Intrauterine lower extremity gangrene in a newborn with Tetralogy of Fallot

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DESCRIPTION

Intrauterine gangrene of an extremity is a rare problem. Lower extremity gangrene is even rarer.[1] We present a case of lower extremity gangrene in combination with Tetralogy of Fallot (TOF) and postaxial polydactyly of the hand. A live male infant was delivered by caesarean section. Delivery occurred at the 34th week of gestation due to meconium aspiration and intrauterine growth retardation. The mother was 35 years of age and had three healthy children. She had been diagnosed with gestational hypertension during this pregnancy. The newborn patient weighed 1740 g (10th centile) and his length was 43 cm (25th centile). After birth, he developed respiratory distress and was intubated. Examination revealed postaxial polydactyly with palpable phalanxes on the left hand, with an attached necrotic soft tissue mass (figure 1), as well as lower extremity gangrene with a visible demarcation line on the left middle thigh (figures 2 and 3).

Blood test results were normal except for a low platelet count of 73 000/mm^3 (normal range 130 000–510 000/mm^3), elevated D-dimer of 1154.88 ng/mL (normal range <500 ng/mL), low antithrombin III activity of 55% (normal range 70–125%) and low protein-S activity of 22% (normal range 33–93%).

Doppler ultrasonography revealed blocked arterial flow below the level of the tibioperoneal trunk. The main, deep and superficial femoral and popliteal arteries were patent. The patient also had TOF, which was diagnosed by a cardiologist. A transfemoral amputation and polydactyly excision were performed on the third day after birth. The histopathological investigation of the tissue blocks from the amputation material was not helpful in identifying the aetiology. On the 32nd day after birth, the patient was operated for TOF. He died on the seventh postoperative day due to cardiac arrest.

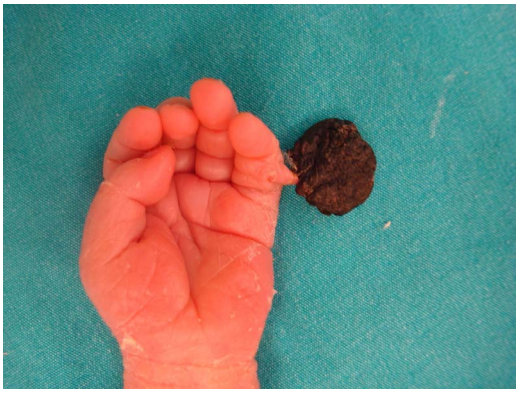

Figure 1 Postaxial polydactyly with an attached necrotic soft tissue mass.

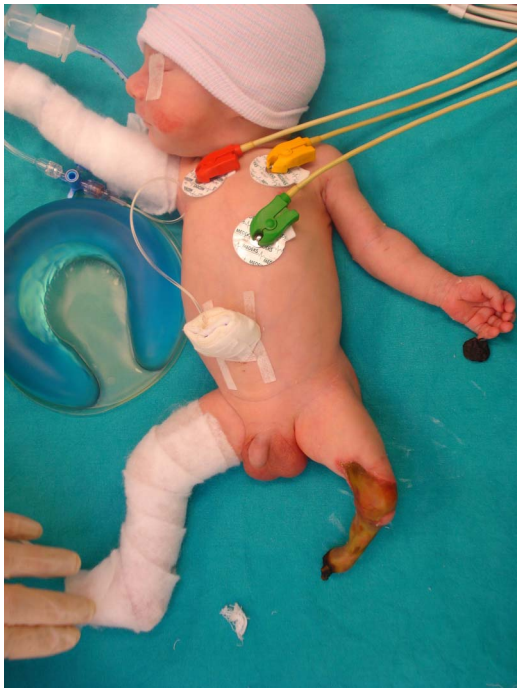

Figure 2 Infant with Tetralogy of Fallot, intrauterine lower extremity gangrene and postaxial polydactyly.

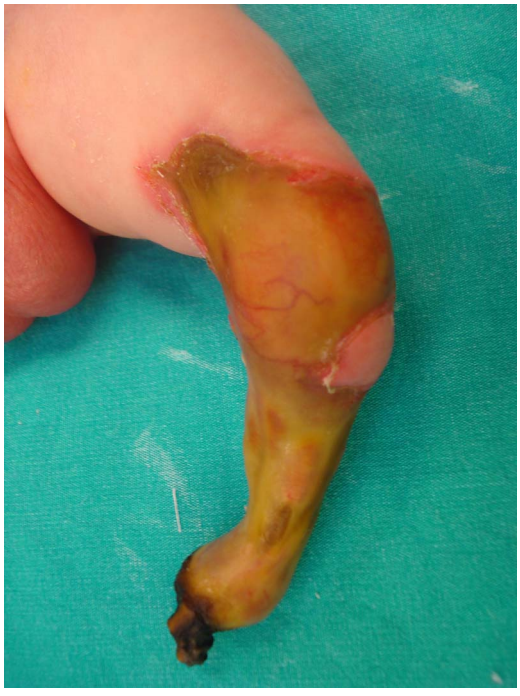

Figure 3 Visible transfemoral demarcation line.

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Various prenatal and perinatal factors can contribute to intrauterine gangrene, including compression or ischaemia. Compression can be caused by uterine anomalies, abnormal fetal presentation, and oligohydramnios or amniotic bands, while ischaemia can result from thrombosis or embolism.

Maternal diabetes, pregnancy-induced hypertension, preterm delivery, dehydration, polycythaemia, congenital heart disease, placental emboli, coagulation abnormalities and twin-to-twin transfusion syndrome may also be underlying causes. The patient had a low platelet count, but his antithrombin III and protein S activity were also decreased, which can be low in neonates and may cause a transient thrombophilic period. The addition of anomalies or other predisposing factors, such as those listed above, would increase the risk of thrombus formation despite a low platelet count.

In this case, another recognisable predisposing factor was gestational hypertension, which may cause ischaemia.[2] Although TOF is an accepted risk factor for upper extremity ischaemia and upper extremity anomalies,[3] there is no such report related to lower extremity ischaemia.

The treatment of intrauterine gangrene consists of maintaining homoeostasis and preventing infection, followed by amputation in the presence of a visible demarcation line.

Learning points

- Intrauterine ischaemia is a rare entity that may lead to devastating consequences such as amputation.
- Gestational hypertension is an important predisposing factor for intrauterine gangrene.

Contributors AS operated on the patient and wrote the manuscript with MEK. MM took the photographs. AK checked the final manuscript.

Competing interests None declared.

Patient consent Obtained.

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