

Brief Report

Thrombotic occlusion of the main stem of the left coronary artery in a neonate

Luby Abdurrahman, Steven M. Schwartz, Robert H. Beekman III

Division of Cardiology, Children's Hospital Medical Center, 3333 Burnet Avenue, Cincinnati, Ohio, USA

Abstract Thrombotic coronary arterial occlusion, and myocardial infarction, are rare in the newborn. We report such a happening presenting shortly after birth with cardiogenic shock, no left ventricular output and a systemic circulation dependent on flow from a patent arterial duct.

Key words: Coronary; thrombus; infarction; neonatal

WE REPORT A RARE OCCURRENCE OF thrombotic occlusion of the main stem of the left coronary artery presenting at birth in an otherwise normal neonate.

Case report

The patient was the 2.88 kg product of an uncomplicated 42-week gestation. The placenta weighed 315g and was histologically normal. Hypotension and bradycardia were present at birth with Apgar scores of 2, 4, and 6 at 1, 5 and 10 minutes respectively. Physical exam revealed an irregular tachycardia of 150–190 beats per minute, a single second heart sound and no murmurs. The chest radiograph was normal. The electrocardiogram demonstrated 4–6mm ST depression in leads V1–V4, and 3–4mm ST elevation and qS pattern in leads V5 & V6, suggestive of infero-lateral myocardial injury.

Echocardiogram and Doppler evaluation showed a large, dilated, poorly contractile left ventricle. The leaflets of the aortic valve were thin but immobile with no antegrade flow. There was however, a continuous jet of regurgitant flow across the aortic valve, indicating that the valve was not atretic. The mitral valve was severely regurgitant,

and Doppler interrogation of the regurgitant jet suggested a left ventricular systolic pressure of 25mmHg (estimated left atrial A' wave of 10mmHg). The simultaneous right arm systolic blood pressure was 40mmHg. The aortic arch appeared normal. A large patent arterial duct permitted right-to-left flow. The right coronary artery appeared normal, but only the most proximal portion of the left coronary artery was visualized. Serum creatine phosphokinase, and its MB fraction, were markedly elevated (creatinine phosphokinase 2808 I.U/L, normal 61–519 I.U/L; MB fraction 356 I.U/L, normal 24 I.U/L).

Cardiac catheterization revealed a left ventricular systolic pressure 15mmHg lower than the simultaneous systolic pressures in the ascending and descending aorta and the right ventricle (45mmHg). The left ventricular end diastolic pressure was 20mmHg. A left ventricular angiogram showed a poorly contractile, dilated left ventricle, severe mitral regurgitation and no antegrade flow across the aortic valve. An injection in the aortic root revealed multiple filling defects in the aortic arch and brachiocephalic arteries. The right coronary artery and its branches appeared normal. The main stem of the left coronary artery had a proximal filling defect, beyond which the coronary arterial system did not fill (Fig. 1). No collateral coronary circulation was present.

A trial of thrombolytic therapy with Tissue Plasminogen Activator was initiated. An injection in the aortic root 12 hours later showed resolution of the aortic filling defects, but the filling defect in

Correspondence to: Luby Abdurrahman, MD, Division of Cardiology, Children's Hospital Medical Center, 3333 Burnet Avenue, Cincinnati, Ohio 45229–3039, USA. Tel: (513) 559–8267; Fax: (513) 475–3952

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the coronary artery was unchanged. Attempts at direct instillation of Tissue Plasminogen Activator into the left coronary artery failed. The baby died on the second day of life.

Autopsy revealed an organized thrombus occluding the main stem of the coronary artery 5 mm from its origin (Fig. 2a). Myocardial necrosis was limited to the distribution of the left coronary artery (Fig. 2b). No other thrombus was found. Myocardial inflammation was limited to the wall of the left coronary artery adjacent to the thrombus. Thrombotic screening could not be performed because the patient had received blood products. Subsequent screening of the parents for prothrombotic disorders revealed both parents to have low levels (25%) of Protein C. The patient's normal platelet count (158,000) and prolonged partial thromboplastin time (>90 sec) however, are suggestive of antiphospholipid syndrome,¹ though administration of heparin prior to these measurements makes this diagnosis uncertain.

