A Case of Total Anomalous Pulmonary Venous Connection to the Portal Vein

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ABSTRACT

An autopsy case of total anomalous pulmonary venous connection to the portal vein was presented and clinical and anatomical features were discussed.

Key Words: *congenital heart disease*; *pulmonary lymphangiectasis*; *patent ductus arteriosus*

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac malformation. This malformation is usually classified into 4 well-defined types, i.e. supracardiac, cardiac, infracardiac and mixed type according to the site of anomalous connection. Of these, the infracardiac type has an especially poor prognosis mainly because a pulmonary venous return is often obstructed at the level of diaphragma in this type of anomaly.

As we have recently experienced an autopsy case of infradiaphragmatic TAPVC, the clinical and anatomical features will be described below.

CLINICAL COURSE

A six-day-old male infant was refered to the Pediatric Ward of Yamaguchi University Hospital because of hyperbilirubinemia and cardiac murmur on November 26, 1977. The baby was the product of a full-term, uncomplicated pregnancy in a 28-year-old gravida 3, para. 2, with normal labor and delivery; he weighed 3380 g at birth.

At admission to the hospital his temperature was 36.5°C. The pulse and the respiratory rate were 174 and 72 per minute, respectively. A grade 3 systolic murmur was heard, maximal in the fourth costal space

at the left margin of the sternal border. The pulmonic second sound was slightly accentuated but clear-cut splitting could not be detected. The liver was palpable 3 finger breadths below the right costal margin. The spleen was not palpable. Laboratory data included a hemoglobin level of 13.5 g/dl; red blood cells of $449\times10^4/mm^3$ and white blood cell count of $7400/mm^3$ with 60 per cent lymphocyte present. The serum bilirubin was 19.6 mg/dl and phototherapy was instituted but without significant effect. The blood group was determined as 0 and Rh-positive (Rh D,c,$\bar{c}$,E,e: each type positive). His mother's blood was also type 0 and Rh-positive but Rh E negative. Therefore, the prolonged hyperbilirubinemia was ascribed clinically to hemolytic disorder due to Rh incompatibility but neither direct nor indirect Coomb's test was performed.

An electrocardiogram demonstrated sinus tachycardia at a rate of 172; the QRS axis was 120°; the precordial leads showed a pattern of right ventricular hypertrophy, with tall R wave in V_1 and upright T

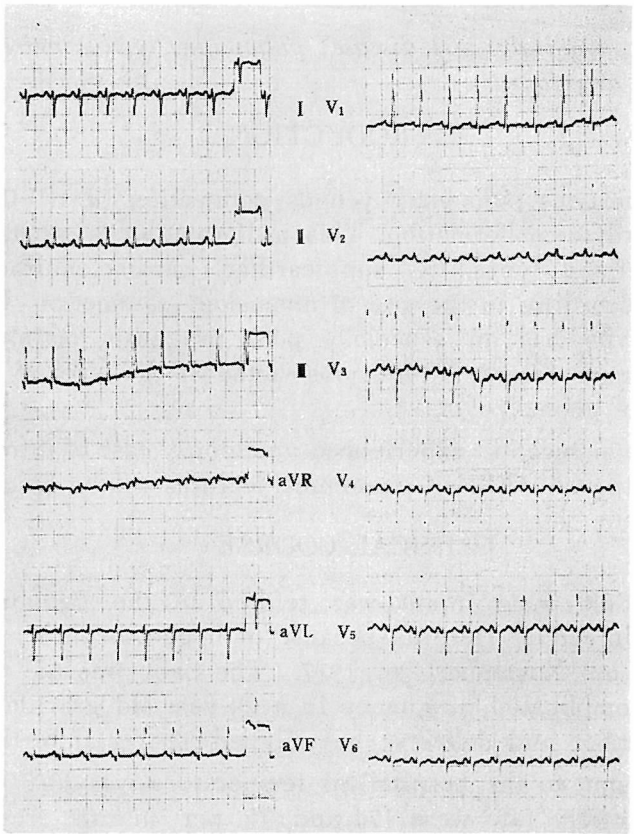

Fig. 1. Electrocardiograms of the patient at age of six days.

