

features were also favoring clinical diagnosis, but there was another lesion in the acetabulum, which was not a typical feature of bone sarcomas. There was no other bone involvement or systemic involvement characteristic of LCH. LCH most commonly involves flat bones. Potepan, et al. [7] reported a series of seven children of LCH, where initial X-ray findings were suggestive of malignancy (large lytic lesion, purely destructive in nature). Lesion sites were pelvis, tibia, femur, clavicle, jaw and scapula. All children were treated with chemotherapy for LCH or curettage and long-term follow-up showed complete remission. Apart from this series, there are rare reports of LCH mimicking osteomyelitis [8].

We reported these two cases to highlight the difficulties in radiological diagnosis of bone lesions in children and the manner in which they can behave like radiological mimics. A limitation of our report is that for the child in case 1, we could not process additional IHC for Ewing sarcoma and ALCL. Radiologically, bone lesions in children can sometimes be confusing and histopathology will help us to reach a final diagnosis.

MAHARSHI TRIVEDI,¹ GURUPRASAD CS,^{1*}

PRIYAKUMARI THANKAMONY,¹

SUBIN SUGATH,² JUBIE RAJ,³ JAYASREE KATTOOR⁴
Departments of ¹Pediatric Oncology, ²Surgical Oncology,
³Imaging, and ⁴Pathology, Regional Cancer Center,

Thiruvananthapuram, Kerala.
* drguruprasadcs@gmail.com

Note: Additional material related to this study is available with the online version at www.indianpediatrics.net

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Spontaneous Neonatal Arterial Thrombosis of Axillary Artery

Neonatal arterial thromboembolism is uncommon [1], and most commonly it is related to arterial catheterization or another iatrogenic injury. Currently only few case reports describe spontaneous arterial thrombosis, manifesting immediately after birth, failure to identify can have serious consequences [2-4]. We describe a neonate with spontaneous upper limb artery thrombosis manifesting immediately after birth leading to massive limb ischemia.

A male infant of a diabetic mother was born at 36 week gestation by cesarean section due to cephalopelvic disproportion (birthweight 3650 g), with an Apgar score of 10/10/10. At birth, the physical examination detected cyanosis of the right upper limb from the middle of the shoulder to the acral parts. The upper limb was livid and paretic. The formation of bullae in the forearm was observed after a few hours of life.

After transport and admission of the patient to the neonatal intensive care unit (NICU), Doppler ultrasound and computed tomography angiography (CTA) was urgently performed. Scans revealed a complete thrombotic occlusion in the distal 3 cm of axillary artery (Fig. 1). Due to the age of the child and the location, interventional catheter-directed thrombolysis (CDT)

could not be technically performed. Systemic thrombolysis was contraindicated because of imminent surgery. Therefore, anticoagulant therapy was started. A bolus of unfractionated heparin (UFH) was administered, followed by continuous heparinization with concomitant antithrombin III (AT III) substitution. Conservative treatment had no effect on the extent of ischemia, so surgical thrombectomy with a Fogarty catheter was done on the first day of life. Despite attempts at revascularization and continuous hematological treatment, irreversible changes of right forearm (mummified fingers, extensive necrosis of the forearm and distal part of the shoulder) persisted.

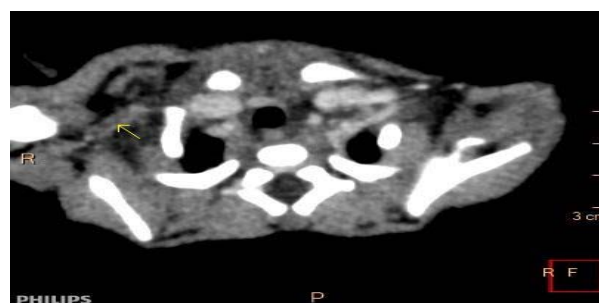


Fig. 1 Computer tomograph angiography scan of complete thrombotic occlusion in the distal 3 cm of axillary artery (white arrow).

On the sixth day of life, the amputation of the devitalized part of arm was done with negative pressure wound therapy. Concomitantly, hematological therapy continued and was later changed to low molecular weight heparin (LMWH). A diagnosis of prothrombotic thrombocytopenic platelet aggregation (sticky platelet syndrome type 1, SPS I) was confirmed, with exclusion of other thrombophilic condition such as AT deficiency, factor II mutation G20210A, factor V Leiden, *MTHFR* gene mutation, and homocysteine deficiency. Growth and development of child at 12 months is age-appropriate.

The actual incidence of spontaneous arterial thromboembolism in newborn is unknown and varies depending on the method used for assessment. It is assumed that arterial thromboses represent 50% from all thrombotic events in neonatal period. Identified risk factors can be categorized into four subgroups maternal (e.g., gestational diabetes), congenital (e.g., placental pathology), acquired (e.g., sepsis) and inherited prothrombotic abnormalities [3,7]. Prematurity, maternal gestational diabetes and presence of SPS I associated with platelet hyper-aggregability were identified as predisposing factors. Although SPS usually occurs in the third and fourth decade of life, cases in childhood are also reported [8].

Management of spontaneous arterial thrombosis can be challenging. Therapeutic approaches and drug dosage are not standardized. The goal of treatment is interrupting thrombus propagation, enhancing tissue perfusion with complete recovery of the affected area, while minimizing risk of bleeding [6]. The most commonly used therapeutic modalities in the literature are heparin, LMWH, tissue plasminogen activator (tPA), urokinase and streptokinase. Microsurgical techniques appear to improve outcome and should be reserved for severe cases [3,5]. Metabolism of the discussed medications is different in the neonate. Heparin is used in a higher dose compared to adults, but there is still a lack of consensus on the ideal dose. The use of thrombolytic agents is promising but has never been proven in randomized controlled trials [3,7]. Interventional radiological management of acute ischemia can be difficult, although catheter-directed thrombolysis has been reported to be beneficial in selected patients [5].

The most important lesson from this case is the need of prompt recognition of symptomatic thromboembolism with urgent treatment. Despite the lack of data on effectiveness of specific treatments, we feel that an early aggressive approach to treatment can improve outcome.

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**FILIP SCHRENK,¹ ZUZANA UHRIKOVA,^{1*}
KATARINA MATASOVA,¹ DALIBOR MURGAŠ,²
MIRKO ZIBOLEN¹**

From ¹Department of Neonatology,

²Clinic of Pediatric Surgery University Hospital in Martin,

Comenius University Bratislava,

Jessenius Faculty of Medicine,

Martin, Slovakia.

**uhrikovazu@gmail.com*

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