Association of Anorectogenital Malformation and Double Aortic Arch in a Child: A Case Report of an Uncommon Coexistence: Original Image

Nadir Bir Anorektogenital Malformasyon ve Çift Aortik Ark Birlikteliği

Vedat BAKAN, MD,[a]
M.Ali KAYGIN, MD[b]

Clinics of [a]Pediatric Surgery,
[b]Cardiovascular Surgery,
Numune State Hospital, Erzurum

Geliş Tarihi/*Received:* 28.07.2008
Kabul Tarihi/*Accepted:* 31.10.2008

This case was presented at the 26th annual meeting of the Turkish Association of Pediatric Surgeons, İstanbul, Turkey, June 16 to 18, 2008

Yazışma Adresi/*Correspondence:*
Vedat BAKAN, MD
Kahramanmaraş Sütçü İmam University,
Faculty of Medicine,
Department of Pediatric Surgery,
Kahramanmaraş
TÜRKİYE/TURKEY
vedatbakan@hotmail.com

ABSTRACT Congenital double aortic arch (DAA) is a very rare vascular anomaly that causes tracheal and /or esophageal compression. A case of recto vestibular fistulae and vertical vaginal septum coexisting with double aortic arch was reported in this paper. Since infancy, the patient was suffering from recurrent airway infections. DAA was diagnosed by computerised tomography. After surgical disruption of the double aortic arch, the respiratory troubles disappeared. This case shows that the vascular ring anomalies should be investigated in patients with anorectal malformations who have recurrent airway infections. Early diagnosis and treatment of such anomalies are important to prevent future morbidity and mortality. To our knowledge, this case is probably the first one in the English literature. The case has been reported in view of its rarity and nature of presentation.

Key Words: Anus, imperforate; aortic arch syndrome

ÖZET Konjenital çift aortik ark (DAA; Double Aortic Arch) trakeal ve/veya özofagial basıya neden olabilen seyrek bir vasküler ring anomalisidir. Bu yazıda rektovestibular fistül vertikal vaginal septum ve çift aortik ark birlikteliği olan bir olgu sunuldu. Hastanın doğduğundan beri tekrarlayan alt solunum yolu enfeksiyonu öyküsü vardı. DAA tanısı bilgisayarlı tomografi ile kondu. Aortik ark divisyonu sonrası hastanın şikayetleri ve tüm semptomlar kayboldu. Tekrarlayan alt solunum yolu enfeksiyonu olan anorektal malformasyonlu hastalar vasküler halka anomalileri açısından araştırılmalıdır. Bu tür anomalilerin erken tanı ve tedavisi morbidite ve mortalitenin önlenmesi açısından önemlidir. Bilgilerimize göre olgumuz, İngilizce literatürde, DAA'ın anorektogenital malformasyona eşlik ettiği ilk olgu olma özelliğini taşımaktadır. Olgu atipik klinik özellikleri ve seyrek görülmesi nedeniyle sunulmuştur.

Anahtar Kelimeler: İmperfore anüs, aortik ark sendromu

Turkiye Klinikleri J Cardiovasc Sci 2008;20(3):232-4

Cardiovascular anomalies occur in about 10-30% of patients with (ARM). The most common cardiovascular anomalies presented in these patients are arterial septal defect, patent ductus arteriosus, tetralogy of Fallot and ventricular septal defect [1]. Double Aortic Arch (DAA) is a very rare congenital vascular anomaly [2], and its association with anorectal malformation has not been previously reported.

Vascular rings are congenital anomalies of the aortic arch complex. These anomalies may be complete (double aortic arch and right aortic arch) or incomplete (innominate artery compression and pulmonary artery sling). DAA is the most frequent anomaly in the complete group [3, 4]. Presenting symptoms correlate with the type of vascular ring. DAA usually presents

with respiratory symptoms due to extrinsic tracheal compression.[3-5,]

Although a wide variety of cardiovascular lesions have been associated with ARM, coexistence of a DAA has probably not been reported previously in English literature.

