

Case Report

Amniotic Band Syndrome After Cervical Cerclage

Hakan KANIT, Dilek ASLAN, Nilgün DİCLE, Gülçin UĞUREL

SSK Ege Doğumevi ve Kadın Hastalıkları Eğitim Hastanesi, Perinatoloji ve Patoloji Bölümleri - İzmir

SUMMARY

AMNIOTIC BAND SYNDROME AFTER CERVICAL CERCLAGE

Background: The amniotic band syndrome is a rare collection of extremely variable fetal anomalies. The type of malformations depends upon the period of gestation during which the bands develop. The possible role of amniocentesis and CVS in the aetiology of amniotic band syndrome has been questioned before in the fact that amniotic irritation or mechanical destruction of fetal tissue.

Case: We present a case with amniotic band syndrome in which the diagnosis is reached with prenatal ultrasonography at 17th gestational week. Ultrasonographic examination of the fetus showed marked defect of the abdominal wall and chest with dislocation of the intestines, liver and heart, as well as posture deformity and multiple amniotic bands. History of the case revealed that, cervical cerclage had been performed at the first trimester, because the patient had had multiple second trimester losses. The patient underwent termination of pregnancy.

Conclusion: Presented case is the first described case for the association of the amniotic band syndrome and cerclage. Case will be discussed according to the possible relationship; and proposed pathogenesis will be evaluated in that way with the reference to the literature.

Key Words: Cerclage, Amniotic Band Syndrome.

ÖZET

SERVİKAL SERKLAJ SONRASI AMNİYOTİK BANT SENDROMU

Konunun önemi: Amniyotik bant sendromu nadir ve oldukça farklılık gösteren bir fetal anomaliler topluluğudur. Malformasyonların tipi, bantların geliştiği gestasyonel döneme bağlıdır. Amniyotik bant sendromu etiolojisinde amniyosentez ve koryonik villus örnekleme'nin rolü, amniyotik irritasyon ve fetal dokunun mekanik hasarı yönünden daha önce sorgulanmıştır.

Olgu: Gebeliğin 17. haftasında prenatal ultrasonografi ile tanı konulan bir amniyotik bant sendromu olgusunu sunuyoruz. Fetal ultrasonografik incelemede abdominal ve torakal duvarda geniş defekt, barsak, karaciğer ve kalp gibi organların bu açıklıktan amniyotik kaviteye dislokasyonu ve postür deformitesi ile çok sayıda amniyotik bantların varlığı tespit edildi. Öyküden, ikinci trimester kayıpları nedeniyle bu gebeliğin ilk trimesterinde servikal serklaj uygulandığı öğrenildi. Amniyotik bant sendromu tanısıyla gebelik sonlandırıldı.

Sonuç: Sunulan vaka, amniyotik bant sendromu ve serklaj birlikteliği olarak ilk tanımlanmış vakadır. Bu yayında, amniyotik bant sendromu-serklaj olası ilişkisi, patogeneze ilişkin literatür ışığında tartışılacaktır.

Anahtar Kelimeler: Serklaj, Amniyotik bant sendromu

The amniotic band syndrome (ABS) has been reported to occur in between 1:15,000 and 1:1200 live, and, 1- 2 % malformed births (1). This syndrome is a rare collection of malformations proposed to be a sequela of intrauterine rupture of the amnion; results in an unpredictable constellation of fetal anomalies (2, 3). Mesodermic amniotic bands resulting from rupture, attach to fetal parts abraded by close contact with the chorion, causing amputations or constriction deformities as the fetus

continues to grow (3). The anatomic region affected and the complexity of the defects are related to the timing of the rupture and the location of the bands (4, 5). Clinical picture of ABS varies enormously; the most innocent type includes only constriction band related skin remarks. Association with an anomaly incompatible with life is also not uncommon in ABS.

CASE

A 28- years old woman has admitted to SSK Aegean Maternity and Women' s Health Teaching



Figure 1. Dislocation of abdominal organs from the marked defect of abdominal wall. a) Stomach and liver, b) Intestines and kidney

Hospital, Department of Perinatology, for the second level ultrasonographic evaluation at 17th gestational week. Obstetric history of the patient was gravida: 5, parity: 1, and three second trimester losses. Because of the cervical insufficiency, cervical cerclage (Mc Donald style) had been performed in another center at the first trimester. History was negative for any drug consumption, tobacco, or ethanol. Family history was also negative for anomalies. Pathologic and cytogenetic evaluation of the former aborted fetuses were normal.

