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Arteriovenous Malformation of the Upper Extremity in a Neonate: Case Report

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Abstract

We report the case of a 12-day-old neonate, born by cesarean section at 36 weeks of gestation, with normal prenatal follow-up and no relevant findings on fetal ultrasounds. At birth, the infant developed transient tachypnea, requiring brief oxygen therapy. During hospitalization, the mother noted extensive ecchymosis of the left upper limb, accompanied by generalized jaundice, severe anemia, and profound thrombocytopenia, prompting intensive management with blood products, sedation, inotropic support, and ventilatory assistance. Active bleeding was observed in the affected extremity, along with severe coagulopathy. Echocardiography and arterial Doppler ultrasound were normal; however, surgical exploration revealed venous bleeding and a dissecting hematoma involving the arm, forearm, and hand, which was drained.

Subsequently, in the NICU the patient required advanced management, including mechanical ventilation and hemodynamic support. During hospitalization, a diagnosis of arteriovenous malformation (AVM) associated with kaposiform hemangioendothelioma (KHE) was established, with complications including consumptive coagulopathy (Kasabach–Merritt phenomenon), mineralizing vasculopathy, hypoxic-ischemic encephalopathy, and grade II intraventricular hemorrhage. Treatment included folic acid, iron, prednisone, propranolol, and sirolimus, with good clinical progression: reduction of the hematoma, absence of pain or infection, and preserved limb mobility. After 50 days of hospitalization, the patient was discharged in stable condition with good oral tolerance.

Introduction

Arteriovenous malformations (AVMs) are congenital vascular anomalies characterized by direct connections between arteries and veins without an intervening functional capillary bed, generating high-flow shunts and diverse hemodynamic complications (1). Although AVMs may occur in any anatomical region, approximately 10% are located in the upper extremities, with the hand being more frequently affected than the arm, following only the head and neck in anatomical distribution (2).

In the neonatal period, upper extremity AVMs are particularly rare but may present with aggressive clinical behavior. A recently published case described a high-flow AVM in the arm presenting with a soft-tissue mass and high-output cardiac failure in a neonate, confirmed by Doppler and MR angiography,

and managed with respiratory support, sirolimus, and percutaneous embolization, resulting in hemodynamic stabilization and reduction of lesion size (3).

The pathophysiology involves high-flow hemodynamics that may lead to high-output cardiac failure, pulmonary hypertension, and cardiac dilation, requiring urgent intervention in many cases. Diagnosis typically requires a multimodal approach: Doppler ultrasound is essential for identifying rapid flow and characteristic shunting; MRI delineates the anatomical extent of the AVM, its relationship with adjacent structures, and distinguishes high- from low-flow malformations. In selected cases, angiography is valuable for planning interventions such as embolization. Optimal management of neonatal AVMs requires multidisciplinary coordination. Therapeutic options include medical therapy (sirolimus), percutaneous embolization, surgical resection, or endovascular approaches (4).

Case Report

A 12-day-old neonate, born by cesarean section due to breech presentation with nuchal cord at 36 weeks of gestation (Apgar 8/9), required 3 hours of head-helmet oxygen therapy due to transient tachypnea. Prenatal history included a 30-year-old primigravida mother with normal prenatal check-ups, negative serology, and treatment with progesterone, calcium, folic acid, iron, and nifedipine for cervical insufficiency. Prenatal ultrasounds were normal, and complete pulmonary maturation was administered at 29 weeks. Maternal and neonatal blood type was O positive. Anthropometric measures were >50th percentile (weight 3.8 kg, length 50 cm, head circumference 36 cm, abdominal circumference 31 cm).

