Neonatal Compartment Syndrome as a Result of Thromboembolic Event

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Summary: Neonatal compartment syndrome is a rare condition characterized by progressive limb ischemia and tissue necrosis manifesting at birth or in the immediate postpartum period. Early recognition of clinical features and immediate surgical intervention offer the best prognosis. Underestimating the urgency of these findings often results in catastrophic consequences, including limb amputation. We present a case of a neonate with in-utero limb ischemia that resulted in the formation of a proximal arterial thrombus and subsequent compartment syndrome.

Neonatal compartment syndrome (NCS) is a rare condition characterized by progressive limb ischemia and tissue necrosis manifested at birth or in the immediate postpartum period. Early recognition of clinical features and immediate surgical intervention offer the best prognosis. Focal skin necrosis is often the earliest sign of NCS. Underestimating the urgency of these findings often results in catastrophic consequences, including limb amputation. We present a case of a neonate with in-utero limb ischemia that resulted in the formation of a proximal arterial thrombus and subsequent compartment syndrome.

CASE REPORT

A 31 weeks and 1 day baby girl was born to a 28-year-old mother via emergent cesarean section due to preeclampsia with severe features and polyhydramnios. Maternal medical history was significant for poorly controlled diabetes mellitus and spina bifida. Within 1 hour of birth, the patient developed diminished right arm distal pulses with areas of cyanosis and ecchymosis extending from the upper arm to the hand. Conservative management overnight with observation found return of pink coloration to the hand (Fig. 1). Initial ultrasound imaging was negative for arterial thrombus and fractures.

On the second day of life, dramatically increased upper extremity swelling, faint radial pulse, nonpalpable ulnar pulse, and pinpoint cyanosis with prolonged capillary refill on the tip of her fifth finger were noted. The patient was immediately transferred to a pediatric tertiary care facility. At physical examination, the patient appeared in significant discomfort and the compartments on the right upper extremity were found to be firm from the forearm to the hand. Severe swelling of the right arm with dorsal forearm blistering, nonpalpable radial and ulnar pulses, and lack of spontaneous right upper extremity movement were also noted. Ultrasound demonstrated a completely occlusive right mid to distal subclavian artery thrombus. Compartment syndrome was diagnosed, and emergent surgical fasciotomies of the right arm, forearm, and hand were performed (Fig. 2). The proximal arm musculature appeared well perfused; however, the forearm muscles appeared white and edematous (Fig. 2). At the end of the procedure, the patient’s hand was warm and pink, but radial and ulnar pulses remained weak. The patient was considered too high risk for tissue plasminogen activator, but was placed on a heparin drip starting at 28 U/kg/h, that was titrated to achieve an unfractionated heparin level between 0.35 and 0.7 U/mL. Full hematologic workup was initiated. Over the next several days, the patient developed worsening edema and epidermolysis and the small fingertip remained dusky without progression to frank necrosis. Despite continuation of occlusive dressings, the areas of epidermolysis progressed to full thickness skin necrosis. Doppler imaging at 14 days of life showed recanalization of the previous subclavian thrombus, and an incompletely occlusive axillary artery thrombus with normal distal flow. Surgical debridement of necrotic tissue from the ulnar aspect of the forearm including part of the flexor carpi...
ulnaris and extensor carpi ulnaris muscles was performed; no signs of infection were noted.

With continued negative pressure wound therapy and therapeutic anticoagulation, the wounds developed healthy granulation tissue and were found to be completely healed at 2 months (Fig. 3), at which time the patient had begun spontaneous movement of the thumb, wrist, and elbow. One month later, Doppler imaging demonstrated complete thrombus resolution and unremarkable hematologic workup. The patient had only partial return of elbow flexion/extension and minimal recovery of hand and wrist motion at 8 months. Contracture along the ulnar fasciotomy site was severe enough to prevent neutral position wrist bracing, necessitating soft tissue release (Figs. 4 and 5).

DISCUSSION

The etiopathogenesis of NCS includes both intrinsic and extrinsic factors composing of in utero mechanical compression, birth trauma, or hypercoagulability, leading to an occlusive thrombus. NCS presents with evolving limb ischemia characterized by ischemic changes in the cutis, ranging from localized ecchymosis to blistering and bullae or large areas of skin necrosis. These changes, commonly mistaken for birth trauma, herald variable degrees of injury to the underlying deep tissues dependent upon the length of tissue injury associated with arterial embolism, trauma, and reflex vessel spasm. Permanent injury to muscles and nerves can result in fibrosis with Volkmann contracture and/or irreversible nerve damage, permanent joint contracture, tissue loss, and functional abnormalities of bone.

Wiseman et al described complete left axillary artery occlusion causing ischemic forearm and hand changes with the absence of brachial, radial, and ulnar pulses and lack of spontaneous wrist and finger movement. Thrombectomy resulted in transient improvement, but

Fig. 1. One day of age; cyanosis and ecchymosis of the right upper extremity. Black line demarcated area of ischemic involvement.

Fig. 2. Postoperative day 4; forearm fasciotomies.

Fig. 3. One month of age; after negative wound pressure therapy.

Fig. 4. Ten months of age; right forearm contracture.
the development of forearm and hand gangrene necessitated transelbow disarticulation. This emphasizes the importance of surgical intervention in neonatal occlusive arterial thrombus causing ischemic changes to avoid limb loss. Ricciardelli et al. reported a patient born with upper extremity skin necrosis, absence of distal pulses, and brachial artery thrombosis on arteriogram, treated with urokinase therapy and forearm soft tissue debridement, ultimately achieving limb salvage. The early diagnosis of NCS and treatment with fasciotomy can prevent the progression of ischemia and injury, increasing the potential for limb salvage and restoration of function.

Given the lack of arterial occlusion on initial ultrasound, the initially fluctuating course here is presumably due to in utero mechanical compression, revascularization after delivery with reperfusion injury, and subsequent occlusive subclavian arterial thrombosis formation, which was detected by ultrasound at 36 hours post delivery. Management with complete upper extremity fasciotomy, antithrombotic therapy, and daily wound care/debridement resulted in restoration of distal perfusion and extremity salvage. Nevertheless, gross sensation in the fingers appears intact and slow progress continues at the time of this manuscript. Given the regenerative capabilities of infants, we are optimistic that functional improvement will continue.

**CONCLUSIONS**

We present a case of right upper extremity NCS caused by complete right subclavian artery thrombotic occlusion resulting in ischemic injury. This case demonstrates that even delayed treatment with systemic antithrombotic therapy, fasciotomy, debridement of affected tissue, and negative pressure therapy can result in limb salvage. Clinicians should be aware of the early characteristic skin findings and promptly diagnose compartment syndrome. Timely and aggressive surgical and medical treatment are crucial to achieve the best overall prognosis.

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