Sonographic Diagnosis of Amniotic Band Syndrome and its Risk Factors

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Abstract: Objective: The aim of this study was to describe our experience with amniotic band syndrome (ABS), define specific sonographic characteristics and common features. Methods: Patients diagnosed with ABS underwent detailed ultrasound evaluation at the time of diagnosis and during follow-up. Their ultrasound examinations and medical records concerning the current pregnancy and past medical records were analyzed. Results: Ten pregnancies were diagnosed with ABS. Most pregnancies were diagnosed at the beginning of the second trimester. Two cases were bichorionic twin pregnancies involving one of the fetuses and these were the only women who continued their pregnancies to term. The other eight cases with ABS chose to terminate their pregnancies. One pregnancy was conceived following trachelectomy. We found a significantly higher rate of prior uterine surgeries (p=0.008) in patient with ABS compared to control. Conclusions: ABS diagnosed in early pregnancy can be a sporadic event. However, there is a higher risk of ABS in pregnancies preceded by uterine procedures.

Keywords: amniotic band syndrome, ultrasound, previous uterine surgery

1. Introduction

Amniotic band syndrome (ABS) is a rare congenital disorder caused by entrapment of fetal parts by fibrous amniotic bands, resulting in devastating cosmetic and functional disability. Incidence varies from one in 1200 to one in 15000 live births. The hallmark of ABS is the presence of fibrous bands; however, the precise mechanism of amniotic band formation is still unknown. Several risk factors for ABS have been suggested, such as smoking, drug use, maternal hyperglycemia, amniocentesis and high altitude. The aim of this study was to describe our experience with ABS diagnosis in a tertiary center ultrasound unit, define specific sonographic characteristics, risk factors and common features of this syndrome.

2. Methods

This retrospective study was conducted at a tertiary care centre over a period of 14 years. All patients with diagnosis of amniotic band syndrome were included in the study. Patient’s past medical and obstetrical history was collected. Data details of ultrasound at the time of diagnosis and follow up was retrieved. All deliveries in the past eight years were used as control (86686 deliveries). Statistical analysis was performed by a two-tailed chi-square analysis with Yate’s correction. The study was approved by the local Institutional Review Board committee.

3. Results

Out of all patients included in the study, 10 patients were diagnosed with amniotic band syndrome. Eight patients were diagnosed with ABS during early second trimester (13–15 weeks) at the first ultrasound scan. Two other cases were diagnosed during late second trimester and early third trimester (22 and 27 weeks). Eight patients discontinued the pregnancy. Two of the 10 continued the pregnancy to term. Both of which were twin pregnancies involving one of the fetuses. Most of the anomalies observed in this study were limb deformities. Other anomalies like cleft lip and cleft palate were also seen in some cases. We compared our study group to all 86686 deliveries in the previous eight years. In the study group, five out of the 10 patients had prior uterine surgery.

Table 1: Characterization of all cases included in this study

<table>
<thead>
<tr>
<th>Gestational week</th>
<th>Obstetrical history</th>
<th>Fetal finding</th>
<th>outcome</th>
<th>Previous uterine surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. 14 G3P2</td>
<td>Amputation of hand</td>
<td>TOP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. 13 G1P0</td>
<td>Hands confined by amniotic band</td>
<td>TOP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. 14 G3P2</td>
<td>Foot and hand confined by amniotic band</td>
<td>TOP</td>
<td>CS</td>
<td></td>
</tr>
<tr>
<td>4. 15 G2P0</td>
<td>Deformation of hand</td>
<td>TOP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. 27 G6P5</td>
<td>Amputation of hand</td>
<td>NVD at 37 weeks</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. 14 G1P0</td>
<td>Absence of fingers 2-4 in both hands</td>
<td>TOP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. 15 G2P0A1</td>
<td>Cleft lip and palate, foot and hand confined by band</td>
<td>TOP</td>
<td>Laparoscopic myomectomy</td>
<td></td>
</tr>
<tr>
<td>8. 13 G2P1</td>
<td>Fetus confined by amniotic bands</td>
<td>TOP</td>
<td>Radical trachelectomy</td>
<td></td>
</tr>
<tr>
<td>9. 22 G3P2</td>
<td>Cleft lip and amputated hands</td>
<td>TOP</td>
<td>Previous CS</td>
<td></td>
</tr>
<tr>
<td>10. 15 G4P2A1</td>
<td>Absence of distal phalanx in fingers 2-3 in left hand</td>
<td>CS at 38 weeks</td>
<td>CS</td>
<td></td>
</tr>
</tbody>
</table>
Three patients had prior cesarean sections (CS), one patient had a laparoscopic myomectomy and another pregnancy was conceived following radical trachelectomy. The rate of previous uterine surgery in women with ABS was significantly higher than in the control group (50 versus 14.9%, p<0.008). In three cases, a small vestige in the distal part of the amputated limb was reported (Figure 1) and it was visible after delivery (figure 2).

4. Discussion

Amniotic band syndrome can cause malformations that range from mild deformities to severe anomalies that are incompatible with life. The most common defects associated with ABS are limb defects such as focal constrictions, amputations, pseudosyndactyly, and brachydactyly. An amputation with the distal bone protruding beyond the soft tissue at the site of the amputation is diagnostic of ABS. The two main theories regarding the pathogenesis of ABS are known as the extrinsic and intrinsic models. The most accepted etiology for ABS is the one proposed by Torpin. He proposed a sequence mechanism originating in rupture of the amniotic sac, followed by extrusion of fetal part into the chorionic cavity and resulting in fetal anomalies due to vascular compression by the amniotic bands. Other authors suggested that ABS is formed by vascular disruption and conditions associated with relative hypoxia, such as smoking and living in high altitude, have been identified as risk factors for ABS.

Orioli et al. studied an increased incidence of ABS in the population living at a high altitude, in primipara, in women with a history of febrile illness in the antenatal period, and in women with a history of vaginal bleeding in the first trimester. The pathogenesis of ABS is thought to be disruption of amnion allowing the embryo to enter the chorionic cavity resulting in entanglement of fetal parts creating various malformations. When any of the malformation involving craniofacial region is seen ABS should be suspected while doing the ultrasound. A search for amniotic band (although not diagnostic) should also be made in ultrasound.

In our study we found that history of prior uterine surgeries (50%) is associated with high risk of ABS in the subsequent pregnancy. Previous uterine surgery leading to uterine vascular compromise (placental hypoxia) may be the cause. This study also emphasize the importance of ultrasound examination in first trimester specially in women with history of uterine surgery.

5. Conclusion

We found that there is a higher risk of ABS in previous uterine surgeries and also it can be diagnosed in ultrasound by characteristic fetal malformations involving craniofacial region, trunk, extremities alone or in combination.

References


