pain. Even if the neonate appears asymptomatic, the umbilical artery can form an abnormal peritoneal band extending from the posterior aspect of the umbilical ring to the SMA. The presence of this peritoneal band can cause the loops of the intestines to twist, resulting in an intestinal volvulus and eventual obstruction [5,6]. In a study described by Postoloff, which was based on surgical findings, three types of persistent vitelline arteries are seen. One is a persistent band between the anterior abdominal wall and ileal mesentery, one is present around the Meckel's diverticulum, and the last is a free-hanging cord. Whenever a pediatric surgeon is operating on a volvulus in a neonate, knowledge of these abnormal peritoneal bands is essential [8].

With only a few case reports in literature, the prognosis of fetuses with Type II SUA is generally poor, especially when combined with other congenital anomalies. Table 1 shows the differential diagnosis of SUA [9].

CONCLUSION

Very few cases of this abnormality have been described in the literature. Of these, most were detected in the second trimester.

On review of the literature, this is probably one of the first cases detected during the first-trimester anomaly scan. Our case report will help in the early detection of this anomaly which often has an unfavorable outcome.

AUTHOR'S CONTRIBUTORS

Early detection of Type II Single Umbilical Artery in the first-trimester anomaly scan, followed by subsequent scan at 16–17 weeks which confirmed the findings.

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