

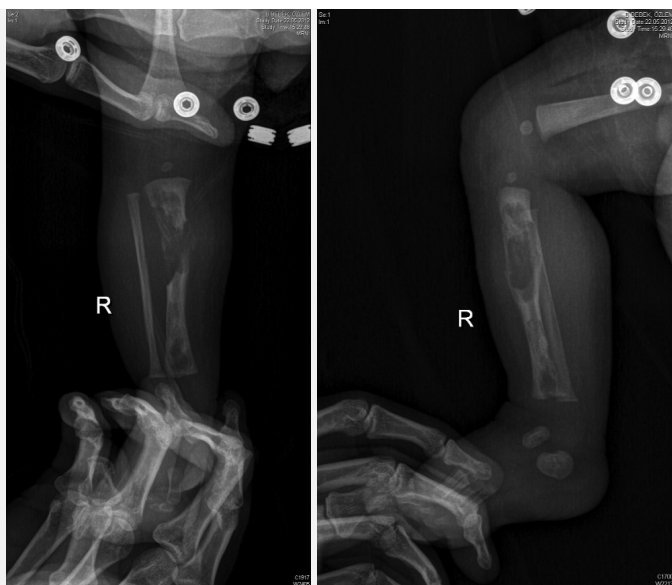


Newborn with pathologic fracture of the tibia: osteofibrous dysplasia

To the Editor,

Osteofibrous dysplasia is a very rare tumor-like developmental fibroosseous condition of the long bones (1). As far as we know a total of eighth newborn cases with osteofibrous dysplasia have been reported in the literature three of which had pathologic fracture (2,3,4,5,6). In this article, a patient with osteofibrous dysplasia who was found to have pathologic fracture at the second hour after birth and who was diagnosed at the earliest period according to the literature.

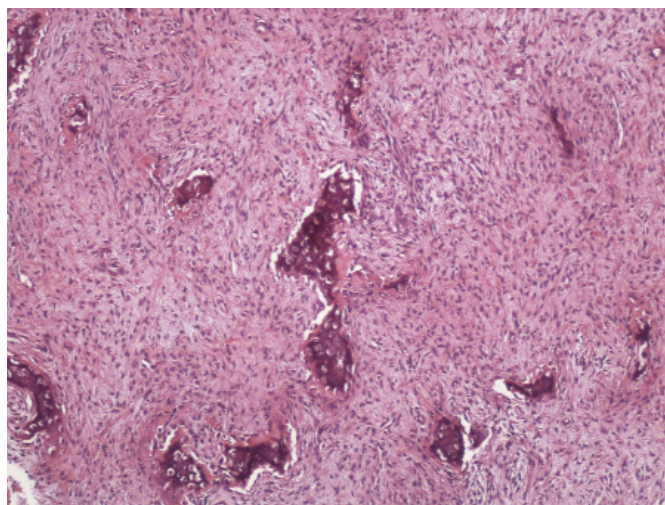
There was no pathology in the medical history of the male infant who was born by cesarean section at the 37 5/7th gestational week because of footling breech presentation. The



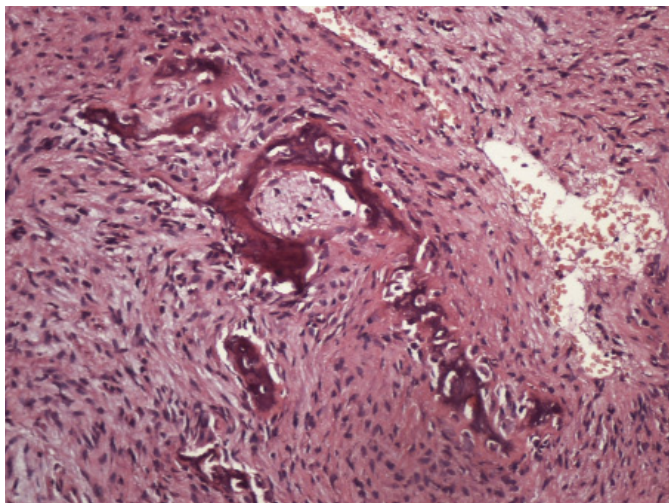
Picture 1. Right tibial anterior and lateral graphies reveal multiple radiolucent areas with lobulated contours, loss of cortical integrity at the 1/3 proximal-middle diaphysis level and angling interpreted in favour of pathologic fracture. The cortical integrity is maintained in the right tibia included in the imaging area

birth weight was 3305 g, the height was 52 cm and the head circumference was 35 cm. When swelling, echymosis, pathologic movement, crepitation and tenderness in the right leg were found two hours after birth in the second examination, two-way tibia-fibula graphy was taken (Picture 1). When pathologic fracture was found on direct graphy, bone biopsy was obtained at the 24th hour after birth and sent to pathology. A fibro-osseous lesion containing ovoid-fusiform cells and new bone formation with a trabecular structure were observed in the biopsy specimen (Picture 2a-2b). A marked osteoblastic array (rimming) was observed in the midst of the newly forming bone trabecules (Picture 2b). The laboratory results were found to be within normal limits. Fibrous dysplasia, congenital syphilis, histiocytosis X, congenital pseudoarthrosis (neurofibromatosis) were considered in the differential diagnosis. With the existing clinical, radiologic and histopathological findings a diagnosis of osteofibrous dysplasia was made.

