Carotid Arterial Elastic Hyperplasia in a Newborn

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Abstract: Cerebrovascular insufficiency in infancy and childhood is well documented and has a diverse and often unknown etiology. Reported here is a polycythemic, microcephalic infant girl of 43 weeks' gestation with bilateral cerebral infarction occurring in the perinatal period. Infarction was the result of bilateral carotid artery stenosis produced by massive reduplication of the internal elastic lamina. Review of the literature failed to reveal any reports of similar arterial lesions. The angiographical picture was also unique.

The etiology of this unusual defect is not known, but we believe the problem was congenital perhaps due to an intrauterine infectious process.

Additional Key Words: cerebral infarction, congenital vascular disease

Numerous reports of cerebrovascular insufficiency in infancy and childhood have been recorded, and this problem has been the subject of comprehensive evaluation by the Joint Committee for Stroke Facilities in its recent report.1 The etiology of congenital cerebrovascular insufficiency is obscure in many cases, since mortality rates are relatively low and tissue specimens, therefore, are usually unavailable.

The following is a report of bilateral cerebral infarction in a polycythemic newborn infant with stenosis of the carotid arteries resulting from massive reduplication of the internal elastic lamina.

Case Report

The patient was a 2,830 gm, 43-week gestation, infant girl (small for gestational age), who was the product of an uncomplicated pregnancy and delivery. The 33-year-old mother had two living children and four prior miscarriages. There was no maternal history of infection or ingestion of teratogenic agents during the pregnancy.

The infant was in no acute distress at birth, although meconium staining of the skin and fingernails was noted. Within five minutes of birth the patient developed generalized tonic-clonic seizures, which were controlled with intramuscular phenobarbital (5 mg per kilogram). A bulging fontanelle was noted. The initial head circumference (OFC) was 32.2 cm, greater than two standard deviations below the mean for an infant girl of 43 weeks' gestation. No focal neurological deficit was present.

At one day of age the fontanelle was still bulging, and intermittent, generalized seizure activity persisted. The patient was then admitted to the University of Utah Medical Center. Physical examination revealed a temperature of 98°F, respirations of 50 per minute, and a pulse of 120 per minute. The head transilluminated normally. The right pupil was dilated and fixed to light, and there was a left sixth nerve palsy. The latter findings were interpreted as signs of transtentorial uncal herniation. Hyperactive, symmetrical deep tendon reflexes and intermittent decorticate posturing were noted as well.

On admission the hematocrit was 75, polycythemic for a child of this age. Skull x-rays revealed diastasis of the sutures but no intracranial calcification. Serum calcium, phosphorus, glucose and electrolytes, urine amino acid screen, serum immunoglobulin-M, chest x-ray, karyotype, and cultures of the blood, cerebrospinal fluid and urine were normal.

Seizure activity was controlled with intramuscular phenobarbital. Increased intracranial pressure was treated with dexamethasone 2 mg I.V. every eight hours. Subdural and ventricular taps were performed in an attempt to determine the cause of the increased intracranial pressure. The subdural taps were negative, and the ventricular system could not be entered. Eleven hours after admission (two days of age) the patient had an episode of bradycardia and apnea and was promptly resuscitated. At that time bilateral fixed and dilated pupils were noted, as well as intermittent decerebrate posturing. Mannitol (1.5 gm per kilogram) was given without apparent improvement, and the patient was prepared for cerebral angiography.

A right carotid angiogram (fig. 1) revealed a stenotic lesion in the right external carotid artery a few millimeters beyond the carotid bifurcation, and an overall decrease in caliber of the right internal carotid with multiple discrete and diffuse stenotic lesions. The right ophthalmic artery filled well, but virtually no dye extended beyond the right carotid siphon. An aortic arch study revealed multiple similar stenotic lesions in the left internal carotid artery, minimal flow to the anterior circulation, and moderately decreased flow in the posterior circulation. An intravenous pyelogram was normal.

The patient died at two and one-half days of age.

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Pathological Examination
The brain was that of a term infant. There were bilateral recent infarcts in the distributions of both the middle and anterior cerebral arteries, as well as cerebral edema with evidence of transtentorial herniation. The area supplied by the posterior circulation was relatively well preserved. Microscopically these infarcts contained pericellular vacuolization, spongy change and petechiae, with no evidence of an inflammatory reaction. The left cerebellar hemisphere contained a vascular malformation formed of a collection of large, thin-walled vessels, but was otherwise normal. The brain stem was intact.

The walls of all four carotid arteries in the neck were markedly thickened. Stenosis was estimated at 70% to 80%. Microscopically there was dramatic reduplication of the internal elastic lamina, with no increase in smooth muscle cells or adventitia (fig. 2). No thrombi were seen. Carotid arteries above the siphon had normal walls grossly. Vessels within the brain and the rest of the body were normal on gross and microscopic examination.

Discussion
A number of pathological processes may lead to insufficient of the cerebral circulation in infancy and childhood. These include arteriosclerosis, arteritis of infectious or collagen vascular origin, embolism associated with congenital or rheumatic heart disease, subacute bacterial endocarditis, sickle cell disease, thrombosis associated with congenital heart disease, trauma or infection. The etiology in the majority of cases is unknown.

Harvey and Alword, in a classification of cerebral arterial lesions associated with "acute hemiplegia of childhood," cite several examples of cerebral arterial dysplasia including intimal hyperplasia, medial hypoplasia or atrophy, and elastic hypoplasia or atrophy. Fibromuscular dysplasia and Moyamoya disease also have been described as causes of cerebral ischemia, but we have
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FIGURE 2

Cross section of the right internal carotid artery, demonstrating dramatic reduplication of the internal elastic lamina, × 250.

found no reports of dramatic reduplication of the internal elastic lamina of the carotid arteries resulting in stenosis such as was seen in the case reported here. Search of the literature also reveals no similar-appearing arteriographical abnormalities of the carotid arteries. The lesion is unlike that seen in fibromuscular dysplasia.10–14

Our patient presents an intriguing combination of cerebral arterial dysplasia, microcephaly and polycythemia in a postmature infant, small for her gestational age. We speculate that a primary carotid artery dysplasia caused intrauterine cerebral ischemia and resulted in microcephaly.

Both histological and radiographical evidence supports the conclusion that carotid artery stenosis (70% to 80%) resulted in a significant decrease in cerebral blood flow. Polycythemia may have contributed to cerebral ischemia. Stasis of blood due to increased viscosity has been reported to produce seizure activity and focal neurological deficits such as hemiplegia in infants without arterial stenosis.15 The observed increase in intracranial pressure in this patient was the result of cerebral edema accompanying massive bilateral cerebral infarction.

The etiology of the complex constellation of findings in this patient is not known. She was microcephalic and demonstrated intrauterine growth retardation, both possible stigmata of intrauterine infection. Her mother did have four prior miscarriages, suggesting an adverse intrauterine environment or genetic defect. The patient's karyotype was that of a normal female, and the urine amino acid analysis showed no evidence of homocystinuria as a cause of the arterial disorder. These results, however, would not rule out a genetic defect.

References


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