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DIRECT COMMUNICATION BETWEEN PULMONARY ARTERY AND LEFT ATRIUM*

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In this article an autopsy finding of a rare congenital anomoly is presented. We found only 28 cases in the literature. Four anatomic variations have been defined, and our case was of type II. The morphogenetic mechanism is a matter for discussion. The previous reports include inadequate pathological examinations. Our histologic findings support the arterial nature of the aneurysmal sac. *Key words: pulmonary artery-left atrium communication, congenital heart disease.*

Direct communication between the pulmonary artery or its branches and the left atrium is a rare anomaly, and only 25 cases had been published as of 1986[1]. Since then three cases (two cases in 1986 and one case in 1989) have been reported[1-3]. The age range of the patients is between one day and 45 years, and only six cases under the age of one year are reported. This anomaly has been classified into four types on the basis of two main features: the presence or absence of an aneurysm in the communication, and the anatomy of the pulmonary venous drainage[4,5]. Our case was compatible with type II.

Because it is a rare congenital anomaly, we present this case with a review of the literature.

Case Report

A three-day-old male baby was admitted to the Neonatal Unit of Erciyes University Faculty of Medicine with complaints of purplish skin color, failure to suck and uneasiness. His past history was noncontributory. There was no consanguinity between his parents. the had three brothers who were alive and healthy.

He was 49 cm in length and his weight was 2800 g. His body temperature was 36 °C. Blood pressure was 60 mmHg by flush method, and pulse was 120 beats per minute. Tachypnea with perioral and peripheral cyanosis was evident. Significant II/IV systolic murmur was heard in the mesocardiac area during auscultation.

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Hemoglobin concentration was 17.7 g/dl. Urinalysis results were within normal limits. Blood gas analysis revealed pH 7.34, pCO_2 45 mmHg, pO_2 25.2 mmHg, HCO_3^- 24.6 mEq/L, O_2 saturation 42.3%, and BE -80.9. The chest roentgenogram showed a double contour on the right border of the cardiac silhouette. The pulmonary vascular markings were decreased. Electrocardiogram revealed spiked p waves at D II.

In the 38th hour of hospitalization, the patient died. Autopsy was then performed:

Macroscopic findings of cardiopulmonary system: Valves were normal. The foramen ovale was open (ostium secundum type). Patent ductus arteriosus was present. The aorta and its branches were normal. The left atrium was dilated and communicated with an aneurysmal sac toward the lower lobe of the right lung. The pulmonary artery was dilated up to 1.5 cm beginning from its exit from the right ventricle and opening to the left atrium which communicated with the aneurysmal sac (Fig. 1). There were three pulmonary veins draining directly into the left atrium below the aneurysmal sac. The left lung consisted of two and the right of three lobes, and there was no pulmonary lobe agenesis. However, there was an aneurysmal sac of 2.5 x 2 x 1 cm dimensions present in the lower lobe of the left lung (Fig. 2).

Fig. 1: Macroscopic appearance of the heart. There is a marked dilatation at the beginning of the pulmonary artery. After the juncture with the left and right branches, the pulmonary artery communicates with the left atrium. The aorta and its branches are normal. Patent ductus arteriosus is also present.

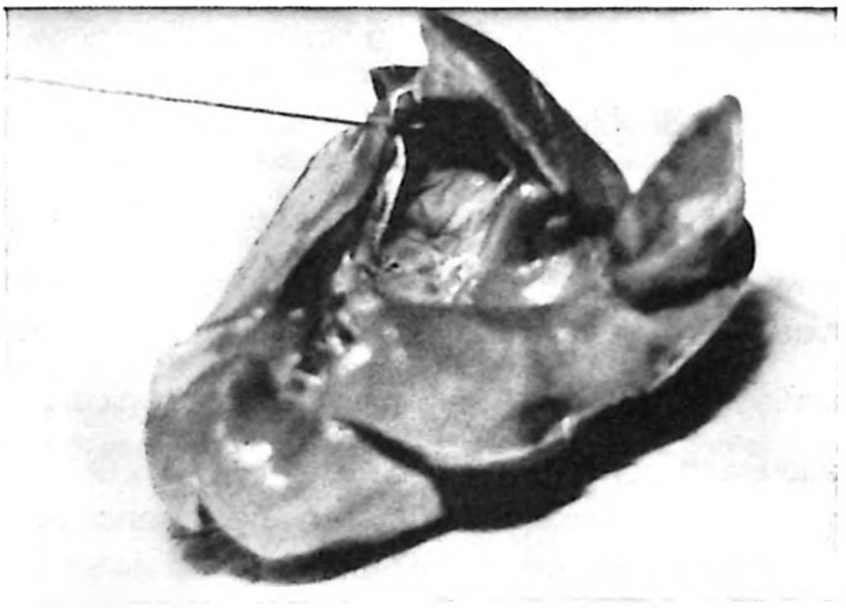

Fig. 2: Macroscopic appearance of the left lung. Note the aneurysmal sac located in the lower lobe.