The mother noted ecchymosis involving the entire left upper limb (Figure 1), and the newborn developed generalized jaundice managed with phototherapy. Initial labs showed severe anemia and profound thrombocytopenia, requiring central venous access for intensive management with blood products, fluid resuscitation, sedation, and inotropic support. Coagulation tests (PT, aPTT, and fibrinogen) were non-coagulable. Active bleeding from the

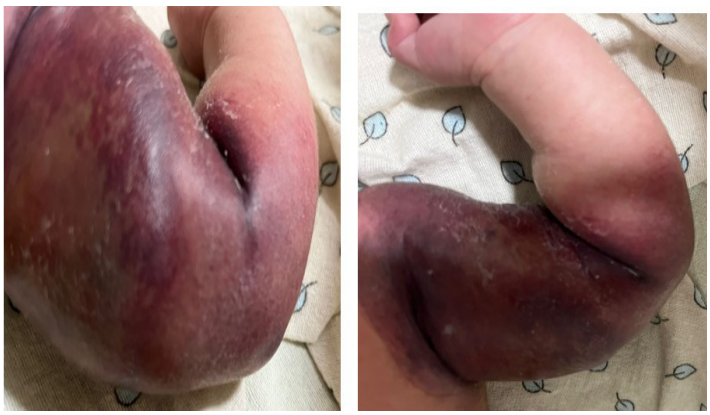

Figure 1: *Extensive ecchymosis of the left upper limb*

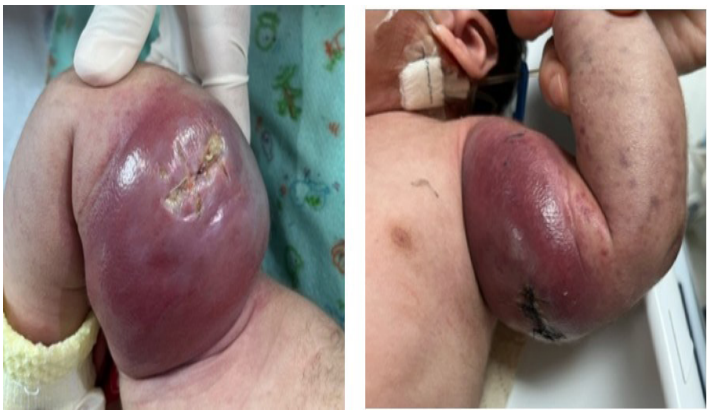

Figure 2: *Post-surgical evolution*

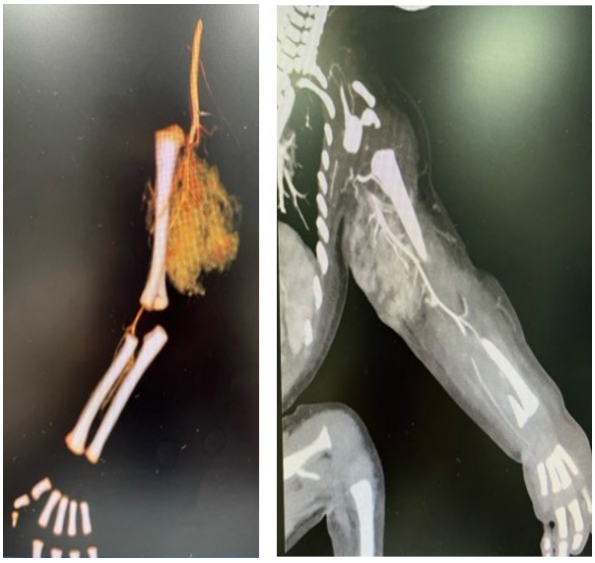

Figure 3: *CT angiography of the left upper extremity*

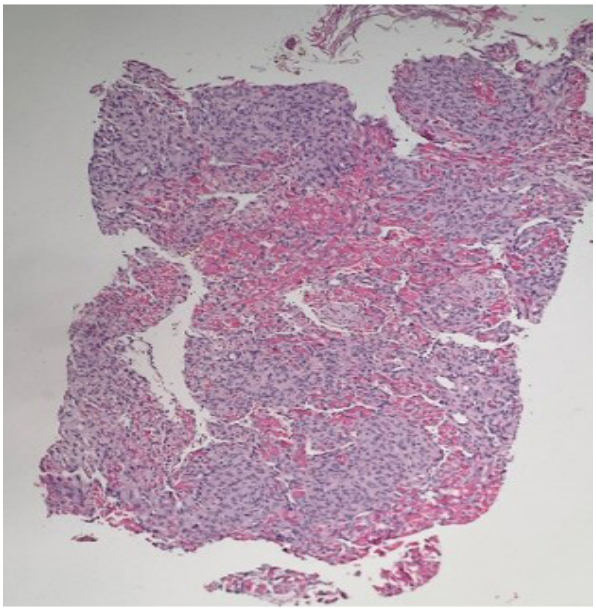

Figure 4: *Histological findings of the biopsy*

Figure 5: *Timeline of events*

left arm lesion was noted. Upon admission (06/06/25, 12th day of life, corrected GA 37.5 weeks), the patient exhibited hemodynamic instability with poor perfusion, grunting, bradycardia, and shallow respirations, leading to advanced airway management with a 3.5-mm endotracheal tube placed at 10 cm.

