

Tracheoesophageal Fistula and Anomalies of Thoracic Vessels

Their Occurrence in a Case of Congenital Rubella

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• Virologic, gross, and microscopic studies of rubella embryopathy were performed. The presence of esophageal atresia with tracheal fistula allied with anomalies of the thoracic vessels were seen. We believe that both the mechanical action of the anomalous vessels on the developing organ, and the vascular degenerative aortic lesions, which are identical to those described in rubella syndrome, are probable causes of the esophageal malformation.

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The use of virologic and serologic methods to assess rubella permits the evaluation of the pathologic potential of an intrauterine infection with a large range of possible injury. Although there is a large diversity of associated malformations, references to esophageal atresia^{1,2} and to tra-

cheoesophageal fistula³ are scanty.

We studied a case of esophageal atresia with tracheoesophageal fistula allied with malformations and degenerative lesions of some thoracic vessels that are identical to those described in the rubella syndrome.^{4,5} The diagnosis was based on the isolation of the virus from autopsy material.

REPORT OF A CASE

At 44 hours of age, a boy, who weighed 2,370 g, was admitted to the Surgical Pediatric Department of Instituto Fernandes Figueira, Rio de Janeiro. He was found to have mild icterus, normal but ectopic anus, and severe respiratory distress. A thick secretion was continuously excreted by the nostrils and mouth. Roentgenograms showed esophageal atresia without tracheoesophageal fistula, normal cardiac shadow, and pneumonia in the right lung. A gastrostomy was set up and the baby continued to receive parenteral feeding. Based on the results of the roentgenographic examination, he was fed human milk; there was immediate increase of oral and nasal secretion and he suddenly died.

At the time of autopsy, besides mild icterus and ectopic but perforated anus, he displayed low-set ears and enlarged fontanelles.

Internal examination revealed hepatosplenomegaly and several malformations: esophageal atresia and distal tracheoesophageal fistula (Fig 1), anomalies of aortic arches with anomalous implantation of the right subclavian artery (which emerged from the aorta after the left subclavian artery, passing just below the atresic upper portion of the esophagus before reaching the right superior limb [Fig 2 and 3]), patent ductus arteriosus and preductal aortic stenosis, duplication of the right renal artery, and abnormal rotation of the left kidney.

Viscera were fixed in 10% formaldehyde solution. Hematoxylin-eosin stain was used routinely, as well as specific stains, such as PAS, Sudan III, orcein for elastic tissue, and Perls' and von Kossa stains for identification of iron and calcium deposits, respectively.

The microscopic examination of the viscera permitted identification of inflammatory and degenerative lesions described in congenital rubella, such as chronic meningoencephalitis with perivascular mineralization, chronic active hepatitis, precocious stimulus of the lymphoid tissues, iridocyclitis, fibrosis of the choroid, and ectopy of splenic and parathyroid tissues. The presence of degenerative lesions in the thoracic and abdominal aorta, evidenced by degeneration or fragmentation of elastic bands that were surrounded by amorphous mate-

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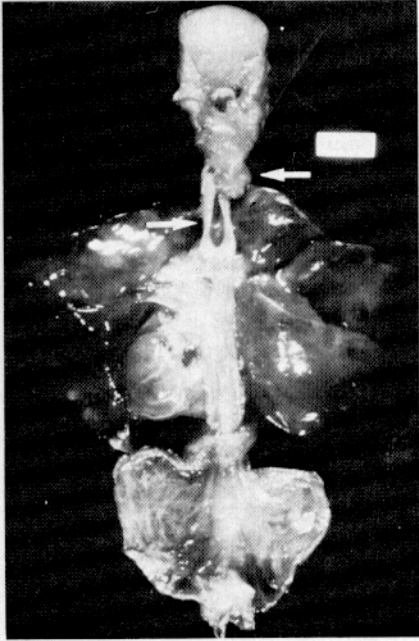


Fig 1.—Upper portion of esophagus is seen, as well as wide tracheoesophageal fistula (arrows).



Fig 2.—Atresic upper portion of esophagus (1) is limited by two subclavian arteries (2, 3).

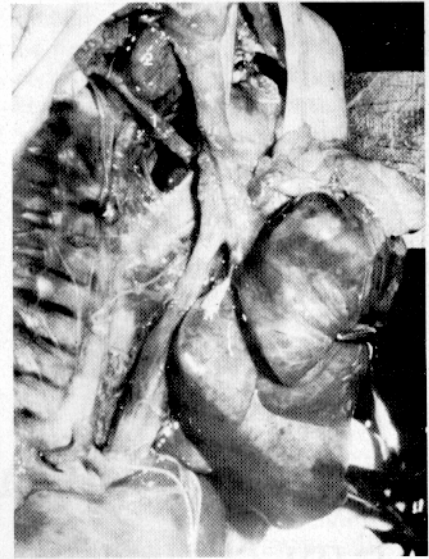


Fig 3.—Lateral view showing right subclavian artery (1) just beneath atresic portion of esophagus (2).

