BMJ Case Reports

Rare disease

High-output heart failure in a newborn

Maria Inês Mascarenhas,[1] Marta Moniz,[1] Sofia Ferreira,[1] Augusto Goulão,[2] Rosalina Barroso[1]

[1]Neonatal Intensive Care Unit, Department of Pediatrics-Hospital Prof. Doutor Fernando Fonseca, Lisbon, Portugal
[2]Neuro-Imagiology Department, Hospital Garcia de Orta, Lisbon, Portugal

Correspondence to Dr Maria Inês Mascarenhas, ines.mascarenhas@gmail.com

Summary

High-output cardiac failure is rare in newborns. Emergent diagnosis and management of this pathology is crucial. We report the case of a child, currently 12-months old; obstetric background is non-contributory. Clinic observation on D1 was normal except for the presence of a systolic cardiac murmur; cardiological evaluation revealed mild ventricular dysfunction of the right ventricle. On the third day of life, she developed cardiac failure with gallop rhythm, hepatomegaly and a murmur in the anterior fontanel; an echocardiogram confirmed clinic aggravation with biventricular dysfunction and right cavities and superior vena cava dilatation. The cranial MRI confirmed the presence of a pial arteriovenous malformation (AVM) involving the anterior and middle cerebral arteries with an associated fronto-parietal ischaemic lesion. The infant underwent embolisations of AVM with successful flow reduction and cardiac failure improvement. The multidisciplinary follow-up showed no cardiac dysfunction or permanent lesions but confirmed a severe psycho-motor delay and left hemiparesia.

BACKGROUND

Systemic disorders are the most common manifestations of intracranial arteriovenous diseases in neonatal age and cardiac failure is frequently encountered.[1] In this group the most common is vein of Galen malformation (VGM). However, non-galenic cerebral arteriovenous malformations (AVMs) or pial AMVs (PAVMs), although rare in newborns, present much like VGM, also causing congestive cardiac failure in the newborn.[1 2]

The aetiology remains unknown; their presence in newborns suggests that it is a developmental condition.[3] However, it is unclear whether PAVMs form early or late in prenatal development.

Emergent diagnosis and management is crucial as mortality is high during the neonatal period if no treatment is offered. Generally medical treatment is inefficient, requiring therapeutic embolisation.[4]

Antenatal diagnosis has been associated with improved outcome; however, some of these lesions can remain undetected in prenatal ultrasound.[5]

We report a case of PAVM resulting in neonatal cardiac failure, requiring embolisation, which was not detected in second and third trimester obstetric ultrasound.

CASE PRESENTATION

A 39-week, 2735 g girl was born via caesarean delivery due to breech presentation, to a healthy 36-year-old woman. It was an uneventful pregnancy, with normal TORCH serology, and the prenatal screening ultrasounds performed at 14, 23 and 36 weeks gestational age were described as normal, including the central nervous system. Amniocentesis was performed at 16 weeks gestational age due to maternal age and the karyotype was normal (46, XX).

Apgar score was 9 and 10 at minute 1 and 5, respectively, and she was admitted in co-rooming with her mother.

She was evaluated on D1, without feeding intolerance and a normal physical exam except for a systolic cardiac murmur II/VI. A cardiological evaluation was performed describing a mild ventricular dysfunction especially in the right ventricle and she was admitted to the neonatal intensive care unit for surveillance.

On her third day of life she developed feeding intolerance, tachypnoea and intercostals and supraclavicular retractions, grunting; systolic cardiac murmur III/VI and a continuous murmur in the anterior fontanel; she remained afebrile. Analytical tests were normal except for respiratory acidosis (pH 7.13, PCO_2 45.5, HCO_3 15.8, BE −8.8, lactate 7). Chest radiography revealed an enlarged toraco-cardiac index (figure 1) and echocardiogram demonstrated enlarged right cavities and a superior vena

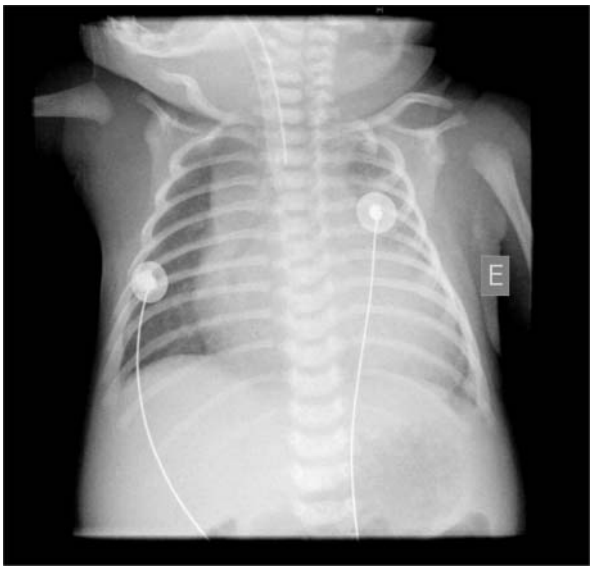

Figure 1 Thoracic radiography with cardiomegaly.

