Radial Artery Pseudoaneurysm in a Neonate with Hemophilia A

SHRENIK VORA, THOWFIQUE IBRAHIM AND VICTOR SAMUEL RAJADURAI
From Department of Neonatology, KK Women's and Children's Hospital, 100, Bukit Timah Road, Singapore.

Correspondence to: Dr Shrenik Vora, Staff Registrar, Department of Neonatology, KK Women's and Children's Hospital, 100, Bukit Timah Road, Singapore 229899, dr_shrenikvora@yahoo.co.in
Received: March 24, 2014; Initial review: June 30, 2014; Accepted: September 02, 2014.

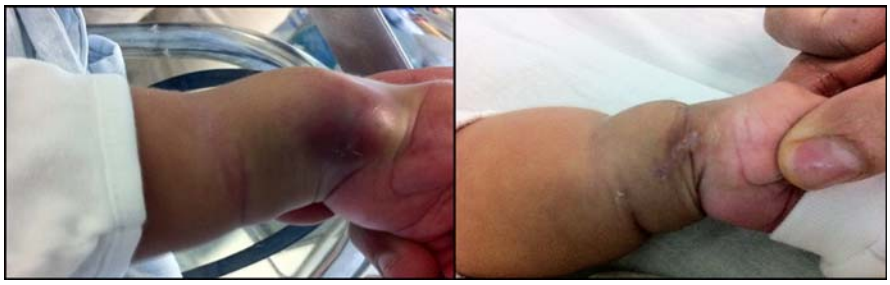
Background: Pseudoaneurysm formation is a rare complication of arterial puncture. **Case characteristics:** 3-week-old male developed an enlarging mass over the anterior aspect of left wrist following radial arterial puncture. **Observation:** Doppler ultrasonography revealed mass to be left radial arterial pseudoaneurysm. Subsequent presentation of ecchymoses and investigations confirmed factor VIII deficiency (Hemophilia A). **Outcome:** Pseudoaneurysm removed with primary end to end anastomosis. Patient is presently on regular factor VIII replacement therapy. **Message:** Hemophilia A can present as pseudoaneurysm in neonatal period.

Keywords: *Arterial cannulation, Complications, Factor VIII.*

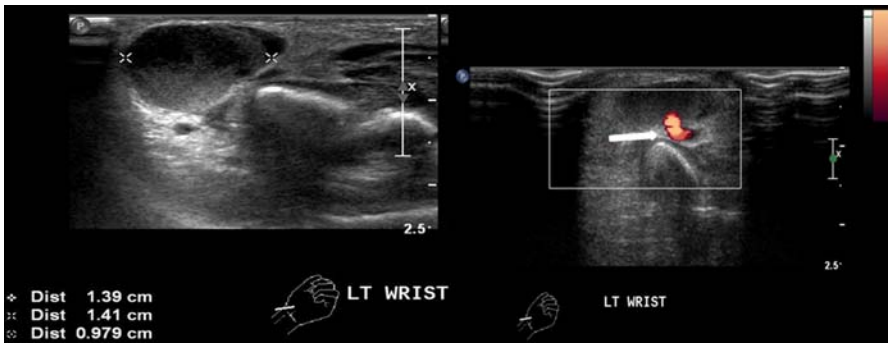
Trauma to the wall of an artery can lead to the formation of pseudoaneurysm, a painful expanding mass overlying the damaged vessel [1]. 30% of severe factor VIII deficiency presents in neonatal period [2]. There are infrequent case reports of hemophilia presenting as pseudoaneurysm in children, more so in neonates [3]. We describe a case report of radial artery pseudoaneurysm following arterial puncture in a newborn infant with hemophilia A.

CASE REPORT

A 3-week-old large-for-gestational age male neonate admitted for presumed sepsis, presented with an enlarging mass at the anterior aspect of left wrist. A week prior to this, an arterial puncture with 24-guage needle had been attempted at the site for blood collection. Physical examination showed a 1×1 cm, firm, non-pulsatile, non-warm swelling over left radial artery, with good capillary refill of distal upper extremity (**Fig.1*a***). Doppler ultrasonography revealed 1.4×1.4×1 cm cystic lesion with moving internal echoes situated adjacent to left radial artery with small connection with the artery suggestive of left radial arterial pseudoaneurysm (***Fig.* 1*b***). The mass progressively increased in size and the patient underwent an operative repair consisting of excision of pseudoaneurysm with primary end to end anastomosis.



(a)



(b)

FIG.1 *(a) Left radial artery pseudoaneurysm- pre- and post- repair; and (b) Sonogram showing cystic lesion with doppler color flow image showing internal echoes (white arrow).*

There were three puncture marks over left radial artery with no evidence of inflammation or infection and there was no difficulty in achieving intra-operative hemostasis. Post-operatively, palpable pulses and good doppler flow were established in the left radial and ulnar artery.

