CASE REPORT

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Osteofibrous dysplasia-like adamantinoma in a 3-month-old male infant: a case report

Halil BULDU[1*], Tuncay CENTEL[1], Hale KIRIMLIOĞLU[2], Yalın DİRİK[1]

[1]Memorial Hospital, Department of Orthopedics and Traumatology, İstanbul, Turkey;
[2]Acıbadem University Faculty of Medicine, Department of Medical Pathology, İstanbul, Turkey

We report a case of a very rarely seen osteofibrous dysplasia-like adamantinoma (OFDLA) of the lower leg in a 3-month-old male infant, making it the youngest case in the literature. OFDLA is typically regarded as a benign lesion; however, due to its convertibility into classical adamantinoma, it is recommended to evaluate it as a pre-malignant lesion. After OFDLA diagnosis with biopsy, our case underwent surgical resection and reconstruction with a large allograft. Patient experienced good outcomes and did not experience any local relapse in the 3-year follow-up.

Keywords: Adamantinoma; osteofibrous dysplasia; osteofibrous dysplasia-like adamantinoma.

Adamantinoma, a low-grade malignant primary bone tumour, is a biphasic tumour containing epithelial and osteofibrous components.[1-13] Osteofibrous dysplasia (OFD) is a rare, benign, fibro-osseous lesion that is typically seen in children. Osteofibrous dysplasia-like adamantinoma (OFDLA) is diagnosed by the presence of scattered nests of epithelial cells imbedded in osteofibrous tissue.[1-5,12-14] The radiographic and morphologic features of adamantinoma, OFD and OFDLA aresimilar.[12] Differential diagnosis between OFDLA and OFD can be difficult and can only be assessed through histological evaluation.

Long bone adamantinoma is rare and accounts for approximately 0.4% of primary bone tumors.[1-3,15] It can be observed in all age ranges from infant to elderly.[1,2,4,8-10] However, it is most frequently observed between the ages of 25 and 35 years.[1] The prevalence and incidence of OFDLA is not currently known.

This case study reports on the diagnosis and treatment of OFDLA in a 3-year-old male infant.

Case report

A male infant of age 3 months and 18 days was brought to our outpatient clinic with a history of thickening and reduced movement in the left leg over the previous two weeks. Physical examination revealed a normal general status. However, there was thickening, deformation, and local tenderness over the left tibia. We did not observe any signs of active infection including local warming or any skin change. No other abnormality was detected. His body temperature, blood pressure, pulse and other vital parameters were all within normal ranges. X-ray and magnetic resonance imaging (MRI) revealed a cortical, lobulated lesion approximately 3 cm in length, with an anterior angulation in the tibial diaphysis (Fig. 1). Based on the radiologic

**Current affiliation: İstanbul Surgical Hospital (Nişantaşı Branch), Department of Orthopedics and Traumatology, İstanbul, Turkey*

Correspondence: Halil Buldu, MD. İstanbul Cerrahi Nişantaşı Hastanesi, Ortopedi ve Travmatoloji Bölümü, Nişantaşı, İstanbul, Turkey.

Tel: +90 212 – 291 91 22 e-mail: halilb77@hotmail.com

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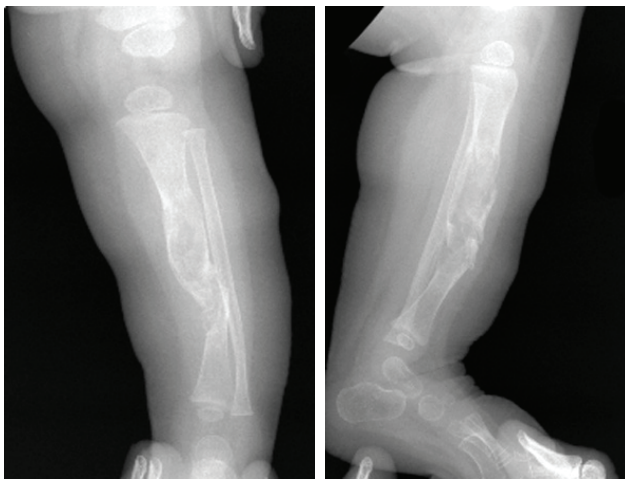

Fig. 1. Pathological fracture angled to anterior and medial side has been seen in cruris anteroposterior and lateral X-ray images.

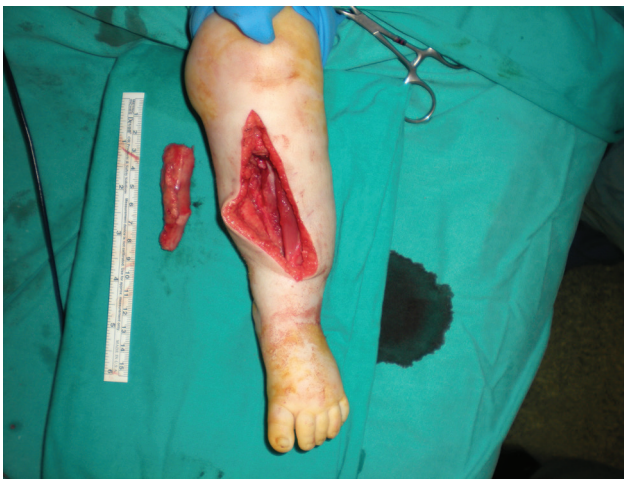

Fig. 3. The 6 cm long resection from tibia. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

examination, we considered differential diagnoses of osteofibrous dysplasia (OFD), nonossifying fibroma and eosinophilic granuloma.

