EXTREMITY GANGRENE IN UTERO

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A neonate with extremity gangrene resulting from intrauterine embolization of infarcted placental substances is discussed. This rare clinical entity is thought to be most commonly a manifestation of embolic phenomenon during maturation of the neonatal circulatory system. Management of neonatal gangrene is conservative, delaying amputation as long as possible since the line of demarcation tends to migrate distally. Evidence of multiple emboli should be carefully sought prior to definitive treatment.

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Neonatal extremity gangrene is rare, and probably decreasing in incidence with improvements in perinatal care. Even rarer are those children born with evidence of intrauterine vascular occlusion. We present a case which illustrates the need for recognition of the signs of embolic disease and conservative management of peripheral infarction in neonates.

CASE REPORT

A 2.22 kg male infant was born with necrosis of the right forearm and hand (Fig 1). His 34-year-old mother was healthy but had a history of three previous spontaneous abortions. The pregnancy had been normal until the 34th week when the mother noted decreased fetal movements. Despite a normal non-stress test, she went into premature labour during the 35th week and delivered the child vaginally. Placental pathology included multiple infarcts and several sclerotic chorionic villi.

On examination, there was a large necrotic area on the ulnar border of the right forearm and hand (Fig 1). The tips of the right thumb, ring, and little fingers were ischaemic and the thenar eminence was oedematous. There was no ulnar pulse by Doppler at the wrist but the radial pulse was palpable. Radiographs of the limbs were normal (Fig 2), although a technetium bone scan showed absent uptake in the distal ulna (Fig 3).

The initial diagnosis was a hyperviscosity syndrome leading to ulnar artery thrombosis since the haematocrit was 0.7, so the patient underwent partial exchange transfusion to bring the haematocrit to 0.52. The coagulation profile including proteins C and S and antithrombin III was normal.

At 3 weeks of age, he underwent débridement of necrotic areas including the distal ulna and amputation of the ring and little fingers. Pathology showed coagulative necrosis.

Physiotherapy and right hand splinting were instituted and at 7 months the physiotherapist documented increased tone and neglect on the left side. A computed tomogram revealed multiple large cystic lesions in the right cerebral hemisphere suggesting massive intrauterine infarction in the right anterior and middle cerebral artery distribution (Fig 4).

At 1 year, he underwent a four-flap Z-plasty of the first web space and release of the ulnar aspect of the wrist with a full thickness skin graft. Splints were used postoperatively but the child developed progressive ulnar deviation of the forearm and wrist (Figs 5 and 6).

At 3 years his dominant right hand was functioning well and the left was used for two handed tasks. The ulnar wrist flexion deformity was more pronounced, and repeat radiographs showed little increase in the length of the right radius.

Fig 1 Right arm of neonate illustrating necrotic area on ulnar border, with gangrenous middle, ring, and little fingertips.

Fig 2 Normal radiograph of patient's right arm shortly after birth.

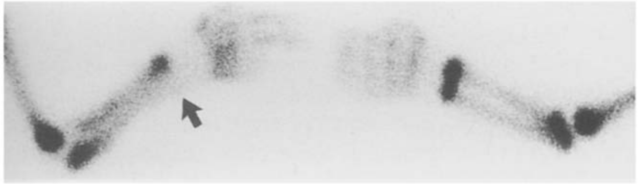

Fig 3 Technetium 99 bone scan of both arms showing absent uptake at right distal ulna (arrow).

