A Case Report of Prenatally Diagnosed Duplication Cyst Located at Posterior Mediastinum

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Abstract

Background: Duplication cysts originated from gastrointestinal system (GIS) are rare anomalies (1/4500) and these cysts are most commonly encountered from ileum. In this case report, differentiation of cystic lesions located at posterior mediastinum and evaluation of duplication cyst in the view of literature is aimed.

Case: A pregnant women at 28 week of gestation was referred to our clinic as fetus presenting with a cystic lesion located at posterior mediastinum which was suspected as diaphragmatic hernia. In the evaluation of fetal thorax, normal diaphragm and a smooth cystic structure located in the front of the descending aorta and in the right side of posterior mediastinum were seen in which it was thought to be a duplication cyst. In addition to the above ultrasonographic features, single umbilical artery was observed. After vaginal delivery, surgical operation was performed and pathological evaluation of the cyst confirmed prenatal diagnosis of intestinal duplication cyst.

Conclusion: Cooperation of obstetrician, geneticist and thoracic surgeon during evaluation of such cases has an importance on understanding of prenatally diagnosed disorder by family and planning of postnatal operations.

Keywords: Duplication cyst, ultrasonography, fetus.

Prenatal tanısı konmuş, posterior mediastinal yerleşimli enterik duplikasyon kist olgusu

Amaç: Gastrointestinal sistem (GİS) kaynaklı duplikasyon kisti, konjenital olarak nadir görülür (1/4500) ve en sık ileumdan orjinini alır. Olgu sunumunda posterior mediastene yerleşen kistik lezyonların ayırımının yapılması ve literatür eşliğinde duplikasyon kistinin değerlendirilmesi amaçlanmıştır.

Olgu: 28. gebelik haftasında, sağ posterior mediastende yer alan kistik oluşum sebebi ve diafragma hernisi ön tanısı ile kliniğimize sevk edilen olgunun ultrasonografi ile fetal toraksın değerlendirilmesinde; diafragma konturlarının düzenli olduğu görüldü. Sağ posterior mediastende, inen aorta önüne yerleşen düzgün konturlu kistik oluşumu nedeniyle duplikasyon kisti düşünüldü. Ek ultrasonografi bulgusu olarak, tek umbilikal arter tespit edildi. Doğumdan sonra yenidoğana yapılan operasyon sonucu elde edilen materyalin patoloji değerlendirmesinde; prenatal tanı, mediastinal yerleşimli intestinal duplikasyon kisti olarak doğrulandı.

Sonuç: Obstetrisyen, genetik uzmanı ve toraks cerrahının işbirliği ile yapılan değerlendirme; prenatal tanıların aile tarafından anlaşılması ve olası postnatal operasyonların değerlendirilmesi açısından önem taşımaktadır.

Anahtar kelimeler: Duplikasyon kisti, ultrasonografi, fetüs.

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Background

Duplication cysts originating from gastrointestinal system (GIS) are anomalies which are rarely seen as congenital (1/4500) and which proceed light male dominance (1.2:1) in neonatal period. It can be observed in anywhere at GIS and less than 2% of it settles into posterior mediastinum. GIS duplication cysts are mostly result from ileum.[1,2] Common qualities of duplication cysts are reported by Ladd as; a- including muscle layer, b- including epithelial structure having intestinal system, c- being hang on intestinal system by a part of growing structure.[3]

GIS duplication cysts are clinically and pathologically named as; 1. Enteric duplications and cysts (covered by intestinal epithelium), 2. Bronkogenic cysts (covered by respiratory epithelium), 3. Neuroenteric cysts (related with vertebral anomaly or connected with nervous system).[4,5] The situation that duplications belonging to intestinal system have many forms can not explain the theory of one embryological formation. Problems occurring in simultaneous diverticulization of notochord with GIS, not exposing to regression of intestinal diverticulization which takes place in normal embryological are shown as the reasons.[5,6]

Single umbilical artery (SUA) is formed of nonexistence of one artery of umbilical cord which is normally formed of two arteries and one venous. Its incidence frequency is reported as 0.9-1.2% in white people.[7,8] SUA may be monitored together with structural and chromosome anomalies. While chromosome anomaly risk is low in isolated SUA cases, aneuploid monitoring chance increases in cases by additional ultrasonographic diagnosis.[9,10]