The child subsequently developed spontaneous bruises and ecchymoses at lower limbs. The activated partial thromboplastin time was prolonged (>180 secs) and diagnosis was confirmed as hemophilia A (factor VIII levels < 1%). Factor IX, XI, XII, von-willebrand factor and ristocetin factor levels were in acceptable range. Mother's activated partial thromboplastin time and factor VIII levels were within normal range, and there was no family history of hemophilia. Post-discharge, the child was started on regular factor VIII replacement therapy. On follow-up at one year of age, clinical examination revealed palpable left radial pulses and normal growth of the left upper extremity without any neurological deficit.

DISCUSSION

Arterial trauma may lead to occlusion of the vessel by thrombosis, development of arterio-venous fistula or the formation of an arterial pseudoaneurysm [4]. Pseudoaneurysm formation is a delayed complication arising due to disruption of the vessel wall with containment of blood by surrounding tissues and appearance of sac in direct continuity with arterial lumen. The most common cause of pseudoaneurysm is blunt or penetrating trauma to artery from cannulation or puncture, while less common etiologies are connective tissue disorders, bleeding tendencies, infection and inflammation [5]. Severe factor VIII deficiency mainly presents in neonatal period as excessive hematomas, post-delivery cephalhematomas, post-surgical bleeding and intracranial bleed [2]. Pseudoaneurysm in hemophilia is known, but there has been a limited description of bleeding disorders presenting as arterial pseudoaneurysm in the neonatal period [6].

Most common means of presentation of pseudoaneurysm is that of a palpable pulsatile or non-pulsatile expanding mass with palpable distal pulses and a vital extremity. Pressure applied to the mass will result in decompression of the blood filled sac whereas release leads to rapid refill, thereby distinguishing from a solid mass and it should be carefully examined for a palpable thrill and audible bruit. An effort should always be made to look at other diagnosis like connective tissue disorder, coagulopathy or arteritis [7]. In our case, the appearance of spontaneous bruises and ecchymosis along with the formation of left radial artery pseudoaneurysm lead us to investigate further and confirm the diagnosis of bleeding diathesis. Doppler sonography or CT scan is required to assess the presence of vascular flow within the mass and to differentiate the lesion from either a tumor or abscess [8]. Traditionally arterial pseudoaneurysms are managed in various ways: observation, compression bandages, ultrasound-guided compression, ultrasound-guided thrombin injection, and surgical repair [9]. Factors requiring urgent surgery include bleeding, evidence of vascular compromise or increasing size of the pseudoaneurysm (as in our case). Patients with arterial pseudoaneurysm need to be followed up for a prolonged period to assess possible development of growth disturbances, neurological deficit, and limb length discrepancies [10].

To conclude, radial artery cannulation being a very common procedure in clinical practice for arterial blood gas analysis and cardiovascular monitoring in neonates and infants, physicians and nurses in pediatrics units should be aware of the possibility of incurring arterial injuries, necessitating early diagnosis and prompt intervention of pseudoaneurysms. Though a small-caliber needle was used for radial arterial puncture in our case, his underlying bleeding diathesis contributed to the formation of radial arterial pseudoaneurysm. One should be vigilant to rule out other rarer causes of pseudoaneurysm like bleeding disorders, inflammation and infection.

Acknowledgement: Dr Por YC, for his help in the surgical management of the patient.
Contributors: SV: Literature review, manuscript drafting, review and editing, and patient management; TI: Literature review, manuscript editing and patient management; VSR: Guidance and final editing of manuscript with extensive literature search. All authors approved the final manuscript.
Funding: None; *Competing interests:* None stated.

REFERENCES

1. Ganchi PA, Wilhelmi BJ, Fujita K, Lee WP. Ruptured pseudoaneurysm complicating an infected radial artery catheter: case report and review of the literature. Ann Plast Surg. 2001;46:647-50.
2. Chalmers E, Williams M, Brennand J, Liesner R, Collins P, Richards M. Guideline on the management of hemophilia in the fetus and neonate. Br J Haematol. 2011;154:208-15.
3. Rodriguez-Merchan EC. Pseudoaneurysms in haemophilia. Blood Coagul Fibrinolysis. 2013;24:461-4.
4. Rey C, Manache P, Watel A. Iatrogenic false aneurysm of the brachial artery in an infant. Eur J Pediatr. 1987;146:438-9.
5. Ivo D, Josip U, Davor M, Kresimir B. Pseudoaneurysms of the brachial artery following venipuncture in infants. Pediatr Surg Int. 2004;20:594-7.
6. Fields JM, Saluja S, Schwartz DS, Touloukian RJ, Keller MS. Hemophilia presenting in an infant as a radial artery pseudoaneurysm following arterial puncture. Pediatr Radiol. 1997;27:763-4.