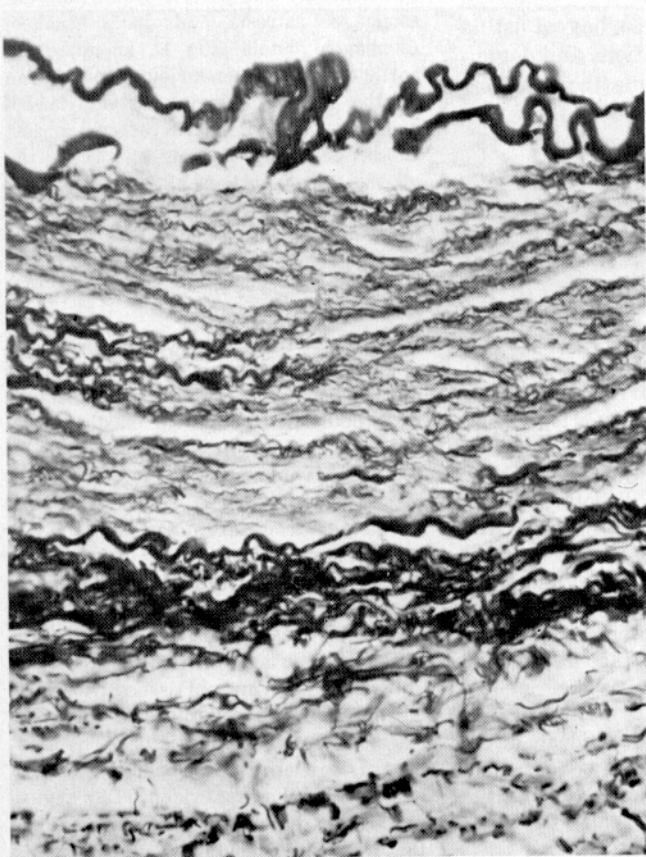


Fig 4.—Irregularity and fragmentation of elastic bands of thoracic aorta (orcein, $\times 560$).

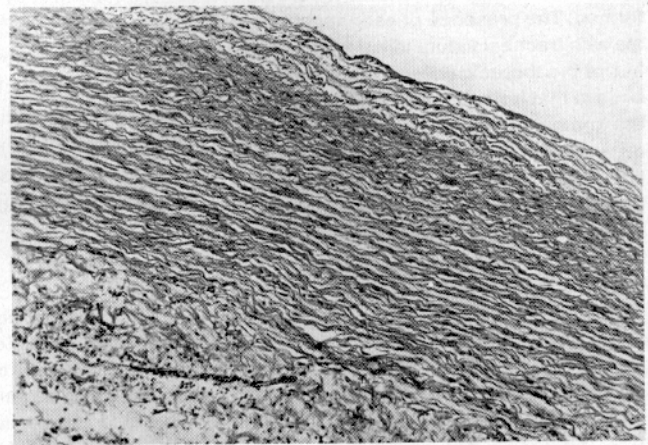


Fig 5.—Fibromuscular proliferation of intima of thoracic aorta (Gomori stain, $\times 125$).

rial were observed (Fig 4 and 5). In the pulmonary and renal arteries, proliferative fibromuscular lesions were found. Death was probably due to pneumonia by aspiration.

Autopsy specimens from this case (liver, lung, and heart) were obtained and sent for virologic investigations. Tissue culture techniques revealed a cytopathic effect in the human epithelial (HEP)-II cell line from the liver and lung specimens. Identification was made by specific neutralization test and confirmed to be rubella virus.

COMMENT

Obstructive lesions in the gastrointestinal tract have been associated with congenital rubella^{6,7}; association with rubella was suggested by the possibility of an intrauterine lesion of the mesenteric artery, which resulted

in an atresia analogous to the intestinal lesions that follow mesenteric artery ligation during pregnancy.⁸

The angiotropism of the rubella virus is considered one of the most potent pathogenic mechanisms of the intrauterine infection.⁹ The presence of the virus in the vasculature of the embryo and the chorionic tissues has been demonstrated,^{10,11} and is considered to be the cause of the pleiotropism of prenatal rubella and of the late vascular lesions described in certain congenital virus infections.¹²

With respect to the cause of esophageal atresia, various opinions have been formulated. Roy et al¹³ considered that the presence of this type of malformation in only one or both monozygotic twins points to nongenetic origin, whereas Keith,¹⁴ Lang-

man,¹⁵ and Fluss and Poppen¹⁶ attributed the amputation of that organ to the mechanical effect of anomalous vessels on the developing esophagus. In this case, besides the pressure exerted by the vascular ring, the degenerative lesions of the thoracic aorta may have influenced the anomalous development of the esophagus. The isolation of rubella virus from the lungs and the liver, as well as the presence of multiple anomalies and the microscopic visceral and vascular lesions, may be considered as manifestations of the wide dissemination of the virus in the embryo.

The misinterpretation of the thoracic roentgenograms allowed feeding by gastrostomy, which caused dissemination of the pneumonia by aspiration, leading to death.

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