Cavernous haemangioma causing high-output cardiac failure in a neonate

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Infantile haemangiomas are benign tumours originating from the vascular endothelium, representing one of the most common neoplasms in childhood. We present the unique occurrence of a neonate born at term with a large, cavernous haemangioma on the scalp. On the second day of life, the neonate developed high-output cardiac failure, characterised by echocardiographic findings of a dilated right atrium and right ventricle, as well as tricuspid regurgitation, without evidence of structural abnormalities or arteriovenous fistulas. The neonate also developed Kasabach-Merritt syndrome. Despite optimal therapy for cardiac failure, the neonate's condition deteriorated, leading to his unfortunate demise. An academic postmortem confirmed the presence of a cavernous haemangioma, highlighting the need for heightened awareness of atypical presentations of neonatal vascular anomalies.

Keywords. infantile haemangiomas; high-output cardiac failure; neonate; Kasabach-Merritt syndrome.

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Infantile haemangiomas (IHs) are benign tumours arising from the vascular endothelium and are one of the most prevalent tumors during childhood.[1] Despite their generally benign and self-limiting nature, certain haemangiomas can lead to complications, such as ulceration or significant disfigurement, with the potential to impact the affected individual's quality of life, both biologically and psychologically. In some instances, haemangiomas may even compromise vital organ function or occur concomitantly with developmental anomalies, as elucidated in the subsequent case report, where they can precipitate high-output cardiac failure (HOCF) leading to morbidity.

IHs represent the predominant soft-tissue tumours observed in infancy, typically manifesting within the initial weeks of life, undergoing a phase of active growth, and subsequently exhibiting spontaneous involution.[1] Most IH cases follow a benign course, necessitating no therapeutic intervention, as they regress autonomously. However, a subset, ~10% to 15% of IH cases, demands treatment owing to complications such as the risk of disfigurement, functional impairment, obstruction of vital structures, or ulceration.[2]

Childhood vascular anomalies encompass a diverse range of disorders, including vascular tumours and malformations, spanning minor 'stork bites' to severe, life-threatening conditions with consequential morbidity and mortality.[3] Neonatal vascular tumours, comprising haemangiomas or kaposiform haemangioendothelioma (KHE), alongside vascular malformations, manifest as developmental aberrations affecting arterial, venous, or lymphatic vessels which is described by the International Society for the Study of Vascular Anomalies.[1]

KHE is commonly associated with the Kasabach-Merritt phenomenon (KMP), characterised by low platelet levels and hypofibrinogenaemia, posing a substantial risk of bleeding secondary to consumptive coagulopathy, with a mortality rate ranging between 20 and 30%.[4,5] The prevalence of KHE is noted at 0.9 per 100 000 children, with ~50% of cutaneous lesions being evident at birth.[5] Tufted angiomas (TA) present as infiltrated firm patches, plaques or nodules and may also be linked to KMP.[6] Neonates exhibiting KMP typically display progressive lesion enlargement and purpura, with 42 to 71% of neonates with TA and KHE developing KMP, especially if the lesions exceed 8 cm in diameter.[4] Surgical excision is the preferred treatment for small, localised tumours.

For large, non-resectable tumours, the recommended therapeutic approach involves vincristine and corticosteroids.[7,8] Neonatal vascular tumours precipitating HOCF early in life are not extensively documented in the literature. The current case report presents a unique and rare instance of a cavernous haemangioma contributing to HOCF within 48 to 72 hours of life. Owing to the large size of the lesion, the low platelet count was attributed to KMP.

Case

Our case was a 3 200 g male neonate who was delivered at 39 weeks' gestation via normal vaginal delivery to a healthy 29-year-old multiparous woman at a community clinic. The mother was RVD-reactive and virally suppressed, with a CD4 count greater than 200 cells/μL at a community health clinic. No prenatal ultrasound documentation was recorded. The mother booked her pregnancy at 21 weeks' gestation at a local clinic. Her antenatal booking bloods were noted to be RPR-non-reactive and rhesus-positive. In terms of medication, the mother was on iron supplements, folate and antiretroviral therapy.