CASE REPORT

A 20 month-old girl was admitted to our hospital because of ARM presentation. On the physical examination, she had rectovestibular fistulae, vertical vaginal septum, and a right transverse loop colostomy (Figure 1). She had been underwent a right transverse loop colostomy at another hospital in the neonatal period. The child had a history of persistent barking cough and partial responsive to medical treatment since infancy. She had been treated unsuccessfully for suspected bronchitis for several times before coming us. In addition to airway symptoms, she also had a history of choking and dysphagia with solid food.

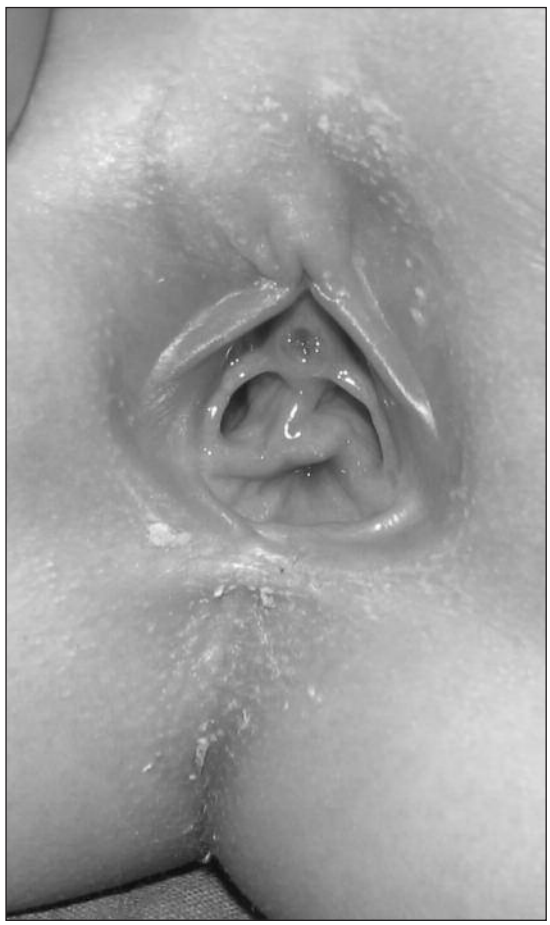

FIGURE 1: Rectovestibular fistulae and vaginal septum.

At admission to our clinics, she was well and had no respiratory symptoms except mild barking cough. There was no positive finding on the respiratory examination. Clinical and echocardiographic assessments of the cardiovascular system were normal.

Leukocyte count was 13,800/mm^3, other laboratory findings, urogenital ultrasound and PA chest X-ray were normal. Posterior sagittal anorectoplasty was performed as a definitive surgery. Both diagnostic vaginoscopy and vaginal operation were refused by parents until marriage due to the cultural and religious causes. Also, the definitive operation of patient had been postponed by parents until the age of 4 because of low socio-economic condition and lack of interest of them. The postoperative course was uneventful. Three months later, her colostomy was closed and the patient was discharged on the 5th postoperative day. Because respiratory symptoms continued postoperatively, a congenital anomaly of the lung was suspected. A contrast-enhanced computed tomographic (CT) scan of the thorax was obtained, which showed DAA encircling and moderately narrowing the trachea and esophagus (Figure 2). Division of the DAA was done by thoracotomy. After surgical disruption of the DAA, the respiratory troubles disappeared.

