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A CASE OF COR BILOCULARE ACCOMPANIED BY POLYSPLENIA

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Bilocular heart or cor biloculare is a rare congenital malformation in which the septum between both the atria and ventricles fail to develop. Consequently, the patient's heart consists of a single atrium, a common atrioventricular canal and valve, and a single ventricle. This malformation may be accompanied by anomaly of great vessels and features of mongolian idiots. Some 50 per cent of the cor biloculare patients are mongolian idiots (3).

An autopsy was recently performed on a case of cor biloculare in the Pathology Department of Tehran University Medical School (autopsy no. 366/49). The following is a brief account of the findings.

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Clinical history

The patient was a 3 $\frac{1}{2}$ year old girl who was brought to the hospital because of dyspnea, cyanosis, and growth retardation. Her birth had been in full term, normal, and without obstetrical trauma. The mother had not suffered any viral disease during pregnancy. The child's dyspnea and cyanosis had appeared from the third month of birth.

Observations

The patient had intense generalized central cyanosis. The jugular vein had pulsation. The thorax had projected forward but the respiratory, urinary, and digestive systems were all normal. Auscultatory findings indicated systolic murmur in grade IV. Systolic murmur could be heard in all four centers. The murmur had radiated toward left axilla. The second heart sound was normal. The liver was not palpable. The spleen and lymph nodes were also normal.

Cardiac globus hypertrophy was noticed in radiography of the chest. The patient's electrocardiogram showed right axis deviation and hypertrophy of the right ventricle. Treatment was based on diagnosis of tetralogy of Fallot. The patient died, however, with intense dispnea and cyanosis.

Necroscopic findings

The anterior and posterior part of the trunk appeared normal, but the strenum and the ribs adjacent to the heart were projected outside. The tip of the fingers were cyonized and enlarged. The superficial lymph nodes and subcutaneous adipose tissues were normal. The body weighted 12,100 grams and was 75.5 cm. long. The head, face, eyes and nose were also normal and without any particular mark.

CARDIOVASCULAR SYSTEM: The pericardium was normal. The heart was enlarged. The right ventricle myocardium was hypertrophied and one centimeter thick (fig. I). The two atria formed a single socket and there was no septum between the atria (fig. I). The pulmonary veins had separately entered the right side of the single atrium (figs. 2 and 3). The superior and inferior vena cava each ended in its normal place toward the right side of the single atrium. The single atrium was connected to a single ventricle by a common atrioventricular canal and valve. The ventricular septum was entirely absent. The ventricles had thus formed an undivided ventricle -- type III of the classification of Van Praagh and associates, 1964 (fig. I). The papillary muscles were numerous and the cordate tendineae of the tricuspid valve was attached to the largest of them (fig. 3). In the single ventricle the aorta occupied a position anterior and the right of the pulmonary artery (i.e., transposition of the great arteries -- d-type of the classification suggested by Brandt and Leibow, 1958; Lev and Rowlatt, 1961: and Van Praagh et al., 1964) (fig. 2).

In the microscopic section of the heart the cells of the myocardium were slightly vacuolized.