Color Doppler echocardiography and arterial Doppler ultrasound were normal, with no signs of active arterial bleeding. Pediatric and vascular surgery teams performed surgical intervention, revealing venous bleeding and extensive dissecting hematoma involving the arm, forearm, and hand, which was drained.

The patient was admitted to the NICU with mechanical ventilation and inotropic support. One week later, vasoactive medications were weaned off, the infant no longer required supplemental oxygen, and enteral feeding was initiated. Along with the diagnosis of AVM, additional findings included mineralizing vasculopathy, hypoxic-ischemic encephalopathy, and grade II intraventricular hemorrhage. Follow-up CT angiography showed a kaposiform hemangioendothelioma versus tufted angioma with probable vascular shunt in the superior lingula.

Biopsy confirmed kaposiform hemangioendothelioma. Medical treatment included folic acid, polymaltose iron, prednisone, propranolol, and sirolimus. After 50 days of hospitalization, the patient was discharged with good oral tolerance, preserved mobility, and a regressing hematoma without signs of infection or pain.

Discussion

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor characterized by locally aggressive behavior and strong association with consumptive coagulopathy known as Kasabach–Merritt phenomenon (KMP). Its estimated incidence is approximately 0.071 per 100,000 children, and more than 70% of cases develop KMP, significantly increasing neonatal morbidity and mortality (5).

In this case, localization in the left upper extremity is consistent with the literature, which reports the trunk and extremities as the most common sites (approximately 30% each) (6). Clinically, KHE typically presents as a firm, infiltrative, purpuric mass with a tendency for rapid progression. The presence of severe thrombocytopenia and consumptive coagulopathy strongly supports the diagnosis of KMP, a finding commonly reported in the literature (7,8).

Imaging plays a key diagnostic role. Doppler ultrasound generally shows hypervascularity with low-resistance flow; MRI typically demonstrates heterogeneous masses with high T2 signal and irregular contrast enhancement (9). These findings, along with histological confirmation—characterized by spindle-shaped endothelial cell proliferation positive for CD31, CD34, and D2-40—help differentiate KHE from other vascular tumors such as infantile hemangioma (10).

Management of KHE with KMP is challenging. Corticosteroids and vincristine were historically used as first-line therapy, but over the past decade sirolimus has emerged as the preferred treatment, demonstrating high sustained clinical and hematologic response rates both in monotherapy and combined regimens (11,12). In this case, sirolimus administration was associated with progressive improvement of the lesion and resolution of coagulopathy, in line with current evidence.

Atypical presentations have been reported, including prenatal diagnoses with non-immune hydrops fetalis and hemodynamic compromise, as well as deep lesions without

visible cutaneous manifestations (13,14). These cases illustrate the clinical heterogeneity of KHE and highlight the need for high clinical suspicion when evaluating neonatal vascular masses, particularly if associated with severe hematologic abnormalities.

In summary, this case reinforces the importance of early diagnosis and multidisciplinary management of neonatal KHE. The use of sirolimus as first-line therapy is well supported by current literature and constitutes an effective strategy to improve prognosis and reduce complications related to KMP.

Conclusion

This case represents a typical presentation of KHE with KMP in a neonatal upper extremity, closely aligned with the clinical profile described in the literature. Accurate diagnosis required targeted imaging and histopathological evaluation, both essential components according to recent evidence. Management with sirolimus—particularly as one-year monotherapy—is consistent with current guidelines and studies demonstrating high efficacy and tolerability. Atypical presentations (prenatal or without visible cutaneous involvement) underscore the importance of maintaining high suspicion in cases of unexplained coagulopathy or vascular masses in neonates.

Informed Consent: Informed consent was provided by the mother, the patient's legal representative.

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