The disease has been mostly found in children below the age of 10 years (1,7). It is a condition of growing bones. It has been found that it occurs a little more frequently in males. It has been reported in advanced ages and rarely in the newborn period. The



Picture 2a. Bone trabeculation, fibroosseous lesion containing ovoid-fusiform fibroblastic cell proliferation (x200 HE)



Picture 2b. Some bone trabeculae are surrounded by osteoblastic cells, while osteoblastic rimming is prominent in some

fact that the disease occurs rarely in advanced ages indicates that the disease regresses spontaneously or is maintained regular with the actualization of bone maturation (1,8,9). It has been reported that genetic effect may be involved in the etiology (10). Generally, the disease is manifested with tibia extension and curling of the tibia anteriorly or anterolaterally with a mild or moderate degree (8). The lesion is generally painless. A part of the cases are determined with pathologic fracture as a result of minor trauma (1,7). Our patient is the earliest diagnosed case in the newborn period and the fourth case with pathologic fracture in the newborn period in the literature. The reason of minor trauma leading to pathologic fracture was probably pulling the foot during cesarean section performed because of footling breech presentation. Recognition of swelling and ecchymosis for the first time at the second hour supports this. Eccentric intracortical osteolysis is the rule in radiologic examination. The outer surface of the cortex can be enlarged moderately or severely or thinned to a high degree because of the tumor which shows extension. The tumor has a bullous appearance which consists of a single cyst composed of confluent lytic areas or multiple cysts. It is typically located in the diaphysis and may extend to the metaphysis. The tumor matrix usually has a ground-glass appearance. If pathologic fracture is present, fracture findings may be observed and if pathologic fracture is not present flexion defect may be observed frequently (4,7,8). Although radiological findings are not pathognomonic, osteofibrosis substantially suggests dysplasia. The periosteum is always protected well histopathologically and the cortex may be as thin as parchment. Microscopic examination reveals immature bone tissue surrounded by osteoblasts in the fibroblastic stroma. This finding is named as “zonal architectural pattern”. The fibrous part contains small and scattered fusiform cells (2,7,8,11).

There is no generally accepted treatment method. Generally, it has been reported that the lesion may regress spontaneously and may be followed up with “conservative” treatment especially in children below the age of 15 years (4,7,8,12). Surgical

treatment is usually recommended in cases of excessive growth and pathologic fracture. Surgical treatment is not recommended in children below the age of one (4,7,8,12). Regional recurrence has been found in a portion of the patients in whom subperiosteal resection was performed (1,8). In one study, extraperiosteal excision was recommended in all subjects with a diagnosis of osteofibrous dysplasia, since adamantinoma was found in recurrent biopsy in the follow-up (11).

The pathologic fracture of our patient was stabilized by pediatric orthopaedics with plaster splint. Surgical excision or curettage was not performed in accordance with the literature information. The patient is still being followed up by pediatric orthopaedics.

Conclusively, osteofibrous dysplasia should be considered in presence of lesions located in the anterior side of the tibial diaphysis and in presence of congenital intracortical osteolytic tibia lesions, though it is a very rare condition.

**Serdar Al¹, Hasan Tarkan İkizoğlu², Metin Uzun³,
Mebrure Bilge Bilgiç⁴, Harzem Özger⁵**

¹Medipol University Medical Faculty, Department of Pediatrics, İstanbul, Turkey

²Acibadem Maslak Hospital, Department of Pediatrics, İstanbul, Turkey

³Acibadem Maslak Hospital, Department of Orthopaedics, İstanbul, Turkey

⁴Istanbul University, Istanbul Medical Faculty, Department of Pathology, İstanbul, Turkey

⁵Istanbul University, Istanbul Medical Faculty, Department of Orthopaedics, İstanbul, Turkey

References

1. Campanacci M. Osteofibrous dysplasia of long bones a new clinical entity. *Ital J Orthop Traumatol* 1976; 2(2): 221-237.
2. Anderson MJ, Townsend DR, Johnston JO, Bohay DR. Osteofibrous dysplasia in the newborn. *J Bone Surg Am* 1993; 75(2): 265-267.
3. Zamzam MM. Congenital osteofibrous dysplasia of the tibia, associated with pseudoarthrosis of the ipsilateral fibula. *Saudi Med J* 2008; 29(10): 1507-1509.
4. Çetinkaya M, Özkan H, Köksal N, Sarısözen B, Yazıcı Z Neonatal osteofibrous dysplasia associated with pathological tibia fracture: a case report and review of the literature. *J Pediatr Orthop B* 2012; 21(2): 183-186.
5. Castellote A, García-Peña P, Lucaya J, Lorenzo J. Osteofibrous dysplasia. A report of two cases. *Skeletal Radiol* 1988; 17(7): 483-488.
6. Smith NM, Byard RW, Foster B, Morris L, Clark B, Bourne AJ. Congenital ossifying fibroma (osteofibrous dysplasia) of the tibia—a case report. *Pediatr Radiol* 1991; 21(6): 449-451.
7. Park YK, Unni KK, McLeod RA, Pritchard DJ. Osteofibrous dysplasia: clinicopathologic study of 80 cases. *Hum Pathol* 1993; 24(12): 1339-1347.
8. Campanacci M, Laus M. Osteofibrous dysplasia of the tibia and fibula. *J Bone Joint Surg Am* 1981; 63(3): 367-375.
9. Goto T, Kojima T, Iijima T, Yokokura S, Kawano H, Yamamoto A, Matsuda K. Osteofibrous dysplasia of the ulna. *J Orthop Sci* 2001; 6(6): 608-611.
10. Hunter AG, Jarvis J. Osteofibrous dysplasia: two affected male sibs and an unrelated girl with bilateral involvement. *Am J Med Genet* 2002; 112(1): 79-85.
11. Lee RS, Weitzel S, Eastwood DM, et al. Osteofibrous dysplasia of the tibia. Is there a need for a radical surgical approach? *J Bone Joint Surg Br* 2006; 88(5): 658-664.
12. Resnick DS. *Diagnosis of bone and joint disorders*. 3rd ed. Philadelphia: WB Saunders, 1995; 1001.