REFERENCES

1. Klimek PM, Lutz T, Stranzinger E, et al. Handlebar injuries in children. Pediatr Surg Int. 2013;29:269-73.
2. Nataraja RM, Palmer CS, Arul GS, et al. The full spectrum of handlebar injuries in children: A decade of experience. Injury. 2014;45:684-9.
3. Je B, Kim HK, Horn PS. Abdominal Compartment Syndrome in Children: Clinical and Imaging Features. Am. J. Roentgenol. 2019;3: 655-64.
4. Fichadia U, Sethuraman U. An unusual case of handlebar injury. Int J Case Rep Images. 2014;5:423-6.
5. Ejike JC, Mathur M. Abdominal decompression in children. Crit Care Res Pract. 2012;2012:180797.

Langerhans Cell Histiocytosis and Osteosarcoma in Children: A Radiological Mimic

Osteosarcoma presents in adolescent age with radiologically aggressive bone tumor, while Langerhans cell histiocytosis (LCH) commonly presents with radiological features of a less aggressive bone lesion [1,2]. We report here two cases of osteosarcoma and LCH with an unusual radiological presentation.

***Case* 1**: A 30-month-old boy presented with swelling over proximal left leg and limping of one-month duration without history of trauma. A 2×2 cm hard swelling, fixed to bone, with mild tenderness was present without any systemic abnormalities. *X*-ray showed a well-defined expansile lytic lesion with narrow zone of transition in meta-diaphyseal region without cortical breaks or significant periosteal reaction (**Web Fig. 1A**). Magnetic resonance imaging (MRI) showed multiseptated peripherally enhancing eccentric lesion without significant periosteal reaction, soft tissue component (**Web Fig. 1B** and **C**). Age and radiological findings were suggestive of LCH. Biopsy showed malignant spindle cells with osteoid formation, typical of osteoblastic osteosarcoma. Immunohistochemistry (IHC) for analplastic large cell lymphoma (ALCL) (CD30) and LCH (Langerin) were negative and CD99 was cytoplasmic positive but without crisp membrane positivity, which ruled out Ewing sarcoma. Thus, diagnosis of osteosarcoma was confirmed. Metastatic work-up was negative. He received six cycles (29 weeks) of methotrexate, adriamycin, cisplatin (MAP) chemotherapy. After 10 weeks (2 cycles) of chemotherapy, MRI post contrast T1 image showed interval regression of adjacent marrow changes and periosteal reaction with reduction in thickness and enhancement of internal septations suggesting treatment response (**Web Fig. 2A and B**). As the child was only 30-months-old, wide-excision and endoprosthetic implant was not a feasible option. Parents were unwilling for above-knee amputation. Hence, we proceeded with wide excision, extracorporeal radiotherapy, and internal fixation. At end-of-treatment, disease was in complete remission (**Web Fig. 2C**). Presently, the child is on follow-up without any evidence of disease, 3-year post-treatment.

***Case* 2**: An 11-year-old girl presented with right thigh pain and limping of one-and-half-month duration without history of trauma. Diffuse swelling and tenderness were present with normal overlying skin at the proximal left thigh without any systemic abnormalities. Blood investigations were normal. *X*-ray showed an irregular lytic lesion involving the upper meta-diaphyseal region with periosteal reaction and cortical-break (**Web Fig. 1D**). MRI showed expansile heterogeneously enhancing lesion involving femoral neck with cortical-break and soft tissue component, which was suggestive of an aggressive bone tumor (osteosarcoma or Ewing sarcoma) (**Web Fig.1E** and **F**). Another lytic lesion was noted in right posterior acetabulum. Biopsy showed sheets of cells having moderate cytoplasm with convoluted vesicular nuclei admixed with eosinophils, diagnostic of LCH. There was no evidence of small round blue cells (for Ewings sarcoma or non-Hodgkin lymphoma) or malignant cells with oseoid formation (for osteosarcoma). IHC for CD1a was diffusely strongly positive. Skeletal survey did not reveal any other bone involvement. She was diagnosed to have multifocal bone LCH without risk-organ involvement. She was started on LCH III protocol. Re-evaluation after 12 weeks induction with vinblastin and prednisolone showed residual lytic area, area of mineralization with regression of periosteal reaction and cortical break (**Web Fig. 2D**). End-of-treatment skeletal survey showed areas of remineralization with cortical thickening without any periosteal reaction, soft tissue component or cortical break (**Web Fig. 2E**). Currently, at 18-month post-treatment, she has no evidence of disease, and can walk without support.