Case

The pregnant who was 28 years old G1 P0 and on 28th gestational week was dispatched to Department of Obstetrics and Gynecology of Medical Faculty of Sütçüimam University by initial diagnosis of diaphragm hernia due to cystic lesion in right posterior mediastinum which was not echogenic in fetal thorax. No situs anomaly and cardiac pathology was found in fetal echocardiography done for possible anomaly diagnosis which might accompany.[11] In abdomen and thorax determination by ultrasonography; pit of the stomach was not observed but diaphragm contours were monitored regularly. A smooth cystic formation was found about 25x33 mm which placed in front of the descending aorta and right posterior mediastinum at the same level with the heart (Figures 1 and 2). In the determination of cystic structure in colored Doppler, no certain blood flow was found which will ensure it to become bloodshot, it was thought that it was nourished from tissues at periphery of cyst. Lungs were bilaterally monitored in normal echogenity. In addition to ultrasonographic diagnosis mentioned above, single umbilical artery (lack of left umbilical artery) was found in the determination of umbilical cord. Amnion fluid amount was normal. Genetic counseling was given to the family in order to indicate chromosome number and structure failures to be occurred with accompany of ultrasonographic diagnosis and the family decided cordocentesis for determination of fetal karyotype. Karyotype 46 was found as XX. The family decided to carry on the gestation after karyotype determination. No other ultrasonographic pathology was found except previous diagnoses in routine checks in rest of gestation period. However, appearance of stomach was smaller than normal in the rest of gestation period. The pregnant gave a birth which was a 3200 gr female by a normal spontaneous vaginal birth at 39th gestational week and though there was no problem in vital functions of newborn, it was found that respiration sound of right lung could not be heard clearly. Additionally, single umbilical artery existence was verified in the examination of placenta and its additions after birth. In the examinations done to newborn with accompaniment of prenatal diagnoses, no pathology was found except cystic formation in thorax. Magnetic resonance display was done in order to determine the relation of cystic formation with close tissues clearly; prenatal diagnosis was verified by finding a cystic formation about 3x3 cm which was adjacent to esophagus in posterior mediastinum and took place in front of descending aorta and having no relation with vertebra. It was decided to do the operation as a result of consultations done by thorax surgery.[12] In the operation; thorax was entered from fifth intercostal interval by right postero-lateral thoracotomy. Cystic mass which was about 3x3 cm reaching diaphragm by pedicel and which was adjacent to esophagus, attached to thorax wall and extra parenchymal settled in exploration was totally excised (Figure 3). It was found in the pathological examination after operation that

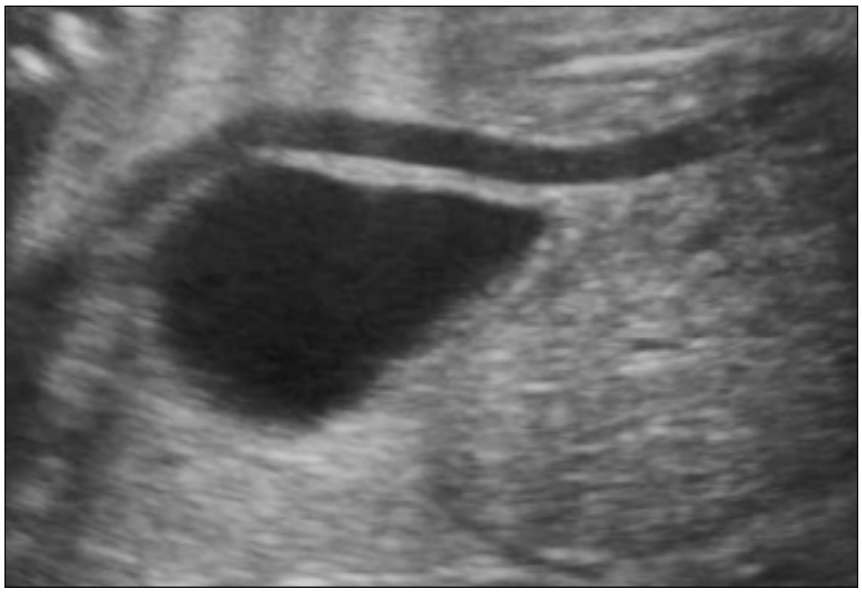

Figure 1. Duplication cyst placing in front of descending aorta and adjacent to diaphragmatic dome.

wall of cystic formation was covered by an epithelial tissue originating from intestine (Figure 4) and prenatal diagnosis was verified as intestinal duplication settling in mediastinum. No complication was found after operation.