DISCUSSION

Double aortic arch is the most common forms of complete vascular rings. Although various forms of double aortic arch exist, the common defining feature is that both the left and right aortic arches are present. By the end of the fourth week of embryonic development, the branchial arches have formed between the dorsal aortas and ventral roots. Subsequent involution and migration of the arches result in the anatomically normal or abnormal development of the aorta and its branches. Vascular rings are formed when this process of regression and persistence does not occur normally, and the resulting

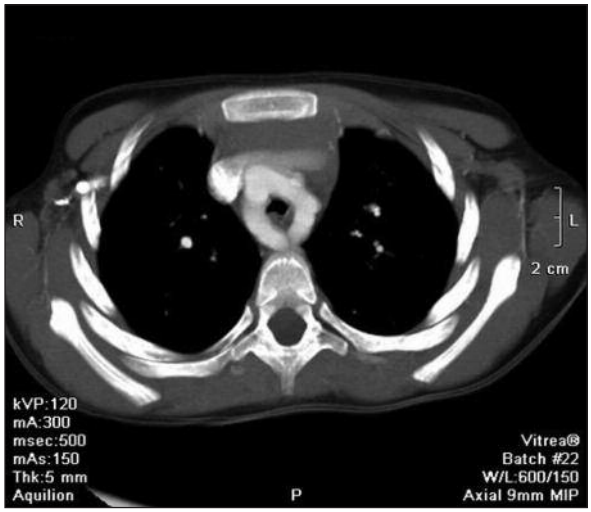

FIGURE 2: CT scan showing the double aortic arch.

vascular anatomy completely encircles the trachea and esophagus. DAA caused by persistence of right and left fourth branchial arches. A Double Aortic Arch occurs when both fourth arches and both dorsal aortas remain present.[2,4]

Patients with vascular rings present with respiratory and/or esophageal symptoms. The symptoms correlate with the type of vascular ring. Presentation is usually within the first 6 months, especially the first month of life.[2,3] In the younger age group, respiratory symptoms and signs such as apnea episodes, cyanosis, stridor, persistent barking cough are predominant, whereas the esophageal symptoms such as dysphagia, reflux, choking and failure to thrive become more apparent when the children begin to eat solid food.[2-4] Aortic arch abnormalities also should be suspected in older children with recurrent bronchitis or pneumonia.[3-5] Our patient had respiratory symptoms due to compression of DAA, predominantly.

Various methods such as plain chest X rays, barium esophagogram, broncoscopy, CT, magnetic resonance imaging are available for diagnosis of vascular rings. Echocardiography is not always reliable in the diagnosis of vascular ring because it sometimes can not visualize the aortic arch clearly .[2-5] Echocardiography has done in infancy in another hospital but it did not helpful in definitive diagnosis of DAA in our case. We diagnosed DAA in our case by CT.

This case shows that the vascular ring anomalies may be seen together with ARM and causes recurrent airway infections. Early diagnosis and treatment of such anomalies are critical to prevent future morbidity and mortality.

REFERENCES

1. Pena A, Levitt MA. Anorectal malformations. In: Grosfeld JL, O'Neill JA, Fonkalsrud EW, Coran A, eds. Pediatric Surgery. 6th ed. Philadelphia, Mosby Inc; 2006. p.1566-89.
2. Devaney E, Ohye R, Bove EL. Congenital heart disease and anomalies of the great vessels. In: Grosfeld JL, O'Neill JA,Fonkalsrud EW, Coran A, eds. Pediatric Surgery. 6th ed. Philadelphia: Mosby Inc; 2006. p.1959-86.
3. Karakoç F, Karadağ B, Kut A, Bakaç S, Dağlı E [Vascular ring as a cause of chronic respiratory symptoms in early childhood]. Turkiye Klinikleri J Pediatr 2000, 9:231-6.
4. Alsenaidi K, Gurofsky R, Karamlou T, Williams WG, McCrindle BW. Management and outcomes of double aortic arch in 81 patients. Pediatrics 2006;118:e1336-41.
5. Bahçeci S, Cengizlier R, Kösebalaban Ö, Önal Z, Çakmak FN [Diagnosis with barium study in etiology of wheezing]. Turkiye Klinikleri J Allergy-Asthma 2000, 2:145-9.