Osteosarcoma presents radiologically with features of aggressive bone lesions like periosteal reaction, periosteal elevation, osteoid formation in soft tissue, wide-zone of transition, cortical break, and pathological fracture [3]. Only 2% of patients present before 5 years of age [1]. In LCH, early lesions appear lytic, expansile, with irregular margin, cortical thickening, and smooth periosteal reaction. As lesions become chronic, they may resolve or appear as punched-out well-defined lesion with sclerotic margins [2]. LCH usually presents at a median age of 3 years and multisystem involvement is more common [4].

Rarely osteosarcoma can radiologically mimic a variety of conditions [5]. In a series of 52 patients (adults and children), six cases of high grade osteosarcoma diagnosed by cytology had atypical radiological appearance and one case of radiological osteosarcoma had histopathological diagnosis of soft tissue sarcoma [5]. In a series by Sundaram, et al. [6] two children with well demarcated osteolytic lesions in tibia diagnosed radiologically as aneurysmal bone cyst, later on were diagnosed as osteosarcoma on open biopsy.

In case 2, child presented at an older age, which is a common age of presentation for aggressive bone tumors. Radiological

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 KATTOOR[4]
Departments of [1]Pediatric Oncology, [2]Surgical Oncology, [3]Imageology, and [4]Pathology, Regional Cancer Center, Thiruvananthpuram, Kerala.
** drguruprasadcs@gmail.com*

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

REFERENCES

1. Ottaviani G, Jaffe N. The Epidemiology of Osteosarcoma. Springer; 2009. p. 3-13.
2. Azouz EM, Saigal G, Rodriguez MM, et al. Langerhans cell histiocytosis: pathology, imaging and treatment of skeletal involvement. Pediatr Radiol. 2005;35:103-15.
3. Fox M, Trotta B. Osteosarcoma: Review of the various types with emphasis on recent advancements in imaging. Semin Musculoskelet Radiol. 2013;17:123-36.
4. Stålemark H, Laurencikas E, Karis J, et al. Incidence of langerhans cell histiocytosis in children: a population-based study. Pediatr Blood Cancer. 2008;51:76-81.
5. Söderlund V, Skoog L, Unni KK, et al. Diagnosis of high-grade osteosarcoma by radiology and cytology: A retrospective study of 52 cases. Sarcoma. 2004;8:31-6.
6. Sundaram M, Totty WG, Kyriakos M, et al. Imaging findings in pseudocystic osteosarcoma. Am J Roentgenol. 2001;176:783-8.
7. Potepan P, Tesoro-Tess JD, Laffranchi A, et al. Langerhans cell histiocytosis mimicking malignancy: A radiological appraisal. Tumori. 1996;82:603-9.
8. Chang W-F, Hsu Y-C, Wu Y-D, et al. Localized Langerhans cell histiocytosis masquerading as Brodie's abscess in a 2-year-old child: A case report. EXCLI Journal. 2016;15:33.

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 administrated, 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.

