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The Ephemeral Femoral Artery: Detection of Aortic Arch Thrombus Presenting as Coarctation

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Abstract

An apparently healthy newborn infant subsequently presented with coarctation and was found to have a partially occluding extensive thrombus within the aortic arch. Non-palpable femoral pulses on the third day of life led to the diagnosis. No underlying explanation was uncovered and thrombophilia was excluded. Early detection resulted in successful thrombus removal and vessel repair with complete recovery. Given the variability of subjective finger palpation consideration should now be given to portable Doppler ultrasound as an inexpensive and, clinically practical adjunct or alternative to the routine neonatal congenital heart screen.

Keywords

Femoral artery; Aortic arch; Coarctation; Thrombus; Thrombophilia; Doppler

Introduction

Coarctation of the aorta is a rare but potentially devastating condition. Detection traditionally has depended on absent femoral pulses detected by finger palpation of the artery in the groin of infants. Variable reliability of this especially subjective part of the newborn examination may lead to missing the diagnosis given a normal pulse oximetry cardiac screen. Yet successful outcome depends on early diagnosis. We propose that a portable Doppler ultrasound backup be considered to supplement the cardiac screen and femoral artery palpation.

Case Study

EV was the product of an uneventful pregnancy, born at 39 weeks to a healthy Gravida 1, Para 1, 28 year-old Caucasian social worker. A transient fetal arrhythmia noted during the third trimester on fetal echocardiogram was thought to represent a few innocuous premature atrial contractions. The mother had previously tested negative for toxoplasmosis, but CMV (Cytomegalovirus) status was not determined and she was asymptomatic throughout her pregnancy. The father of the baby is a healthy African man whose sickle cell and hemoglobin chain status were unknown.

The baby was delivered by an uncomplicated normal spontaneous vaginal delivery. He cried spontaneously and vigorously without cyanosis, had APGAR (Appearance, Pulse, Grimace, Activity, and Respiration) scores of 8 and 9, at 1 and 5 minutes, and received intramuscular vitamin K. No placental abnormalities were noted. He passed his neonatal congenital heart screen. The postnatal course was uneventful and serial physical examinations did not detect any abnormalities. He was discharged to his pediatrician on the second day of life.

On day 3, he was breastfeeding well but the examination revealed a blowing grade 2/6 systolic ejection murmur at the left sternal border without radiation. On careful and persistent physical examination, the femoral arteries were not palpbale. There was no tachypnea, tachycardia, abnormal heart rhythm, pallor or cyanosis. He was referred to the on-call pediatric cardiologist and seen within the hour at which time he was noted to be clinically vigorous and well perfused. An emergency echocardiogram was performed once the original findings were confirmed. The echocardiogram revealed an area of flow acceleration seen in the distal aortic arch. This was initially thought to be a discrete coarctation of the aorta but on further imaging an echo bright structure proximal to the area of flow acceleration was noted in the transverse arch and head and neck vessel junctions. There was severe obstruction at the level of the proximal descending aorta with a peak equaling 70-74 mmHg, and mean of 25-29 mmHg. The proximal portion of the descending aorta appeared to be tortuous. Flow in the pulmonary artery appeared normal without obstruction and without evidence of a patent ductus arteriosus. The initial impression of coarctation of the aorta subsequently appeared to be obstruction secondary to an intraluminal mass, presumably a thrombus, extending from the proximal portion of the transverse aortic arch through the proximal descending aorta.

EV was admitted to the neonatal intensive care unit where he remained stable. Prostaglandin E infusion was initiated. A chest X-ray showed a normal heart size and well

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inflated lungs. Through a properly placed umbilical vein catheter, an aortic CT angiogram was performed. The CT showed tapered narrowing of the isthmus without discrete coarctation. The narrowed portion measured 3.9 × 4.2 mm, while the transverse arch measured 6.3 × 6.2 mm, and the descending aorta just above the diaphragm, 7.6 × 7. 3 mm. The CT reading disclosed a calcified non-occlusive thrombus within the distal descending aorta and transverse arch extending to the proximal portion of the right brachiocephalic artery and the left common carotid artery.

Investigations commenced prior to surgery included carotid artery Doppler, an MRI of the brain, renal ultrasound with Doppler and Doppler of the femoral vessels. In addition, hematology was asked to evaluate potential thrombophilia. The MRI/MRA/MRV of the brain showed no ischemia, but there were 4 and 2 mm foci of signal abnormality possibly representing antenatal thromboembolic events. The renal ultrasound was normal.