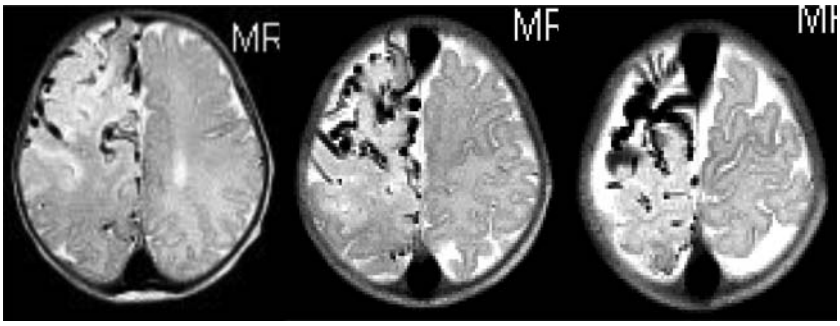

Figure 2 MRI performed on D7 demonstrated right frontoparietal pial arteriovenous malformation involving the anterior and middle cerebral arteries and dilatation of dural venous sinuses. A right ischaemic lesion was also described near the AVM.

cava with moderate ventricular dysfunction. On day 4, she was entubated and ventilated.

INVESTIGATIONS

After clinical stabilisation on life day 7 she performed an MCR-CE which confirmed the presence of a right frontoparietal PAVM involving the anterior and middle cerebral arteries and dilatation of the dural venous sinuses, including vein of Galen, secondary to the AVM. A right ischaemic lesion was also described near the AVM (figure 2).

TREATMENT

She was transferred to a specialised centre and on day 17 of life an angiography with endovascular embolisation was made. A PAVM with multiple high-flow arteriovenous fistulas was found. Embolisation was performed using detachable coils and *N*-butyl cyanoacrylated coil and butylcyanoacrylate glue resulted in partial occlusion of the fistula.

On life day 25, she was submitted to a second embolisation, which was unsuccessful, and 5 days later she was diagnosed with a late-onset sepsis by *Staphylococcus hominis*; she was transferred back to our hospital and vancomycin and ceftriaxone were initiated, completing 2 weeks of antibiotics.

She maintains clinical signs of mild-to-moderate compensated cardiac failure with systolic cardiac murmur, gallop rhythm, mild tachypnoea and hepatomegaly (two fingers below the costal limit), without necessity of ventilatory support and treated with ionotropics and milrinona.

A third intervention was performed on life day 50 with embolisation of four more branches of PAVM, resulting in a marked flow reduction (figure 3). Cardiological evaluation postembolisation revealed a marked improvement in ventricular function with right cavities with a lesser degree of dilatation. The patient gradually improved and inotropic therapy was discontinued.

OUTCOME AND FOLLOW-UP

A development and neurological evaluation was performed: the patient had global axial hypotonia and hyperreflexed deep tendon reflex in lower limbs and mild extrapyramidal signs.

The transfontanelar ultrasound demonstrated extensive cystic encephalomalacia in the right hemisphere with frontoparietal white substance atrophy.

She was discharged home at 2 months of age on no cardiac medications.

At the time of writing, the infant is 12 months of age; she has moderate psychomotor delay and left hemiparesis. The latest MRI revealed right hemispheric atrophy and a residual rim of PAVM.