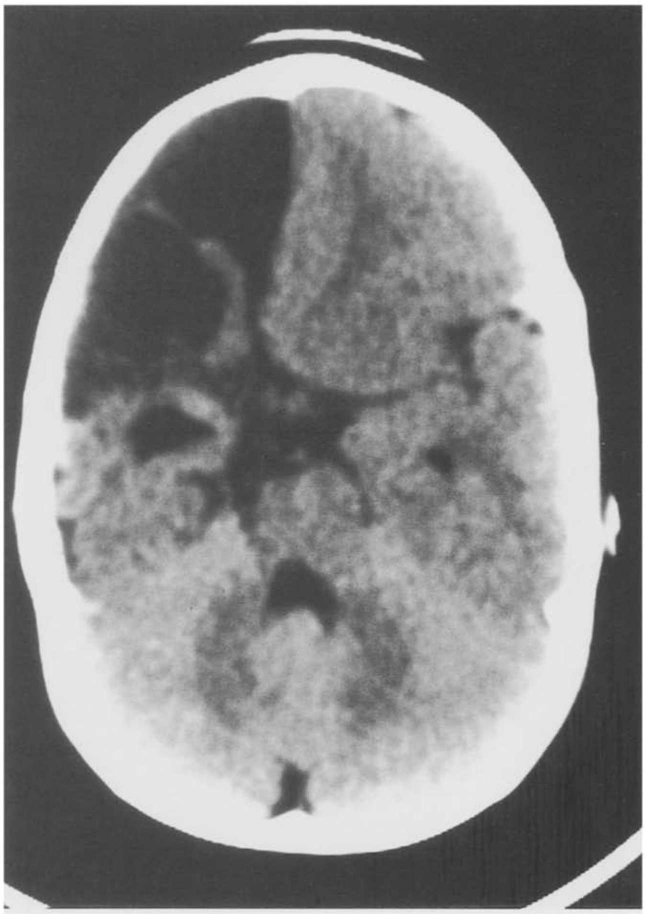

Fig 4 Computed tomogram of patient at 7 months of age. Multiple cystic areas in the anterior right hemisphere signify old infarcts in the anterior and middle cerebral artery territories.

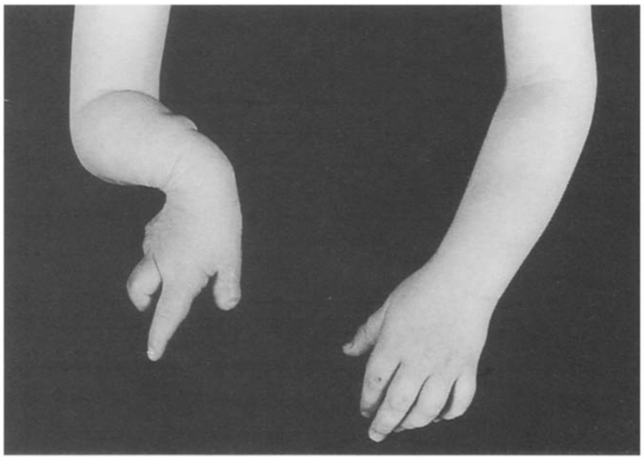

Fig 5 Patient at 18 months of age, illustrating short right forearm, 95° of ulnar deviation at the wrist, and absent ring and little fingers. The left arm, normally formed, is hypotonic.

Fig 6 Radiograph of right arm at 18 months of age, showing absent distal ulna, short bowed radius, and absent ring and little phalanges.

DISCUSSION

Over 60 cases of neonatal gangrene have been reported in the literature but less than one-third appear to have occurred in utero. There has been no attempt in the literature to differentiate between the aetiology and pathogenesis of this clinical entity in neonates versus in utero.

In 1945 Gross analysed the cases in the literature and concluded that although sepsis, congenital heart disease and birth trauma could result in neonatal gangrene, the most common cause was emboli from thrombosis during closure of the ductus arteriosus and umbilical arteries (Gross, 1945; Steiner, 1945). Dehydration and polycythaemia were later added as predisposing factors (Bhettay and Jacobs, 1977; Gross, 1945; Papageorgiou and Stern, 1972). Birth trauma was thought to cause compression of an artery, intimal adhesion or tearing resulting in arteriospasm and thrombosis with emboli formation (Gilbert et al, 1970; Heller and Alvari, 1941; Jaiyesimi et al, 1976; Papageorgiou and Stern, 1972; Perlmutter and Wagner, 1950). Today, emboli are most commonly iatrogenic following cannulation of the umbilical vessels (Bhettay and Jacobs, 1977; Bonifazi and Meneghini, 1980).

