Anomalous origin of single coronary artery with multiple heart malformations

S. Zachariah and R. Reif

From the Department of Pathology, Asaf Harofe Government Hospital, Tel-Aviv University Medical School, Zerifin, Israel

A newborn female infant is reported who had a single coronary artery arising from the innominate artery. In addition, a truncus arteriosus, mitral atresia, and single ventricle were found. Only one case with exactly the same malformation has apparently been published so far.

Single coronary arteries having their origins from the aortic or pulmonary sinuses have frequently been described. Origins of the single coronary artery from sites other than these are extremely rare, and to the best of our knowledge only three cases have been described to date (Mayer, 1827; Forster, 1847; Keeling, 1970). The purpose of this report is to put on record another case of rare anomalous origin of a single coronary artery arising from the innominate artery, in association with multiple heart malformations.

Case report

This 3-day-old female infant was born in Israel to parents of Moroccan extraction on 22 April 1972, with a birthweight of 1.67 kg. The mother's pregnancy was complicated by threatened abortion and the infant was born prematurely. The delivery was normal. The mother had one living son and suffered a previous abortion (details of which are not available). The infant was apparently normal at birth and was rated 10 on the Apgar score. Because of her premature birth and low birthweight, she was transferred to a special premature care centre. On admission there the infant was found to be in respiratory distress with tachypnoea and cyanosis of lips and nail beds. She was placed in an incubator and resuscitative therapy with oxygen, intravenous fluids, antibiotics, and other supportive measures instituted. She failed to respond to any of these measures and died on the morning of 25 April 1972.

Necropsy

The infant weighed 1.52 kg and measured 44.5 cm in length. There was cyanosis of the face, lips, and nailbeds. No external malformations were present. The pericardial reflexions were normal. There was cardiac enlargement involving the right atrium and the ventricle.

The venae cavae drained normally into a dilated and hypertrophied right atrium. The foramen ovale was widely patent. The right atrium drained into a single, hypertrophied ventricle via a normal tricuspid valve. Pulmonary venous drainage was normal into a slightly dilated left atrium. There was complete atresia of the mitral valve, in the region of which only a small dimple existed. No second ventricular chamber could be demonstrated. The only existing outlet for the left atrium was therefore to the right atrium via the patent foramen ovale. Drainage of the ventricle was accomplished by a single truncus arteriosus guarded by a tricuspid valve and from which two pulmonary arteries arose separately at the same level from its posterior aspect, corresponding to type III of Collett and Edwards (1949). The truncus ascended, arched backwards and to the left, and then downwards, giving off the innominate, left common carotid, and left subclavian arteries from its arch. No coronary arterial orifices could be demonstrated within the lumen of the truncus. Further dissection of the specimen revealed a single coronary artery arising from the innominate artery and, enclosed in loose areolar tissue, descending anterior to the right pulmonary artery and just posterior to the truncus to the right atrioventricular groove where it immediately gave rise to an inflorescence of small arteries to supply the entire myocardium (Fig.). The distribution of the branches was atypical, not corresponding to that of the normal right or left coronary arteries.

Several branches descended on the anterior surface of the heart, while one branch each continued in the atrioventricular groove in each direction giving off several smaller branches on the lateral and posterior surfaces of the heart as well as to the atria. The coronary artery was accidentally cut across when the right pulmonary artery was opened.

Other macro- and microscopical findings included acute congestion of most internal organs with petechial haemorrhages, moderate fatty change of liver cells as well as aspiration.

FIG. Specimen showing single coronary artery arising from innominate artery. IA=innominate artery; LPA=left pulmonary artery; RPA=right pulmonary artery; and TA=truncus arteriosus.

Discussion

With the rapid advance in diagnostic procedures that are instituted preoperatively, the surgeon today need not enter into totally unknown territory. A good knowledge of possible variations in anatomy is, however, indispensable in planning surgical procedures. Our case here demonstrates another variation in the spectrum of coronary artery

anomalies, this type having been described only once before by Keeling (1970), and the type of malformation found in our case resembles exactly that found in his patient. In the case described by Mayer (1827) the coronary originated from the right carotid artery. In the case of Forster (1847) the single coronary emerged from the inferior aspect of the aortic arch.

Attempts have been made to classify the anomaly of the single coronary artery. Smith (1950) proposed three groups based on whether the artery was distributed according to one or both of the coronary arteries or was atypical. Our case falls into the third of these groups. Ogden and Goodyear (1970) devised an elaborate classification from a study of 95 cases according to basic distribution patterns. They based their classification on the number of major divisions of the single coronary artery and the subsequent course of these major branches. Our case, however, falls outside their classification. It might be interesting to note that in both cases with anomalous origin of the coronary artery the pregnancy was complicated by threatened abortion. The birth weight in Keeling's case was 2.15 kg; in our case it was 1.67 kg.

References

- Collett, R. W., and Edwards, J. E. (1949). Persistent truncus arteriosus: a classification according to anatomic types. *Surgical Clinics of North America*, 29, 1245.
- Forster, T. (1847). A heart consisting of only two cavities. *Transactions of the Pathological Society of London*, 1, 48.
- Keeling, J. W. (1970). Anomalous origin of a single coronary artery. *Journal of Pathology*, 102, 174.
- Mayer, S. F. (1827). Eine Missgeburt mit einem Parasiten auf der Brust. *Journal der Chirurgischen Augenheilkunde*, 10, 44.
- Ogden, J. A., and Goodyear, A. V. N. (1970). Patterns of distribution of the single coronary artery. *Yale Journal of Biology and Medicine*, 43/1, 11.
- Smith, J. C. (1950). Review of single coronary artery with report of 2 cases. *Circulation*, 1, 1168.

Requests for reprints to Dr. R. Reif, Department of Pathology, Asaf Harofe Government Hospital, Zerifin, Israel.