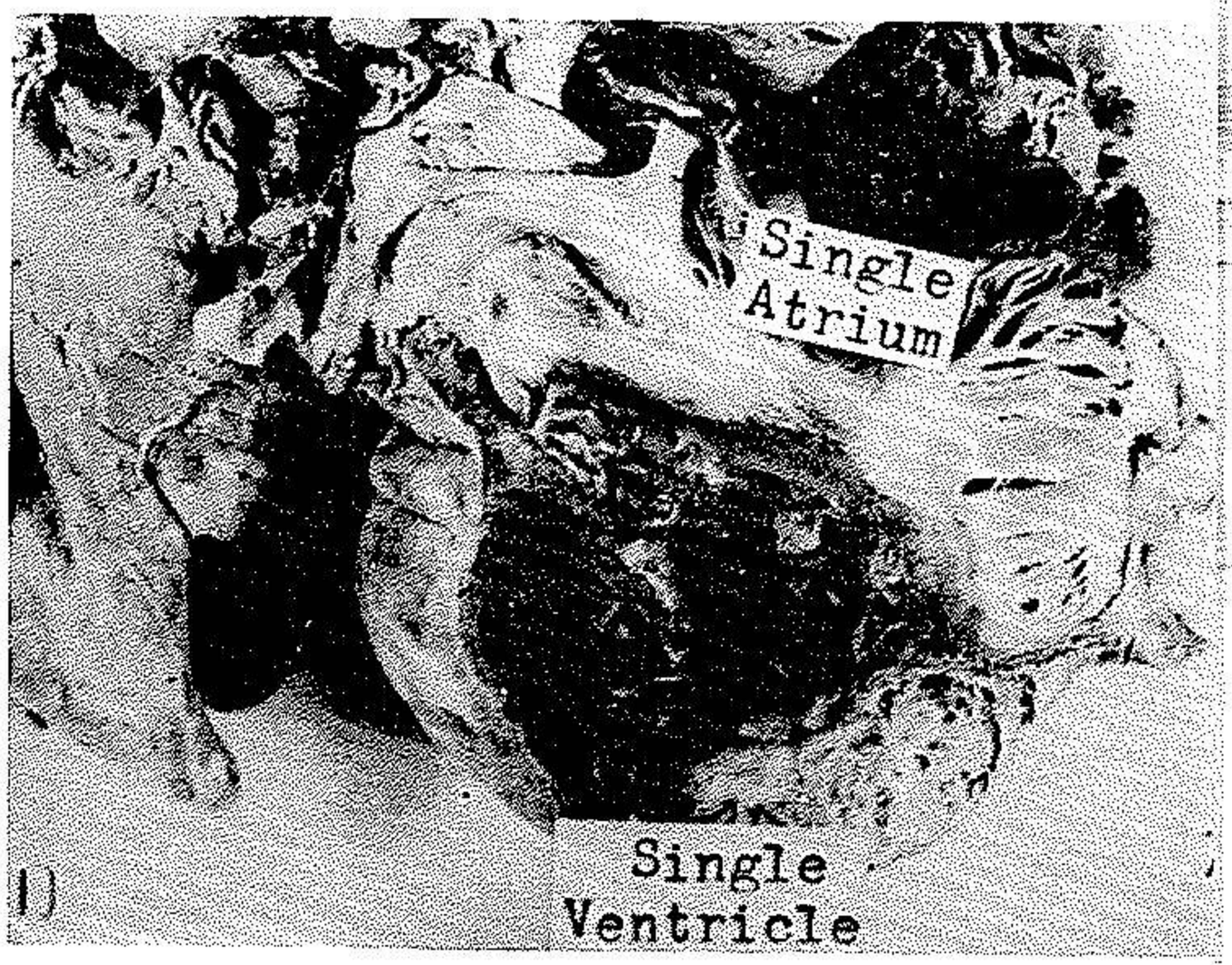

Fig. 1. The patient's heart with a single ventricle and a single atrium.

