Abstract = Authors describe an autopsy case of a 1 month old female infant with symptomatic vascular ring of the aortic arch. The vascular ring was formed by the right aortic arch and left-sided ductus arteriosus connected to the distal part of left aortic arch. Proximal part of aortic arch was not recognizable. Major branches of the arch were, from before backward, the left common carotid artery, the right common carotid artery, the right subclavian artery, and the left subclavian artery arising from a diverticulum of the aorta into which the left ductus inserted. The intracardiac anomalies of ventricular septal defect and persistent foramen ovale were present. The patient suffered from dyspnea, cough, cyanosis on crying and signs of pneumonia, and they could be explained partly by the tracheoesophageal compression of the vascular ring.

Key words: Aortic arch anomaly, Vascular ring, Aspiration pneumonia

INTRODUCTION

Anomalies of the aortic arch complex form a diverse group in their anatomic and clinical manifestations. Major problems related to these malformations are the anatomic types, compression of trachea and esophagus, and associated cardiac anomalies. The morphologic types and classifications are based on the hypothetical double aortic arch proposed by Edwards (1974). By the morphologic types of the malformed aortic arch system, trachea and esophagus can be compressed and clinical syndrome of “vascular ring” is built up.

There have been reports of various types of associated cardiovascular or visceral deformities (Chi et al. 1965; Berman et al. 1977; Binet et al. 1981; Whitman et al. 1982; Pirtle et al. 1983; Minami et al. 1986; Blatchford et al. 1987). The associated anomalies are also related to the morphologic types.

We recently experienced an autopsy case of a 1 month old infant who died with mild signs of pneumonia. The aortic arch anomaly of the infant was not suspected during premortem period. The major anomaly was a vascular ring, and its retroesophageal segment strongly suggested the symptomatogenic nature of the condition.

CASE REPORT

Clinical findings:

This 43 days old female infant presented with coughing and failure to thrive of 15 days duration. She was born by spontaneous vaginal delivery at the gestational period of 41 weeks. Her birth weight was 3.0 kg and there was no specific perinatal problem. Since 15 days prior to the hospital visit, she had suffered from failure to thrive, dyspnea, cyanosis on crying and cough. She visited a local clinic and was managed conservatively without symptomatic improvement. She was referred to Hanil General Hospital for the further treatment and was admitted to Pediatric ward. Family and past medical histories were unremarkable except maternal history of unidentified drug ingestion during the first trimester of this pregnancy. Her body measurements were below 3rd percentile (Body weight; 3.11 kg, height; 49.2 cm, head circumference;
34.4 cm, chest circumference; 30.4 cm). On physical examination, cyanosis on crying, incomplete cleft palate, grade II systolic murmur along the left lower sternal border, and crackles on bilateral lung fields were found. The liver margin was palpated at 3.5 cm below the right costal margin. Vital signs were relatively stable except mild and persistent tachycardia. The laboratory findings were unremarkable. Chest X-ray revealed hyperinflation of the lungs and ventricular and atrial septal defects were found on echocardiography. EKG findings were those of right ventricular hypertrophy.

She persistently suffered from cough and cyanosis on crying, and under the clinical impression of congestive heart failure, digitalization was started. There was symptomatic relief during two days but the clinical course suddenly deteriorated and she died without specific causative episode. The result of chromosomal analysis was 46, XX pattern.

Pathologic findings:
Postmortem examination revealed retroesophageal left descending aorta causing compression of midesophagus. Ascending aorta was connected to left-sided descending aorta through right aortic arch. To the left descending aorta, left ductus arteriosus was connected. The vascular ring was formed by ascending aorta, right aortic arch, left descending aorta, left ductus arteriosus, and main pulmonary artery, from right to the left. The branches of vascular ring were, from before backward, the left common carotid artery, the right common carotid artery, the right subclavian artery, and the left subclavian artery arising from a diverticulum of the aorta into which the left ductus inserted (Fig. 1, 2). The left ductus was patent on probing but functional closure was demonstrated on silicate casting. On opening the heart, perimembranous ventricular septal defect, measuring 0.8 cm in diameter, was present and foramen ovale was patent. The lungs were slightly consolidated and showed patchy, dusky red discoloration. The microscopic examination of the lungs showed features of bronchopneumonia associated with evidence of aspiration such as intraalveolar squames and unidentified, refractile ovoid foreign bodies. Other
Fig. 2. Superior view of the vascular ring. The tracheoesophageal tree is encircled by a vascular ring formed by both common carotid arteries (LC, RC), right subclavian artery (RS), left subclavian artery (LS), and ductus arteriosus (DA).
PT: pulmonary trunk, A: Aorta