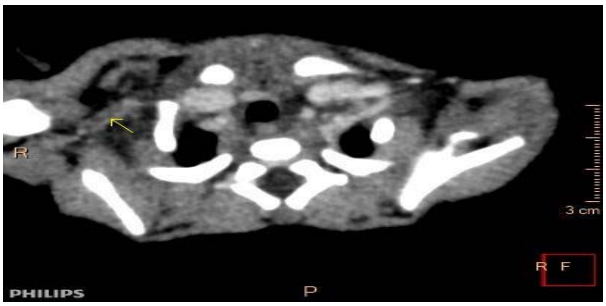
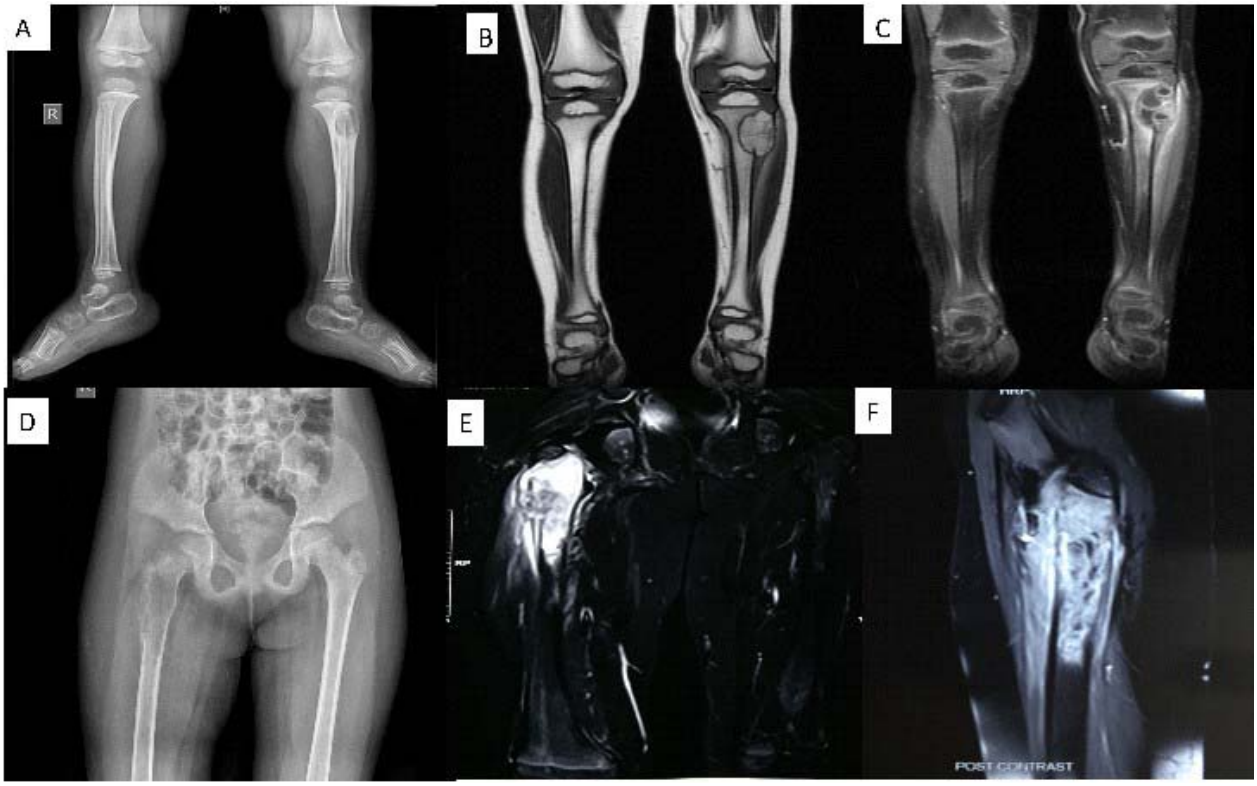
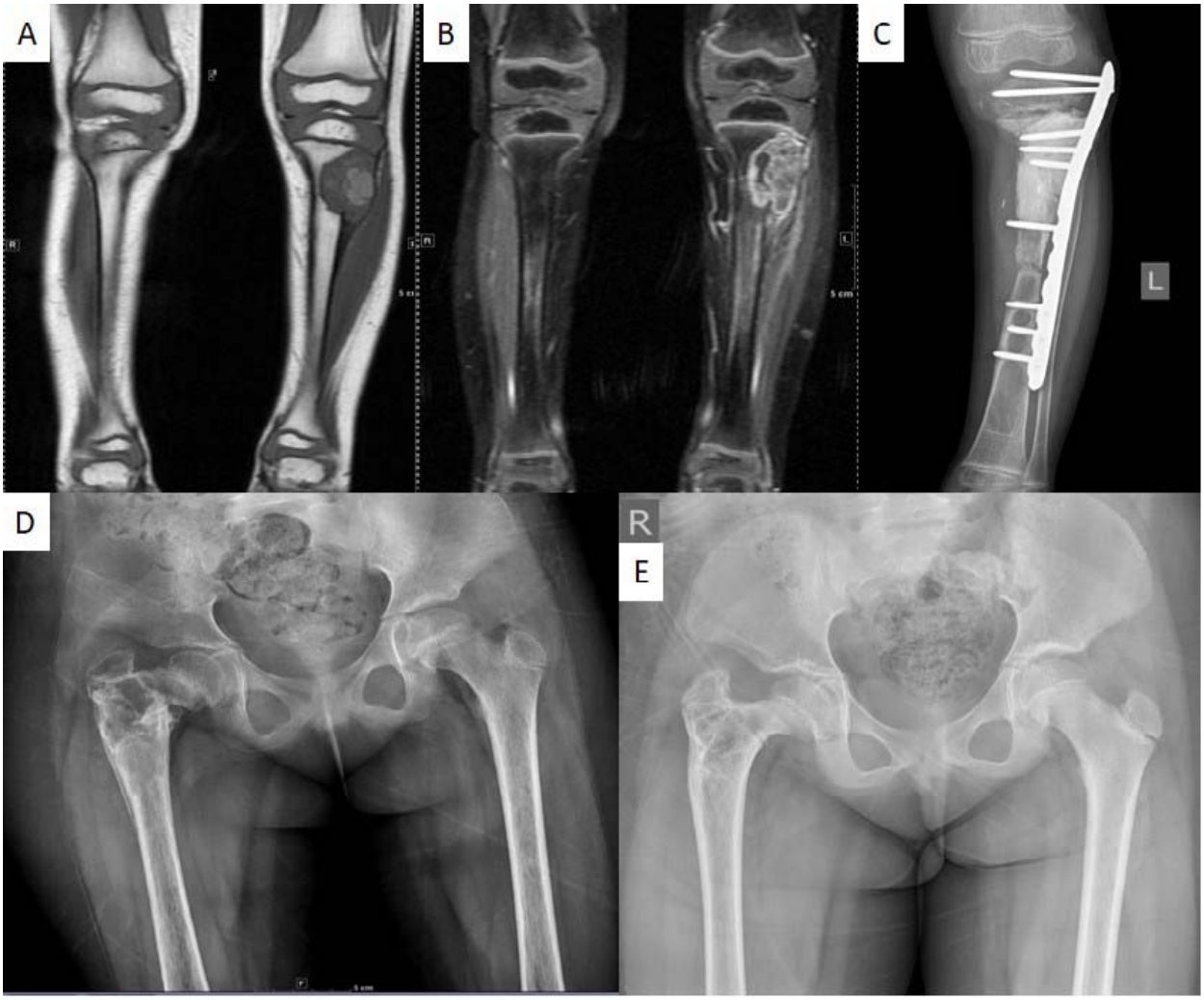