DISCUSSION

PAVM only represents slightly more than one-quarter of cerebral AVMs in newborns;[1] they correspond to abnormal communications between arteries and veins directly and may be located in the posterior fossa or more commonly in the supratentorial space.[2]

The intrauterine diagnosis of this vascular anomaly has been facilitated by prenatal ultrasonography and Doppler screening. However, as occurred in our case, a PAVM can remain clinically occult in utero while the fetus is supplied from the placental circulation and after birth as

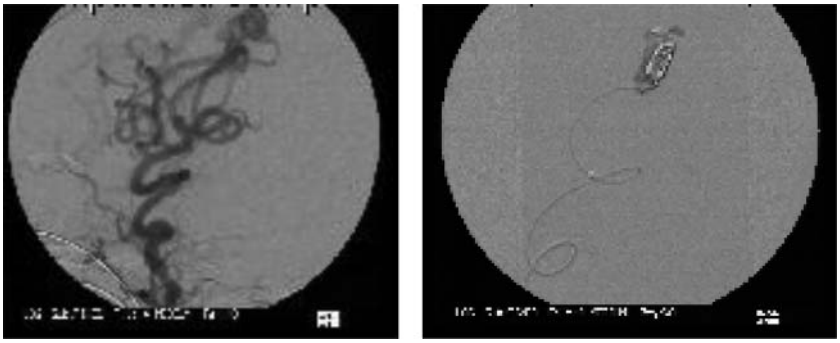

Figure 3 Embolisation of one of the main feeders with coils and glue.

BMJ Case Reports 2012; doi:10.1136/bcr-2012-006289

the arterial pressure increases and venous resistance diminishes, the shunt grows in size and flow, producing haemodynamic decompensation and cardiac failure.[3–5] Damage to the brain can occur, depending on the volume of blood shunted through the malformation and away from the brain parenchyma causing brain ischaemia ('steal phenomenon').[2]

Generally shunts that are detected in fetal life are always associated to cardiac failure at birth and poorer prognosis.

The main differential diagnosis is the vein of Galen aneurismal malformations (VGAM) and this differentiation is of paramount importance. In VGAM, management can be delayed by 5–6 months whereas in PAVM, because cerebral tissue injury can develop rapidly, the aim is to treat as quickly as possible and reduce significantly shunting through AVM, as soon as possible after delivery in order to avoid loss of brain substance and to allow neurocognitive development and normal brain maturation.[2 3]

The chosen treatment is embolisation. Medical management can rarely control cardiac failure[1 2 6] and endovascular therapy can provide good outcomes even in patients with severe cardiac failure and large AVM. Two techniques are most commonly used: (A) Transarterial embolisation is more effective in controlling heart failure mainly when there is only one or a limited number of arteries; this technique was used on our patient. (B) Transvenous embolisation is preferred when there are numerous small arterial feeders and is used by some authors in selected AVM cases.[4 6]

The main goal is to obliterate the AVM completely, as soon as possible. In some cases this is not possible and partial occlusion is also very beneficial for the improvement of cardiac function.[4]

The management of neonates with PAVM remains challenging and implies a multidisciplinary approach to accomplish the best possible outcome.

Learning points

- Systemic disorders are the most common manifestations of intracranial arteriovenous diseases in neonatal age.
- Pial arteriovenous malformation in the newborn presents as a high-output cardiac failure.
- Its identification and rapid treatment are crucial due to risk of severe and possible irreversible cardiac lesions and brain ischaemia leading to impairment in neurocognitive development.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Rodesch G,** Malhet V, Alvarez H, *et al*. Nongalenic cerebral arteriovenous malformation in newborns and infants: review of 26 consecutive cases (1982–1992). *Childs Nerv Syst* 1995;**11**:231–41.
2. **Garel C,** Azarian M, Lasjaunias P, *et al*. Pial arteriovenous fistulas: dilemmas in prenatal diagnosis, counselling and postnatal treatment: report of three cases. *UltrasoundObstet Gynecol* 2005;**26**:293–6.
3. **Potter C,** Armstrong-Wells J, Fullerton HJ, *et al*. Neonatal giant pial arteriovenous malformation: genesis or rapid enlargement in the third trimester. *J Neurointerv Surg* 2009;**1**:151–3.
4. **Koroglu M,** Çil B, Yeseldag A, *et al*. Prenatal diagnosis in intracranial pial arteriovenous fistula and endovascular treatment in neonatal period. *Diagn Interv Radiol* 2006;**12**:64–7.
5. **Reyal F,** Vuillard E, Sibony O, *et al*. Prenatal diagnosis of pial-superficial arteriovenous malformation. *Fetal Diagn Ther* 2002;**17**:255–6.
6. **Hara H,** Burrows PE, Flodmark O, *et al*. Neonatal superficial cerebral arteriovenous malformation. *Pediatr Neurosurg* 1994;**20**:126–36.