minor positive findings were scattered calcifications in the kidney and left ovary, and bilateral extraadrenal cortical nodules.

DISCUSSION

Bilateral aortic arch system connected to both ascending and descending aorta forms complete vascular ring around the esophagus and trachea. But anomalies of aortic arch causing mechanical compression of the esophagus and trachea are collectively called as "Vascular Ring". Therefore, those with left arch and retroesophageal right subclavian artery or right arch with left ductus connected to descending aorta are included in the broad sense of vascular ring.

Edwards (1974) conceived those malformations derived from a functional double aortic arch with bilateral ductus arteriosus, and categorized aortic arch anomalies into four morphogenetic types by the location of ductus and left descending aorta. Anomalies derived from double aortic arch with left ductus and left descending aorta
are the commonest type and the present case belongs to this category. From the hypothetical double aortic arch system, segmental atresia of certain portion of either right or left aortic arch results in the abnormal aortic arch system. Our case can be classified into right aortic arch with retroesophageal component and the ductus is usually connected to the distal part or ‘diverticulum of left arch. The interruption of the left arch can occur at sites lying either between the carotid and subclavian artery as in the present case or distal to the subclavian branch which remains to be hypothetical. The schematic presentations of the left arch in the present case was summarized in Figure 3.

Associated cardiac anomalies vary by the morphologic types of aortic arch anomalies. Double aortic arch is almost always associated with cyanotic congenital heart disease such as tetralogy of Fallot and transposition of great arteries. Left aortic arch with retroesophageal subclavian artery is frequently associated with tetralogy of Fallot. Right aortic arch is frequently seen in cases of tetralogy of Fallot especially when double ductus arteriosus is present. However, right aortic arch with left ductus connected to left innominate artery and right aortic arch with retroesophageal component (as in the present case) are not frequently associated with cardiac defects. One review on the right arch with retroesophageal component revealed 88% of cases without intracardiac defect (Stewart et al. 1983). The cardiac anomalies in the present case were ventricular septal defect and patent foramen ovale. Though ductus arteriosus could be probed forcefully, nonfilling on silicate casting of the arch strongly suggested its functional closure.

The vascular ring may be symptomatic or asymptomatic, but some of asymptomatic cases in infancy may become symptomatic as the size of the arch increases (Binet et al. 1977). Commonly associated manifestations of the vascular ring include both respiratory and digestive symptoms such as stridor, dyspnea, cyanosis, and feeding difficulties with aspiration, and rarely some types can be a focus of traumatic rupture (Mozzi et al. 1979). Those symptoms are mainly due to the tracheoesophageal compression by anomalous circle of the vascular ring. Retrospectively, cyanosis on crying, cough, and dyspnea in the present case are thought to be the result of tracheoesophageal compression, because her intracardiac anomalies were not of cyanotic nature.

The vascular ring in this case caused no significant hemodynamic derangement, and the condition rarely need surgical correction, unless the symptom is severe enough to be disabling. Division of ductus or fibrous cord at the site of atretic segment will provide loosening of the ring. Final autopsy results couldn’t elucidate the definite cause of death, but the aspiration pneumonia and related processes due to compression of tracheoesophageal tree by the vascular ring would be the most plausible explanation.

Because the condition is not exceptionally rare, the patients with unexplained respiratory or digestive manifestations should be evaluated for the possibility of vascular ring whether they have congenital heart disease or not.

REFERENCES