The baby was born at a local clinic with Apgar scores of 6 and 10 at 1 and 5 minutes, respectively. The newborn had a large mass over the posterior aspect of the scalp, extending to the neck, prompting referral to a tertiary referral hospital for further evaluation. Upon arrival, the infant appeared comfortable on room air, pink and well hydrated, with a capillary refill time of less than 2 secs, and alert. The vital signs were recorded as normal with a respiratory rate of 52 breaths per minute, oxygen saturation of 92% (on room air), blood pressure of 62/48 mmHg (mean arterial pressure of 51 mmHg), bedside glucose of 3.9 mmol/L and temperature of 36.5 °C. The neonate appeared mildly tachypnoeic on respiratory examination

with good air entry bilaterally and no evidence of a pneumothorax. On cardiovascular examination, the neonate was noted to have a hyperdynamic precordium with a poorly defined apex beat roughly around the 6th ICS, anterior clavicular line. On abdominal examination, a 2 cm non-pulsatile liver was noted with no bruit. The central nervous system (CNS) examination was unremarkable with no spinal abnormalities. On musculoskeletal examination, an incidental left clavicular fracture was found, with no restriction in range of motion.

A detailed examination of the mass revealed a substantial posterior scalp mass, measuring over 10 cm in diameter and extending from the postauricular area of the left ear to the right ear, and inferiorly to the neck and upper back (Fig. 1). The lesion presented as an erythematous plaque with multiple telangiectasias and hyperkeratotic skin. Notably, the mass was non-pulsatile and devoid of a bruit and ulceration. Additionally, there was evidence of a malformed left ear owing to pressure from the mass. No other lesions were observed on the body. No bruit was detected over the fontanels or over the liver, indicating an absence of a possible arteriovenous malformation. The differential diagnosis included a large congenital haemangioma with Kasabach-Merritt phenomenon arteriovenous (AV) malformation, as well as PHACE (posterior fossa malformations, large facial haemangiomas, cerebral arterial anomalies, cardiovascular anomalies, and eye anomalies) syndrome (in consultation with dermatology at Victoria Mxenge Hospital). Kasabach-Merritt phenomenon was suspected in view of the low platelet count associated with a large haemangioma.

A complete blood count at admission revealed a low platelet count of 72×10^9/L with a normal white cell count and haemoglobin level. The coagulation screen done on day 2 could not be interpreted as the specimen was insufficient. On day 2 of life, the infant developed increasing respiratory distress with worsening hypoxia, necessitating intermittent positive pressure ventilation (IPPV). The neonate was noted to be in HOCF with wide pulse pressures and increasing hepatomegaly.

Despite interventions including diuretics, a beta-blocker (propranolol) and inotropes, the neonate remained in HOCF, characterised by a non-responsive condition and a wide pulse pressure. Within two hours of IPPV, the baby's condition continued to deteriorate. Echocardiography conducted by a paediatric cardiologist revealed a dilated right ventricle and right atrium, as well as tricuspid regurgitation, consistent with HOCF. A chest X-ray showed cardiomegaly (Fig. 2). Cranial ultrasound did not indicate intracranial extension, and no intracranial masses were observed.

Owing to the rapid and early onset of the HOCF, as well as its progression within 24 to 48 hours of admission, the prognosis was poor. Unfortunately, the neonate succumbed to his condition. An academic postmortem was consented to by the mother, revealing a haemangioma containing 85 mL of blood, with no intracranial extension. Given the association with low platelets, Kasabach-Merritt syndrome was considered likely, in view of the large collection of blood in the haemangioma. The postmortem further identified thymus hypertrophy and diffuse organomegaly, including an enlarged heart, liver, kidneys and spleen.

The histology report confirmed a cavernous haemangioma, while lung histology indicated stage 2 pulmonary hypertension. Examination of the brain during the postmortem revealed no evidence of hypoxia.

Discussion

A cavernous haemangioma inducing HOCF is rare, with few documented cases in the literature.[9] HOCF arising from arteriovenous (AV) malformations is exceptionally rare in newborns, emphasising the critical need for prompt diagnosis and management.[10] This form of heart failure is characterised by heightened cardiac output, diminished systemic vascular resistance (attributed to peripheral vasodilation or arteriovenous shunting), and a reduced arterial-venous oxygen content difference. Some cases also involve increased oxygen consumption, reflecting an elevated metabolic demand.[11] The neonate in this case exhibited these characteristics in both clinical presentation and echocardiographic findings.