Discussion

This case illustrates the rare occurrence of thrombosis of the main stem of the left coronary artery, and subsequent myocardial infarction in a neonate. It is only the third case reported in the literature. Shock, immobile leaflets of the aortic valve without antegrade flow, and a systemic circulation dependent on the patent arterial duct all highlight the similarity of this case to critical left-sided



Figure 1. Aortic root angiogram showing the stump of the LMCA with proximal obstruction (arrow); the RCA appears normal. There is no significant collateral circulation.

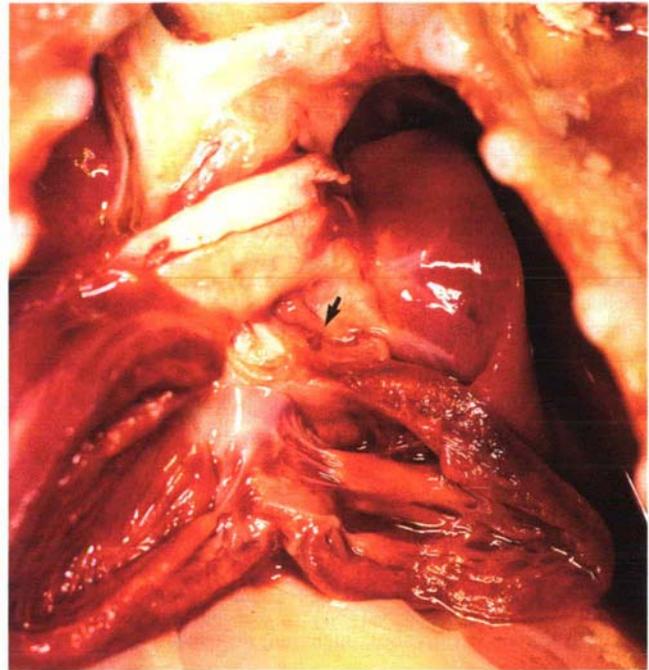


Figure 2a. Dissection through the cavity of the left ventricle and aorta shows the orifice of main stem of the left coronary artery with the thrombus (arrow) visible within its lumen.

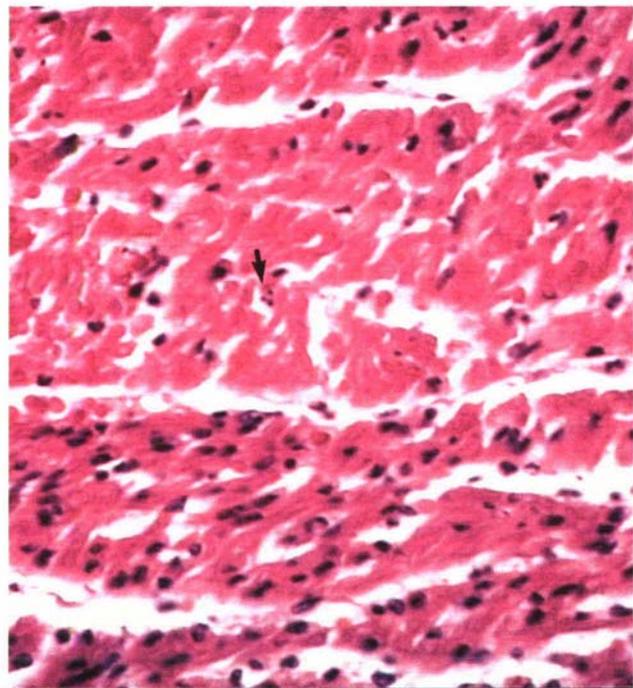


Figure 2b. Section of the left ventricular myocardium showing an area of myocardial ischemia with hypereosinophilia and loss of myocyte nuclei and visible nuclear fragments (arrow) consistent with coagulation necrosis in the lower panel, compared to the adjacent zone of normal myocardium in the upper panel (Stain hematoxylin and eosin 40X).

obstruction of the systemic circulation. In this case, the thrombosis was the primary event resulting in infarction and severe depression of left ventricular function, with consequent inability of the left ventricle to generate a pressure higher than that supplied to the aorta by the patent duct. Thrombus in the aorta may have occurred secondary to low cardiac output,^{2,3} or may have represented a primary thrombotic disorder. Although both parents had low levels of Protein C, and therefore might have been heterozygous for Protein C deficiency, invoking homozygous deficiency in this particular case would be mere speculation. As in previous reports,^{4,5} the etiology of the thrombus remains unknown. This diagnosis should be considered in severe neonatal left ventricular dysfunction without other identifiable cause.

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