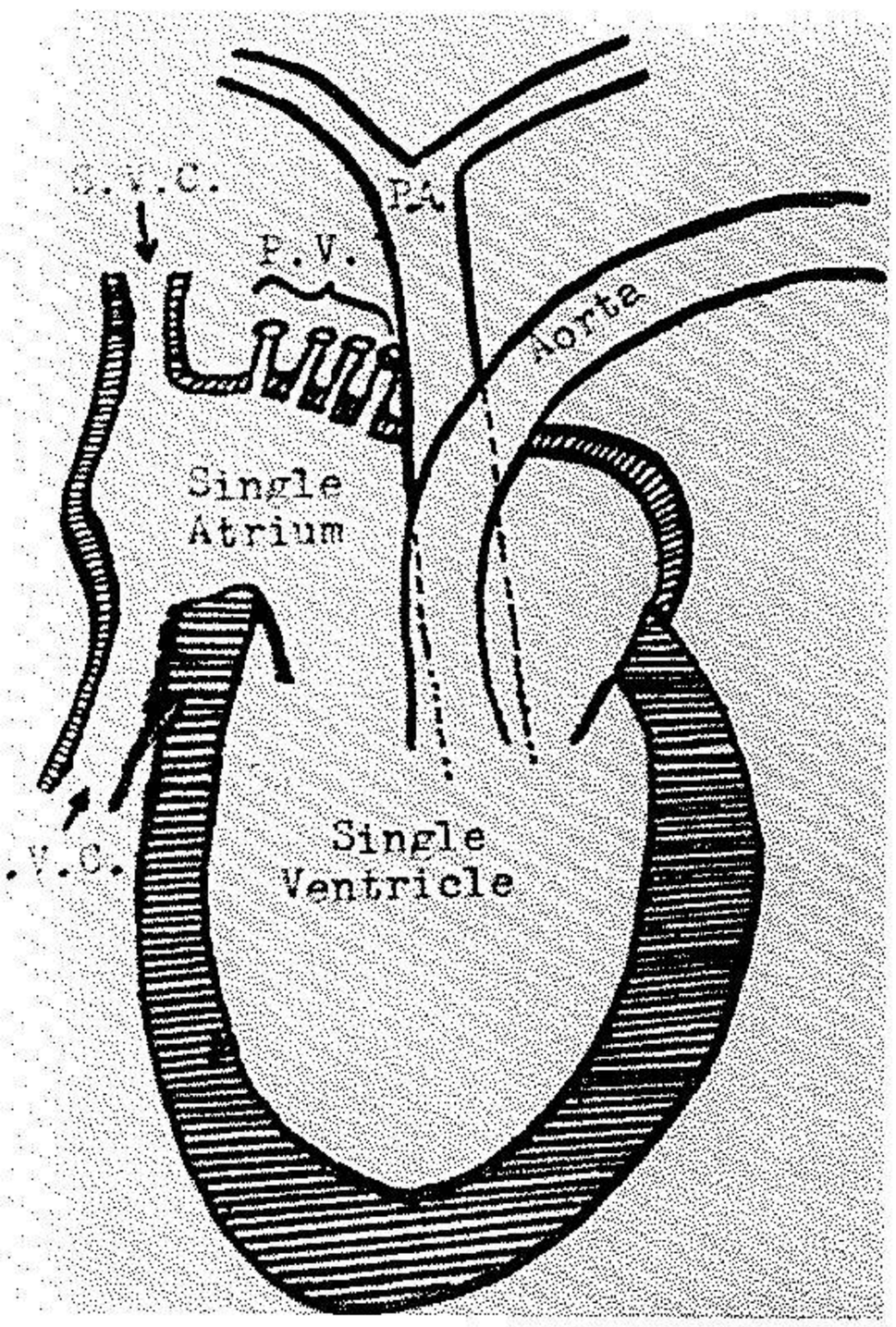

Fig. 2. Diagram of the patient's heart and pulmonary veins connecting to the right side of the atrium.

Fig. 3. The patient's pulmonary viens seen in the right side of the single atrium.

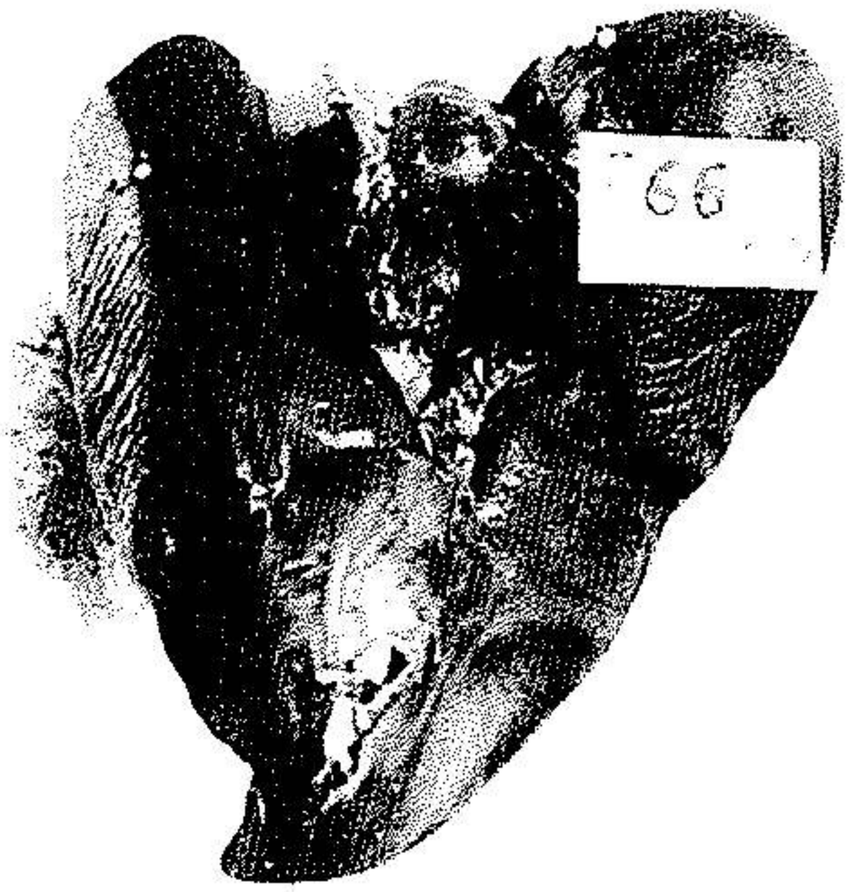

Fig. 4. The patient's large bilobate spleen with two small nodular spleen in its hillus.

RESPIRATORY SYSTEM: The larynx, trachea and bronchi were congested. The pleurae were normal. The cross sections of the lungs were policromatic and lacked even consistence. Bronchopneumonia was diagnosed microscopically.

DIGESTIVE SYSTEM: The tongue, salivary glands, pharynx, and esophagus were normal. The stomach mucous membrane was congested and atrophied. The intestine, vermiform appendix, and mesentric vessels were normal. The liver was enlarged and weighed 1,020 grams. Dilation and passive hyperemia of the veins and sinosoids were observed microscopically. The gall bladder and pancreas were normal.

LYMPH ORGANS: The lymph nodes and thymus were normal but a bilobated spleen weighing 50 grams and two small nodular spleens weighing 23.5 grams were attached to the hilus of the main spleen (fig. 4).

Other system, namely, urogenital, musculoskeletal and endocrines, were also normal.

SUMMARY

The clinical history and necroscopic findings of a $3\frac{1}{2}$ year old girl involved in cor biloculare was reported. In this case dextrotransposition of the great arteries and polysplenia was observed. The majority of cor biloculare cases appearing in the literature being accompanied by asplenia, the present case of polysplenia is relatively rare.

References

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