No pathology was observed macroscopically in the other systems.

Microscopic findings: Congestion due to hypoxia was the only pathology observed in other organs. However, we noted the presence of surrenal ectopia in the testis. Sections prepared from the wall of the aneurysmal sac located in the parenchyma of the lower lobe of the right lung were stained with Voerhoff's elastic stain, Masson's Trichcrom and Hematoxylin-Eosin (Fig. 3). There were no heart muscle fibres in the wall of the sac. The structure was composed of a mixture of connective tissue and smooth muscle fibers. In some sections we clearly observed layers seen in the arterial walls. Furthermore, there were internal and external elastic membranes which led us to suspect that the sac may have originated from an artery.

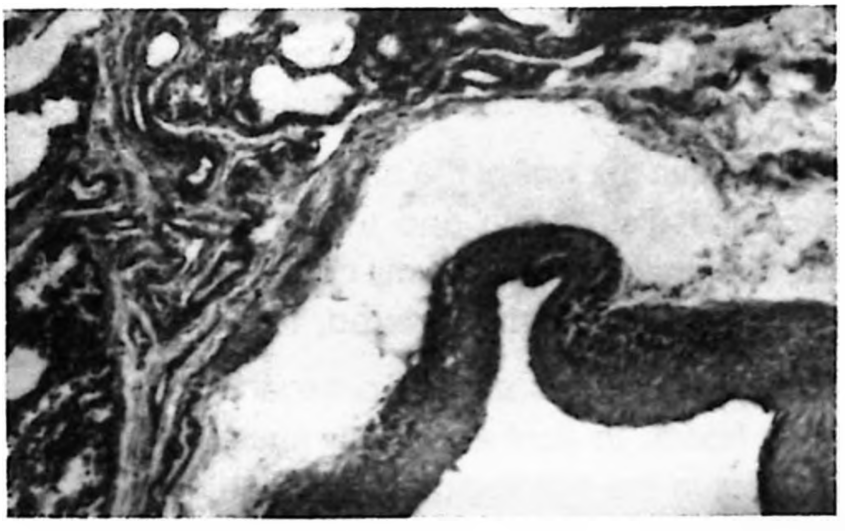

Fig. 3: Microscopic appearance of the aneurysmal sac in the parenchyma of the lower lobe. (H.E. x 40).

Discussion

Direct communication between the pulmonary artery or its branches and the left atrium is a rare anomaly, and only 25 cases had been reported in the literature by 1986[1].

Since then, three cases (two cases in 1986 and one case in 1989) have been reported[1-3]. The age range of the patients reported is one day to 45 years, and only six cases younger than one year have been documented.

Nelson et al.[4] classified the anatomic variations of the communication into three types:

I) Communication with normal veins.
II) Aneurysm with the abnormalities of lobulation and absence of pulmonary veins.
III) All veins draining into the communication.

Ohara et al.[5] added a fourth type to this classification:

IV) Aneurysm with the right veins connected to it.

Our case was compatible with type II. The first case of this type in the literature was presented as a variant of pulmonary arteriovenous fistula in 1961 by Lucas et al[6].

In most of the reported cases in the literature, a surgical approach was chosen rather than an adequate pathological investigation. For this reason the morphogenetic mechanism of the anomaly continues to be a matter of argument. Some have speculated about the persistence of the communication between the pulmonary artery and the pulmonary vein in the first month, when the common pulmonary vein joins the left atrium[1]. Others emphasize the similarity to arteriovenous fistulas originating from agenesis of a pulmonary lobe, the dilated capillary bed giving rise to the communication[1,6]. The presence of abnormal lung lobulation, especially in type II cases, constitutes the starting point of the latter theory. But, as in our case, the existence of normal pulmonary lobulation together with the aneurysm invalidates this theory.

Ohara et al.[5] reported that the wall of the aneurysmal sac can be divided roughly into two layers by the distribution and the population of the elastic fibers. According to this, the inner third of the wall consisted of relatively dense, fibrous connective tissue and regularly multilayered, fine elastic fibers.

The outer two-thirds consisted of a loose collagenous tissue layer with irregular and sparse elastic fibers. In contrast to our case, there were no smooth and skeletal muscle fibers in the histologic examination of the aneurysmal sac. Our findings were mostly different, and led us to consider an arterial nature. Lekuona et al.[1] also stressed that the aneurysmal sac was of arterial origin.

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