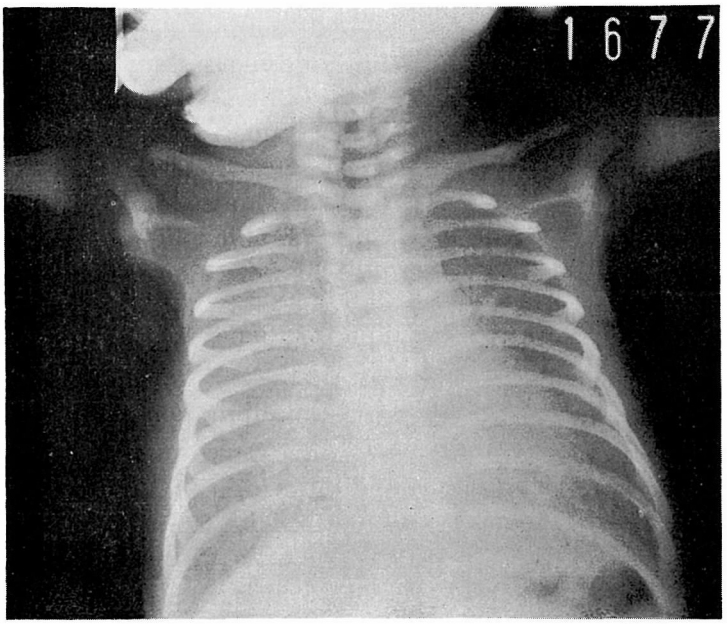

Fig. 2. Chest roentgenogram taken when the patient was eight days old. It shows a normal-sized heart and reticular markings in the lung fields.

waves in $V_{2\text{-}4}$ (Fig. 1).

A chest X-ray film on the 8th day of life showed a reticular appearance due to minute, scattered, irregular arreas of increased density in the lung field. But, no cardiac enlargement was noticed (Fig. 2).

Cyanosis was noted on his lip and nail beds during crying. Feeding was relatively poor throughout his life. He died on the twelfth day of life (the seventh day of hospitalization) despite of therapy with digitalis and oxygen.

PATHOLOGICAL FINDINGS

HEART: Examination of the heart showed marked dilatation and hypertrophy of the right atrium and the right ventricle, whereas the left side of the heart was notably small. The right ventricular wall was 0.4 cm in thickness and the left was 0.2 cm. The pulmonary artery was dilated and communicated with the aorta through a patent ductus arteriosus (Fig. 3). The left atrium was extremely small and received no venous return either from pulmonary vein or from systemic circulation. But instead, there was a septal defect, 0.5×0.4 cm at the height of foramen ovale. The two upper pulmonary veins had joined to form a common venous trunk, which then received the two lower pulmonary veins about 1 cm below and extended downward anterior of the esoph-

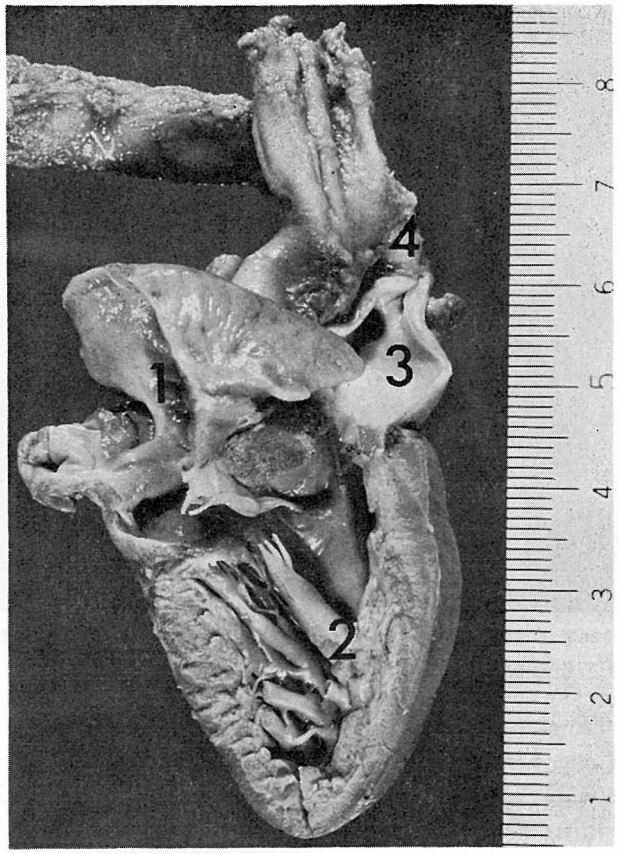

Fig. 3. Photograph of the heart which shows dilatation of the right atrium (1), hypertrophy of the right ventricle (2), dilated pulmonary artery (3) and patent ductus arteriosus (4).

agus to enter the abdomen through the esophageal hiatus and merged to a dilated portal vein at porta hepatis (Fig. 4). The common venous trunk was slightly constricted at the end.

LUNG: Grossly firm and atelectatic. Microscopic examination showed pronounced thickening of the alveolar walls with massive capillary engorgement. Beneath the pleura and in interlobular septum of both lungs, there were numerous dilated thin-walled channels lined with flat endothelial cells (Fig. 5). These channels were mostly empty, but some were filled with blood. The bronchi and pulmonary arteries were not remarkable.