Discussion

Mediastinal structures take places in posterior mediastinum as spherical cystic formations which have no relation with near structures.[1] Prenatal diagnosis of GIS duplication cysts is done rarely and most of the diagnoses are done by symptoms related to compression that cyst made in mediastinum.[2] For distinctive diagnosis of cystic formations in posterior mediastinum found in prenatal period; bronkogenic cysts, intestinal duplication cyst, cystic neurogenic tumors, pulmonary sequestration, cystic adenomatoid malformation and diaphragmatic hernia should be thought.[13-16]

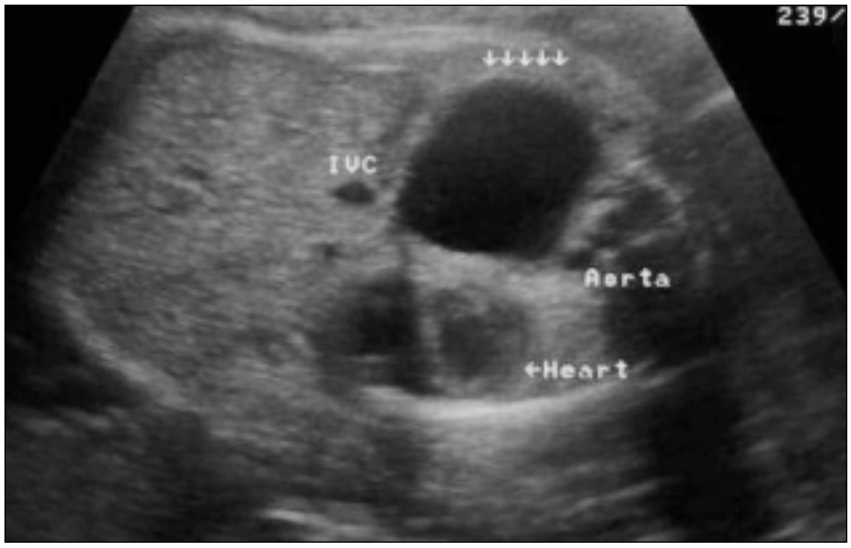

Figure 2. Being at the same level with heart by duplication cyst in coronal section done in heart level of thorax and imitating diaphragmatic hernia as appearance.

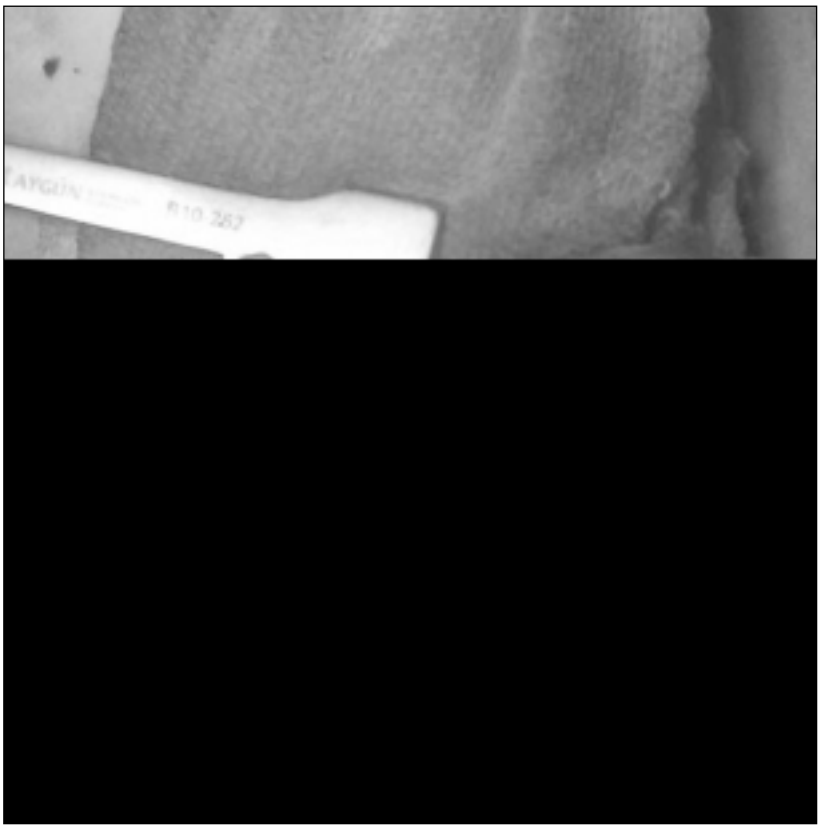

Figure 3. Appearance of duplication cyst during operation.