Fig. 1 Computer tomograph angiography scan of complete thrombotic occlusion in the distal 3 cm of axillary artery (white arrow).



Web Fig. 1 (**A)** X-ray of case 1 showing well defined eccentrically located lytic expansile lesion involving proximal metadiaphyseal region of left tibia with a narrow zone of transition without periosteal reaction **(B)** T2 coronal section MRI of left tibia showing well corticated hyperintense eccentrically located mildly expansile lesion with fluid-fluid levels without periosteal reaction or cortical break **(C)** post-contrast coronal section MRI of left tibia showing heterogenous enhancing lesion with predominantly peripheral enhancement and enhancement of internal septa without any obvious solid enhancing area **(D)** X-ray of case 2 showing poorly defined lytic bone lesion with the significant periosteal reaction 'Codman's triangle' and cortical break in the proximal femur. (**E)** Coronal STIR image of the right femur showing hyperintense heterogeneous lesion with soft tissue hyperintensity(**F)** Post-contrast MRI of the right femur showing heterogeneously enhancing lesion with marrow involvement, cortical break, and soft tissue component.



Web Fig.2 (A) Case 1 MRI T1 coronal section of left tibia showing residual well corticated eccentric lesion with interval appearance of irregular T1 hyperintense area suggestive of intratumoral bleed **(B)** Case 1 Post-contrast MRI coronal section of left tibia showing interval regression of adjacent marrow changes and periosteal reaction with reduction in thickness and enhancement of internal septations**(C)** Case 1 post treatment X ray AP view knee joint post extracorporeal radiotherapy and reimplantation with internal fixation **(D and E)** Case 2 – serial X rays (post week 12 chemotherapy and end-of-treatment X ray) showing progressive mineralization of bone with regression of cortical destruction, periosteal reaction and soft tissue component.