LIVER: The liver weighed 162 g and was yellowish brown in color. The portal vein was dilated at the entrance of the common pulmonary vein. Microscopically, small fat droplets were deposited in the hepatic

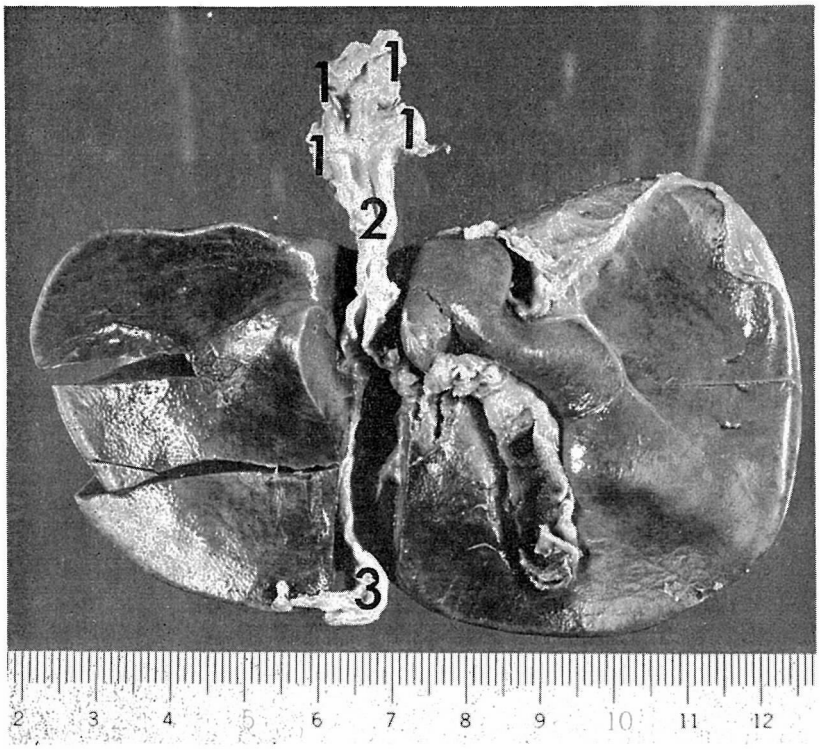

Fig. 4. Inferior aspect of the liver and anomalous pulmonary vein. It shows the pulmonary veins (1), the common anomalous pulmonary vein (2) and the umbilical vein (3).

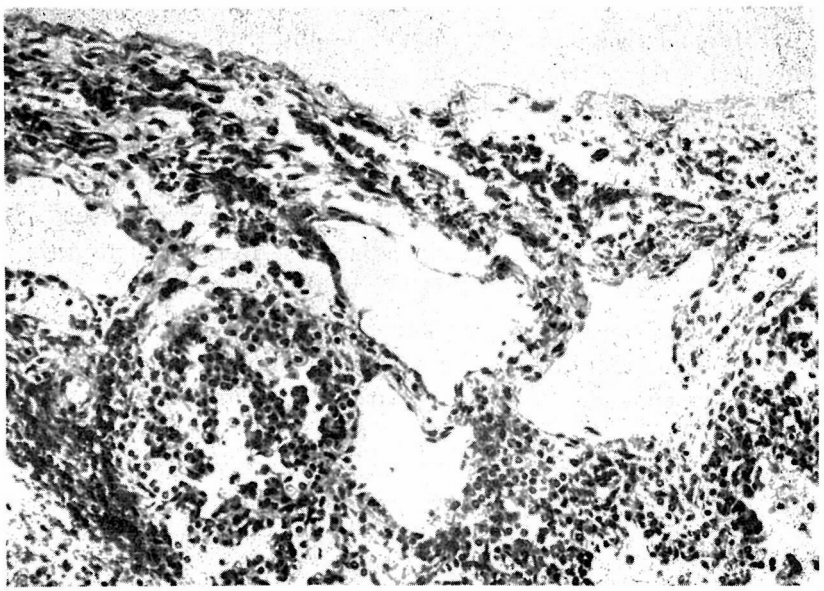

Fig. 5. Photomicrograph of the lung showing dilated lymph channels in interlobular septum. (H. and E. stain, ×100)