Ultrasonographic examination of the fetus showed marked defect of the abdominal wall and chest with dislocation of the intestines, kidneys, liver and heart, as well as posture deformity and multiple amniotic bands. Cytogenetic evaluation of the fetus via cordocentesis was normal. The patient was discussed at the Ethic Committee, and resultantly underwent the termination of pregnancy. Diagnosis was confirmed with the postmortem evaluation. Additionally, lumbar meningocele was detected. On the back of the fetus, there was a fibrous amniotic band remnant lying vertically above

the vertebral line. Further evaluation revealed malrotation of lower extremities together with the dorsal vertebral kyphosis. The final diagnosis was ABS.

DISCUSSION

Several theories regarding the patho-physiology of ABS have been proposed. After the work of Streeter in 1930, the causative role of the amniotic bands was discounted and the malformations, both the bands and the associated absence deformities, were thought to result from tissue necrosis, probably on a vascular basis (3). Abnormal histogenesis was thought to be the primary event resulting in tissue necrosis and faulty embryogenesis (3). A limitation of Streeter's theory, however, is its focus on limb deformities without attempts to explain craniofacial, visceral, or other abnormalities.

In 1960's works of Torpin made a modified form of the amniotic band theory, exogenous theory, which is now most widely accepted (4, 5). Mesodermic amniotic bands resulting from rupture attach to fetal parts abraded by close contact with the chorion, causing amputations or constriction deformities as the fetus continues to grow (4, 5). The variability in defect size and severity has been attributed to differences in timing of the amniotic rupture. Torpin's exogenous theory was further supported by laboratory studies describing the teratogenic effects of amniocentesis in mice and rats, in which experimental withdrawal of amniotic fluid from embryonic rodents caused multiple malformations (6, 7).

Amniocentesis and chorionic villi sampling (CVS) has been implicated, and studies in animal models show an increased incidence of anomalies related to ABS in association with prenatal amniocentesis and CVS (7-9). Proposed mechanism in these invasive interventions is amniotic irritation or amniotic injury. Ossipoff and Hall reported an increased incidence of ABS associated with maternal abdominal trauma (1). One case report describes ABS related to an incompetent cervix (10). Strauss suggested intensive ultrasound evaluation including Doppler studies in patients with augmented risk due to invasive prenatal procedures to exclude ABS (11).

Presented case is the first case for the association of the ABS and cervical cerclage. The possible relation might be explained in the same way with those in amniocentesis and CVS. Further observations can stimulate the research on the subject.

REFERENCES

1. Ossipoff V, Hall BD. Etiologic factors in the amniotic band syndrome: a study of 24 patients. *Birth Defects* 1977; 13: 117-32

2. Streeter GL. Focal deficiencies in fetal tissues and their relation to intra-uterine amputation. *Contrib Embryol* 1930; 22: 1- 44
3. Torpin R. Amniochorionic fibrous strings and amniotic bands. *Am J Obstet Gynecol* 1965; 91: 65- 75
4. Torpin R. Fetal malformations caused by amnion rupture during gestation. Springfield, IL, Thomas, 1968
5. Higgenbottom MC, Jones KL. The amniotic band disruption complex: timing of amniotic rupture and variable spectra of consequent defects. *J Pediatr* 1979; 95: 544- 9
6. Hesfitz SA. Strangulation of the umbilical cord by amniotic bands: report of 6 cases and literature review. *Ped Path* 1984; 2: 285- 304
7. Kino Y. Clinical and experimental studies of the congenital constriction band syndrome, with an emphasis on its etiology. *J Bone Joint Surg* 1975; 57: 636-43
8. Christiaens GC, Van Baarlen J, Huber J, Leschot NJ. Fetal limb constriction: a possible complication of CVS. *Prenat Diagn* 1989; 9: 67- 71
9. Boyd PA, Keeling JW, Selinger M, Mackenzie Z. Limb reduction and chorionic villus sampling. *Prenat Diagn* 1990;10: 437- 41
10. Sachdev R, Ibanex I, Gabriel J, Navarro C. Amniotic band syndrome associated with an incompetent cervix. *Am J Obstet Gynecol* 1984; 148: 243- 8
11. Strauss A, Hasbargen U, Paek B, Bauerfeind I, Hepp H. Intra-uterine fetal demise caused by amniotic syndrome after standard amniocentesis. *Fetal Diagn Ther* 2000; 15: 4- 7