7. Kenneth WG, James M, Ellen LP, Thomas FD. Brachial artery pseudoaneurysm in a 6-week-old infant. Amer Surg. 2004;06:518-21.
8. Restrepo R, Ranson M, Chait PG. Extracranial aneurysms in children: Practical classification and correlative imaging. AJR. 2003;181:867-78.
9. Landau D, Schreiber R, Szendro G, Golcman L. Brachial artery pseudoaneurysm in a premature infant. Arch Dis Child Fetal Neonatal Ed. 2003;88:F152-3.
10. Wolffgang BG, Steven MS, Todd DR. Radial artery pseudoaneurysm in the intensive care unit. Ann Vasc Surg. 2010;24:554e13-16.

Type 0 Spinal Muscular Atrophy with Multisystem Involvement

SANJEEV KHERA AND RANJIT GHULIANI
From Department of Pediatrics, Military Hospital, Agra, Uttar Pradesh.

Correspondence to:
Dr Sanjeev Khera,
Department of Pediatrics, Military Hospital
Agra 282 001, Uttar Pradesh, India.
kherakherakhera@gmail.com
Received: June 03, 2014;
Initial review: June 30, 2014;
Accepted: September 20, 2014.

Background: The classical forms of severe Spinal Muscular Atrophy type is well recognized by pediatricians. **Case Characteristics:** A hypotonic neonate with severe respiratory distress at birth. **Observation:** Homozygous absence of exons 7 of the Survival Motor Neuron I gene. **Outcome:** Died 108 days after admission when respiratory support was withdrawn at the request of the parents. **Message:** Spinal Muscular Atrophy should be kept in mind in the differential diagnosis for unexplained severe generalized hypotonia and severe respiratory distress immediately after birth in the neonates.

Keywords: *Contracture, Exons 7 Survival Motor Neuron, Hypotonia, Neonate.*

The classical form of severe Spinal Muscular Atrophy (SMA) type 1 (Werdnig-Hoffmann disease) has a very consistent clinical phenotype that is well recognized by pediatricians. We report a case of Type 0 SMA. The other notable features in this case report are a rare multi-systemic presentation of SMA associated with intractable seizures not due to hypoxic ischemic encephalopathy (HIE), asymptomatic congenital heart disease, congenital contracture, spontaneous long bone fracture and osteopenia.

CASE REPORT

A male neonate was born to a primigravida mother from a non consanguineous marriage by elective cesarean section at 39 wk of gestation. The pregnancy was unremarkable (except for borderline polyhydramnios) and history of reduced fetal movements reduced for one week before delivery. The neonate did not make any respiratory efforts after delivery and had a weak cry requiring intubation and mechanical ventilation at birth, however bradycardia was never noticed. He was ventilator-dependent, hypotonic, alert and responsive to tactile stimuli with absent deep tendon jerks. Extraocular and facial muscles were spared. He had a bell shaped chest and tongue fasciculation. He was noticed to have contractures of wrist and knee joint immediately after birth (***Figs.* 1, 2**). No other dysmorphism or obvious congenital anomalies were noted.

Serum creatine kinase was 110 IU/L. He required sustained mechanical ventilation. Subtle seizures were noticed on day 4 of life, which required multiple antiepileptic drugs and were refractory. The electrolytes and blood sugar levels were normal on multiple occasions. The seizures were not fully controlled with Injectible Phenobarbitone, Sodium Phenytoin, and high dose Inj Midazolam infusion initially. Later on he was started on oral Sodium Valproate and Levetiracetam without sustained seizures control. Transcranial Ultrasonography was unremarkable. Since the baby was

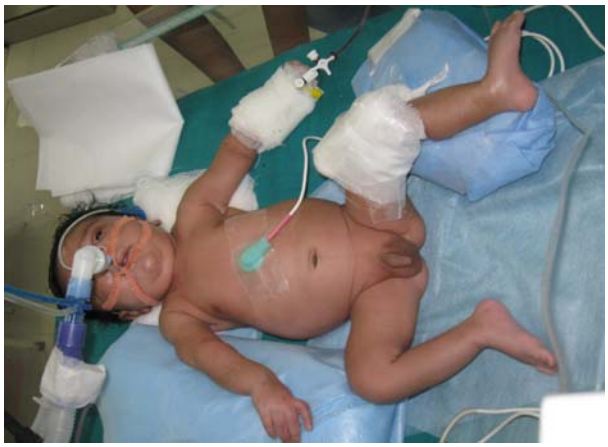


FIG. 1 *Infant on ventilator with alert expression, pithed frog posture, splinted fracture left femur, bell shaped chest and contracture of wrist and knee joint.*