A Tru-cut needle biopsy was performed. Microscopically, the tumour was composed of epitheloid cells within intervening hypocellular osteofibrous tissue. Immunohistochemical examination demonstrated scattered single or thin trabecular patterns of cytokeratin-positive cells. Therefore, the biopsy confirmed a diagnosis of OFDLA according to histological features of the tumour (Fig. 2a, b).

A 6-cm segment was resected from the portion of the tibia with the diagnosis of OFDLA (Fig. 3). After resection, reconstruction using a large allograft was performed.

Histological evaluation of the resected tibia noted similar histologic findings, including microfractures and microcallus areas. The lesion had caused widespread destruction of the cortex, but there was no infiltration outside the cortex.

The patient was followed for 3 years, with no signs of metastasis or local relapse.

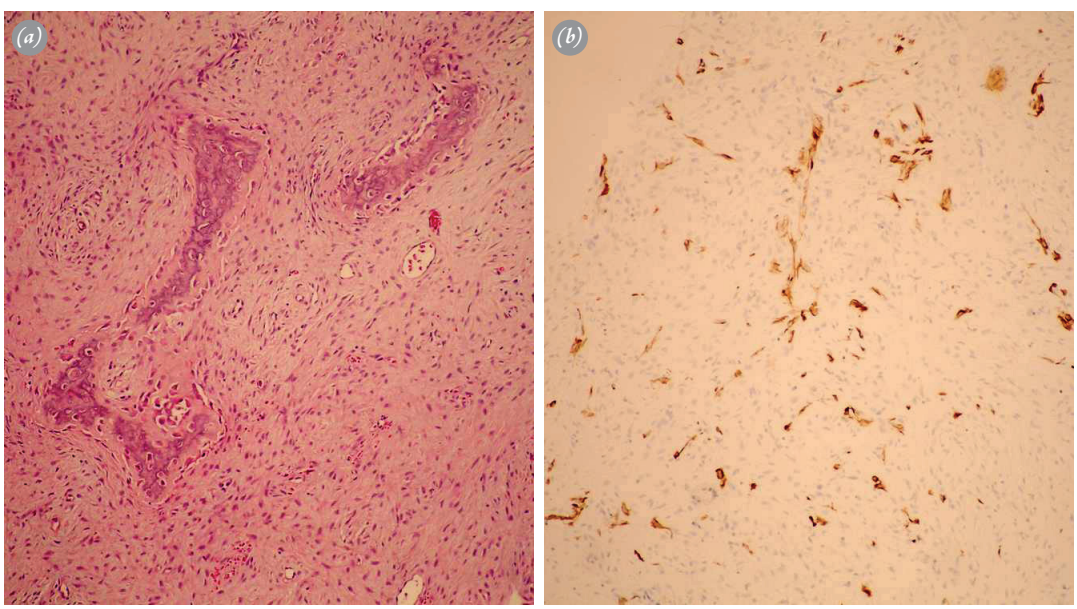

Fig. 2. **(a)** Hypocellular epithelioid shaped cells in the osteofibrous tissues (H&E x100). **(b)** Epithelium cells stained with keratin (x100). [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

Discussion

The present case was unique because OFD, OFDLA and adamantinoma are rare bone tumours. According to Campanacci and Laus, adamantinoma is rarely seen in patients less than 10 years of age.[16] Kuruvilla and Steiner published 5 cases with patients between 4.5 and 14 years of age with OFDLA.[17] To the best our knowledge, this case of a 3.5-month-old male infant is the youngest case with OFDLA published in literature. This is of clinical significance because it highlights the need to screen for these disorders in the infant population.

According to Kuruvilla and Steiner, Sarisozen OFDLA can be a precursor lesion of classic adamantinoma.[7,17] Because adamantinoma has the characteristics of a malignant tumour and can metastasize and exhibit focal and multifocal spread, the patient was assessed similar to a classic adamantinoma case. Similarly, surgical resection was performed, and he was followed up on the basis of classical adamantinoma. This assessment and treatment strategy is based on common conclusions of OFDBA researchers that opined when a lesion is observed, it should be treated as classic adamantinoma.[1-3,7,17]

Contrary to classic adamantinoma, the prognosis of OFDBA is good and generally only follow-up is necessary.[1-3,17] Indeed, no metastasis or local relapse was found in 3 years of follow-up for this case.

Conflicts of Interest: No conflicts declared.

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