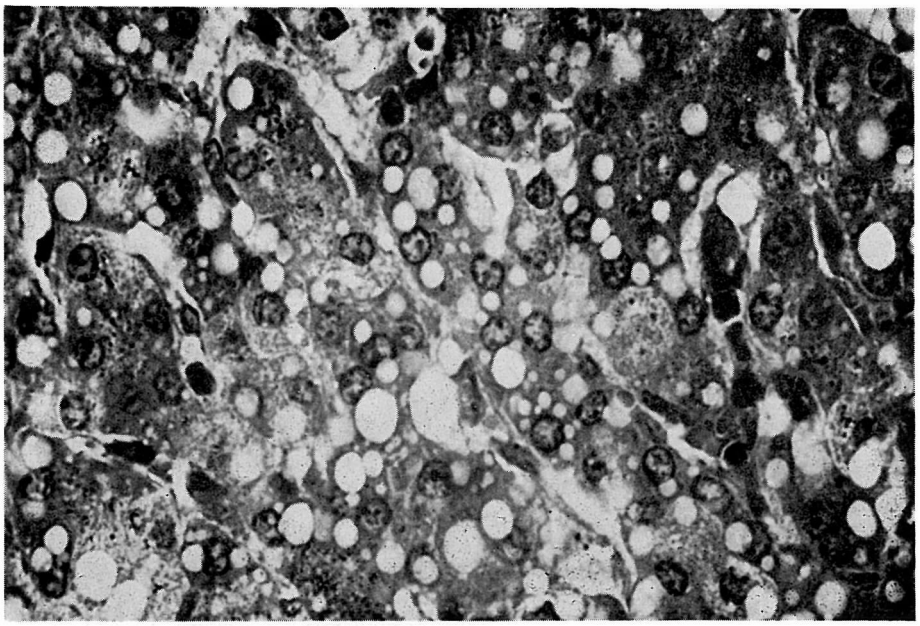

Fig. 6. Diffuse fatty change of the liver. (H. and E. stain, ×200)

cells throughout the entire lobule (Fig. 6). Numerous small foci of extramedullary hematopoiesis were seen in the sinusoid. Bile stasis, although present, was not marked.

COMMENT

Total anomalous pulmonary venous connection to the right side of the heart was first described as an entity by Friedlowsky in 1868 (cited by Darling[1]). Recently, detailed clinical and anatomical features of this anomaly were reported based on the collection of large series[2-5].

The frequency of TAPVC was reported to be approximately 1% of congenital malformations and 4% of congenital heart diseases[6]. In total of 75 autopsies of congenital heart diseases from 1959 to 1977 in our laboratory, only three cases of TAPVC were observed.

TAPVC have been classified according to (1) the anatomic level of the anomalous connection, (2) the length of anomalous route and (3) the presence or absence of other major cardiac defects. For example, Darling et al.[1] based their classification upon the site of anomalous connection. A classification by the length of anomalous route was proposed by Burroughs[7] to correlate each case to its prognosis, but the long route, intermediate route and short route groups correspond to infracardiac, supra-

cardiac and cardiac type of Darling's classification, respectively. The anomalous connection below the diaphragma is in 10 to 20 percent of TAPVC[1,7]. Statistics of TAPVC without other major cardiac defect varies greatly (37% to 77%)[1,2,5,7] depending on whether the case with patent ductas arteriosus is included or not.

The symptoms of TAPVC are not appreciably different from those with other congenital heart disease. According to Gathman et al.[3], cyanosis was noted in 61 percent of infants within 1 month of age, and the symptoms of congestive heart failure such as tachypnea, increased sweating and poor feeding were present in 41 percent. Burroughs[7] emphasized a tendency that the longer the anomalous route and the smaller the inter-atrial comunication is, the earlier is the onset of cyanosis. For precise diagnosis, cardioangiography is essential.

The infradiaphragmatic TAPVC is of extremely poor prognostic type and fatal within 3 months of life because of high resistance to pulmonary venous return[1-3,8-10]. Various causes of resistance have been considered ;i.e. (1) the narrow caliber of the common venous trunk, (2) long route of the anomalous vein, (3) thoraco-abdominal pressure difference and (4) resistance of hepatic capillary bed. A major contributing factor in our case would be the hepatic capillary bed.

According to Harris et al. (cited by Lucas[9]), clinical indication of the pulmonary venous obstruction is afforded by the chest rentgenogram. The findings are characterized by a normal-sized heart in combination with hyperinflated, edematous lungs and these factures are accounted tor not only by the intra-alveolar accumulation of fluid but also by the interstitial edema and lymphatic dilatation[2]. The pulmonary lymphangiectasis was also reported by several authors[9,11,12].

As to the developmental mechanism of TAPVC, agenesis, involution and atresia of the common pulmonary vein were considered to be possible[5].

Seriously ill infants with TAPVC and with pulmonary venous hypertension require emergency operation as soon as the diagnosis is made. In infradiaphragmatic type of TAPVC, however, the mortality rate of corrective surgery is unfortunately high[13,14].

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