In the neonatal population, systemic disorders often manifest as intracranial arteriovenous diseases, frequently resulting in cardiac failure. The most common among these is the Vein of Galen malformation (VOGM). However, non-galenic cerebral arteriovenous malformations (AVMs) or pial AVMs (PAVMs), although rare in newborns, present similarly to VOGM, leading to congestive cardiac failure in affected infants.[12]

Giant cutaneous haemangiomas also have the potential to induce HOCF.[13,14] While 50% of cutaneous lesions are visible at birth, the remaining typically emerge within the first two months of life. In rare circumstances, high-flow arteriovenous shunting in giant or multiple cutaneous haemangiomas can precipitate the development of HOCF.[13] In our case, the histology was suggestive of a cavernous haemangioma.

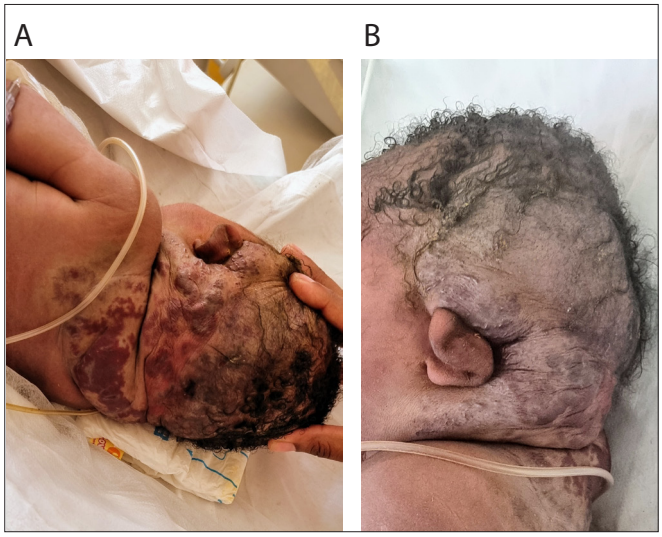

Fig. 1. A mass extending inferiorly to the neck and laterally to the temporal region and beneath the ears, causing anterior displacement of the ear. The overlying skin is hyperkeratotic with multiple telangiectasias.

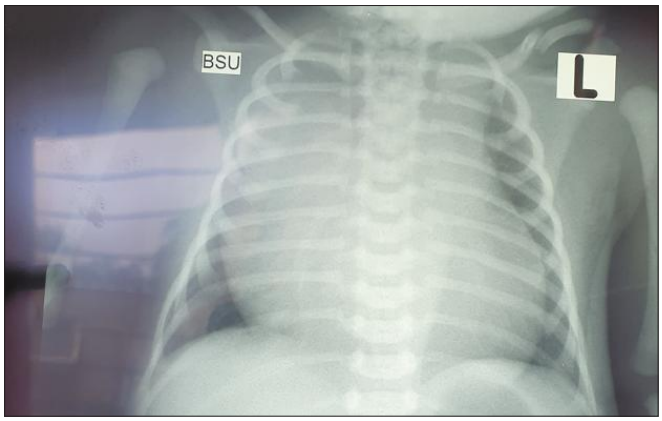

Fig. 2. Chest X-ray showing cardiomegaly and left clavicular fracture.

In this particular case, the infant presented with a solitary and large cavernous haemangioma and was asymptomatic at birth, with a fulminant course to which the neonate succumbed.

For an all-encompassing understanding, a computed tomography angiogram would have been beneficial; however, the neonate succumbed prior to this being done.

Conclusion

The occurrence of HOCF secondary to a cavernous haemangioma with a profound course in a neonate is rare, with few described cases in the literature. In the present report, we have described the first such case in the neonatal unit at Victoria Mxenge Hopsital in KwaZulu-Natal. The precise pathophysiology orchestrating this outcome remains elusive and is a phenomenon all clinicians should be aware of.

Declaration. Verbal and written consent was obtained from the guardian of the neonate for their anonymised information to be published in the present manuscript.

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Author contributions. PS: contributed towards writing up the case report and discussion; RS: literature review, discussion and conclusion.

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