Maternal diabetes is thought to predispose to venous thrombosis (a source of emboli) although this connection was established by a retrospective study of neonatal autopsies and anecdotal evidence (Bhettay and Jacobs, 1977; Hensinger, 1975; Hoffman et al, 1974; Oppenheimer and Esterly, 1965; Papageorgiou and Stern, 1972; Valderrama et al, 1972; Ward, 1977). Congenital rubella and varicella have been reported to cause localized gangrene (Hoyme et al, 1981; Illingworth and Zachary, 1955). Placental infarcts, related to toxaemia, can cause embolization of necrotic tissue or thromboplastic substances through the patent foramen ovale (Jaiyesimi et al, 1976; Wigglesworth and Singer, 1991). Clark and Linell (1954) reported that the internal carotid artery in a dead neonate contained an embolus of necrotic material thought to originate from a small area of placental infarction.

Other factors can cause intrauterine gangrene. Heffelfinger and Harrison (1971) reported a child whose left leg became ischaemic 3 hours after birth, and in whom the involved vessel had a musculoelastic ridge at the site of thrombosis which occluded most of the lumen. Constriction bands associated with distal necrosis have been reported (Askue and Wong, 1952; Braly, 1965; Dohan, 1934).

Regardless of the pathogenesis of gangrene its clinical course in neonates is fairly reproducible. Initial pallor is followed by bluish mottling within a few hours. By 24 to 48 hours a line of demarcation develops which tends to be more proximal than the final level of tissue loss (Gross, 1945). Serial technetium scans reveal that the initial occlusion, correlating to the final degree of necrosis, is more distal than clinically apparent (Puppala et al, 1981).

Certain tenets of management of neonatal extremity gangrene have been accepted for over 50 years. The limb should be kept clean to prevent secondary infection and a variety of dressings have been advocated (Askue and Wong, 1952; Gross, 1945; Hensinger, 1975; Hoffman et al, 1974; Perlmutter and Wagner, 1950; Stadler, 1953; Stavorovsky et al, 1975). Hydration, nutrition, and antibiotics are widely accepted (Askue and Wong, 1952;

Hensinger, 1975; Hoffman et al, 1974; Siegel, 1971; Stavorovsky et al, 1975). Sympathetic blockade, anticoagulants, vasodilators and hyperbaric oxygen may improve circulation in the affected limb and have been used sporadically (Askue and Wong, 1952; Gross, 1945; Hoffman et al, 1974; Stavorovsky et al, 1975; Wiseman et al, 1977). Thrombectomy has proven successful in some cases (Braly, 1965; Wiseman et al, 1977). A conservative approach is considered important and amputation should be delayed as long as possible (Askue and Wong, 1952; Gross, 1945; Heller and Alvari, 1941; Hensinger, 1975; Hoffman et al, 1974; Perlmutter and Wagner, 1950; Siegel, 1971; Stavorovsky et al, 1975). The resilience of these young tissues is so impressive that some authors postulate that the vascular occlusion is merely a temporary agglutination of intimal surfaces, which pops back open much like a compressed garden hose (Perlmutter and Wagner, 1950). Once healing has ensued, splinting and range of motion exercises help prevent joint contractures (Hensinger, 1975).

We believe that in the child reported here substances from placental infarcts were carried in the fetal circulation, traversed the patent foramen ovale and ultimately lodged in the right cerebral system and right ulnar artery days prior to delivery. The brachiocephalic artery, feeding ultimately the right middle cerebral and ulnar arteries, is the first branch of the aortic arch, and so the first conduit an intracardiac embolus can enter. The large right cerebral vascular accident (CVA) was not noted initially since affected neonates tend to present with generalized hypotonia but no focal neurological deficits (Barmada et al, 1979). The child's haematocrit reflects his premature status and may not have played a role in the infarction.

The importance of follow-up in such unusual clinical situations is shown by the discovery of the right CVA, explaining why the child would ultimately function with his abnormal hand and practically ignore his perfectly formed left hand. Unfortunately, by the time the patient was born it was too late to attempt restoration of ulnar arterial flow since necrosis was already present. Amputation of the entire right hand would have been a tragedy.

Further treatment for this child may include radial osteotomy and wrist stabilization to improve the strength of his dominant hand.

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