Cystic adenomatoid structures are a growth anomaly of lung which shows variable qualities from echogenic structures to cystic structures and occurring related to excessive growth of terminal respiratory bronchials. Type 1 especially advances with macrocysts and it may get involved with diaphragm hernia and duplication cyst cases. Cystic adenomatoid lesions are related with tracheobronchial system.[14] Cystic adenomatoid malformation was removed from type 1 diagnosis due to the fact that cystic formation found in our case was taking place in lung periphery at posterior mediastinum and it had no relation with bronchial system. The stomach was not in normal dimension during gestation and diaphragm hernia diagnoses was not thought due to the fact that diaphragm contours were observed clearly in normal structure and intestinal movement in cystic formation could not be observed. We thought that observing stomach small up to therm in abdominal area at normal localization in ultrasonography was related to dysphagia occurred due to possible compression of mediastinal cyst to eusophagus. Moreover, no polyhydroamnios was found in observation of our case up to the therm.

Bronkogenic cysts are frequently cystic formations adjacent to trachea and they may or may not related with traecheo-bronchial system.[15] Bronkogenic diagnosis was also removed due to the fact that bronchial cysts were in the middle of mediastinum and peripheral localization of cystic structure in our case.

In the examination of cystic formation in posterior mediastinum, its relation with vertebra and vertebra anomaly were not found. Trachea was observed at its normal localization. These diagnoses removed neuroenteric cyst diagnosis.[4-5]

We did not think pulmonary sequestration due to causing echogenity increase in lung and frequently taking blood flow from directly aorta.[16]

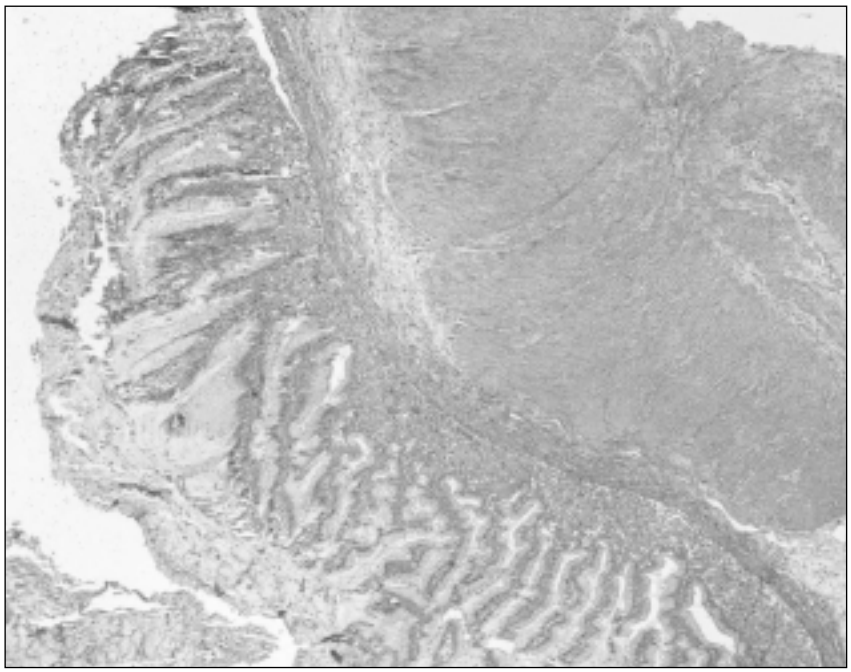

Figure 4. Structures showing duplication cyst is intestinal originated in histological determination.

SUA may be together with structural anomalies (genitor-urinary, central nervous system, cardiovascular, muscle-skeleton system) and chromosome anomaly.[7-10,17] In case of plural gestation and maternal diabetes, incidence frequency of SUA increases.[17] No maternal reason was found which will cause SUA in our case. Though we thought the accidental association of SUA with cystic lesion in posterior mediastinum, cordocentesis with the purpose of prenatal diagnosis was applied due to finding ultrasonographic diagnosis in addition to SUA.[9,10] Karyotype 46 was found as XX.

Duplication cysts may cause deadly complications,[18] thus, full excision of cystic structure is a preferred treatment method. Diagnosis of fetus was done in prenatal period and it was verified by surgery and pathology.

Conclusion

Evaluation done by the cooperation of obstetrician, genetic expert and thorax surgeon is important in terms of understanding of prenatal diagnoses by the family and evaluating possible postnatal operations.

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