At the time of surgery on the 4th day of life the baby was indeed found to have aortic arch obstruction, distal ascending aortic obstruction, obstruction of the left carotid artery and the left subclavian artery, partial obstruction of the innominate artery, and a patent foramen ovale. Appropriate endarterectomies and anastomoses, aortic arch reconstruction and autologous patch augmentation of the arch were carried out successfully under cardiopulmonary bypass. EV spent 9 days in the pediatric intensive care unit recovering from surgery and was discharged to his parents. He had no apparent deficit or abnormality on physical exam. EV was prescribed Aspirin and furosemide initially and continues on the former. At age eight months, he is developing and gaining weight normally and meeting or surpassing expected milestones.

EV was found to be heterozygous for the *97G>A prothrombin mutation, and his mother was homozygous. The baby's risk for venous thrombosis was considered increased only by 3-5 fold. That risk was judged insufficient to warrant anticoagulation or a post-natal diagnosis of thrombophilia. The remainder of the laboratory studies were essentially normal: In particular, prothrombin time, partial thromboplastin time, Protein C and S, D dimer, fibrinogen, plasminogen, antithrombin III, anti cardiolipin antibodies, lupus anticoagulant, factor VIII activity, Factor V Leiden PCR prothrombin (F2) 2020A PCR, all obtained preoperatively, were within normal ranges.

Discussion

Only a smattering of neonatal aortic arch thrombosis cases presenting as coarctation has been reported [1,2]. The majority of these infants have died or suffered cerebral infarction secondary to carotid artery extension of the thrombus and embolization. A few have been shown to have predisposing thrombophilia and one was reported to have had intra-uterine CMV infection [3]. The vast majority of neonatal thrombotic events are associated with umbilical vessel catheterization. In our case, an extensive review of the pregnancy and neonatal hospital course, hematologic studies to detect thrombophilia, and infectious disease and rheumatology consultation did not reveal a source for the thrombus formation. Being heterozygous for the *97G>A mutation did not indicate thrombophilia [4]. Thrombus formation was likely to have occurred sometime before birth, given organization of the thrombus. Propensity of a clot to lodge at, or around the isthmus may be influenced by differential blood flow to the carotid artery or to other complex hemodynamic physiology singular to this location [5]. The definitive approach to this particular condition in an otherwise stable infant appears to be surgery undertaken acutely to remove the thrombus and reconstruct intact, functional vasculature. Anticoagulation therapy is available, but less likely to be effective when the thrombus is old and organized.

Clinical postnatal detection of coarctation continues to rely on one or two finger palpation of the femoral arteries in the groin of the neonate. This is an accepted, but highly subjective measurement, one often carried out in less than ideal circumstances in a busy, loud newborn nursery. When challenging, it may well be deferred. Both the sensitivity and thickness of the examiner's finger varies widely from one individual to another [6,7]. Lacking objective feedback, it is possible to imagine the presence of a femoral pulse, even confusing that with one's own finger pulse. The neonatal congenital heart screen may or may not identify a coarctation.

Thus, the pediatrician is the first line agent for exclusion of a rare though possibly under reported, but rapidly devastating condition, in which early detection provides the best chance for survival. Closure of a patent ductus arteriosus, an associated cardiac anomaly, or simply the passage of time can lead to untoward severe consequences. Just as the pulse oximeter has largely replaced the visual diagnosis of subtle cyanosis, it may now be time to consider Doppler identification of a normally pulsating femoral artery as part of the newborn examination.

Handheld Doppler units are now commercially available and inexpensive alternatives to a challenging and perhaps outmoded clinical practice. There currently is no published evidence validating the sensitivity and specificity of a Doppler pulse evaluation compared against simple finger palpation. However, the author (FRB), has found it to be a simple and more reliable measure of the femoral artery pulse in a limited series, and application in other contexts such as radial artery cannulization is established practice. We conclude that a portable Doppler unit might be of use shortly after birth to rule out pre-bifurcation coarctation of the aorta and provide relatively better information on which to base an urgent referral decision than finger palpation.

Conclusion

Coarctation of the aorta in the neonate is a serious, albeit rare, condition requiring early detection for successful correction without dire sequelae. Diagnosis by femoral artery palpation is subjective and variably sensitive and specific. Addition of portable Doppler pulse detection could be a valuable adjunct to the pulse oximetry cardiac screen.

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