



Case Report

Osteofibrous Dysplasia (OFD) may progress to OFD/ Like adamantinoma (OFD/LA)-to-adamantinoma if long-term follow-up is done: A case report

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Abstract

A patient, aged 18 years male complained of painful swelling of lower forearm. X-ray examination showed an expansile osteolytic lesion with internal trabeculations and preserved cortex in diaphysis of ulna. Mild soft tissue swelling of overlying parenchyma was also seen. Histopathological examination showed osteoblastic rimming of bony trabeculae. The lesion was finally diagnosed as osteofibrous dysplasia (OFD) of ulna.

Keywords: Osteoblastic lining, bone trabeculae, fibrous stroma, giant cells, adamantinoma.

Introduction

Osteofibrous dysplasia (OFD) is an expansible diaphysial osteolytic benign lesion. It begins in the cortex and later spreads in the long axis of bone. Tibial involvement may show anterior bowing. Rarely, both tibia and fibula may be involved. Finding of keratin positive cells in fibrous stroma may suggest its relationship with adamantinoma. Later, it was proposed that OFD

may be a regressing adamantinoma^[1]. However, its relationship with adamantinoma could not be confirmed. Herewith, we report a case of osteofibrous dysplasia.

Case Report

A patient, aged 18 years male complained of a painful swelling of lower half of forearm. X-ray examination showed expansion of shaft of ulna

with internal septa and lysis (figure 1). Later, he was operated and several bony pieces, measuring 3×2×2cm were sent to us for histopathological examination. On cutting, gritty sensation was felt. Section showed several thickened sclerotic bony trabeculae with prominent osteoblastic rimming. The intervening fibrous stroma showed few thin-walled blood vessels with areas of hemorrhage. Fibrous stroma showed few macrophages and

small number of lymphocytes. Several multinucleated giant cells were seen. A small piece of cartilaginous tissue was also seen. There was no evidence of a malignant change. The lesion was provisionally diagnosed as osteofibrous dysplasia. Further, immunohistochemical (IHC) examination using Anti-cytokeratin antibody could detect small number of keratin positive epithelial cells in osteofibrous stroma (figure 2).



Figure 1: AP and lateral views of ulna showing expansion of middle of its shaft with osteolytic changes and internal septa formation. Lesion is shown by arrows.

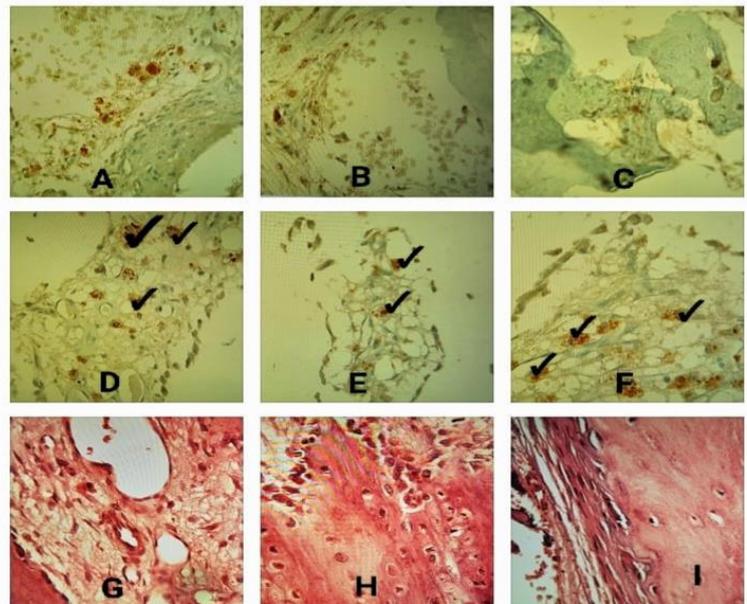


Figure 2: Tumor cells were stained using anti-cytokeratin antibody by IHC. (A) showed an arteriole and positive cells. (B) showed several keratin positive cells. (C) showed woven bone formation. (D) showed several keratin positive cells. (E) showed a layer of osteoblasts and few keratin positive cells. (F) showed approximately 10 keratin positive cells and rimmed layer of osteoblasts. (G) showed fibrous tissue, blood vessels and small portion of trabeculae (H&E×100). (H) showed trabeculae rimmed with osteoblasts. (I) showed trabeculae and osteoid formation. Keratin positive cells have been highlighted by a pointer (H&E×100).

Discussion

Osteofibrous dysplasia (OFD) is a benign lesion which may involve ulna^[2,3] and/or tibia or fibula^[4]. Disease mainly develops in pediatric age-group. Prognosis of the lesion is good. However, Scholfield et al found no evidence of its progression from OFD to adamantinoma^[4]. Therefore, previous authors had advised observation^[5]. However, aggressive behavior of the lesion has been reported in an earlier report^[2]. The disease mainly presents as a pathological

fracture. Another complication might be the existence of adamantinoma in OFD, suggesting a more aggressive behavior^[2]. Radical excision of the lesion might be advised if the disease appeared aggressive. Hahni et al suggested OFD to be a neoplasm while others reported it to be a residue of regressing OFD^[6]. Rarely, OFD/like adamantinoma may develop in fibula with pathological fracture^[7]. Bethopudi et al suggested that OFD/ Like adamantinoma (OFD/LA) and classical adamantinoma might be related. OFD

might be at the benign end of the spectrum and OFD/LA may lie at mid-point of spectrum. In addition, adamantinoma may lie at the malignant end of the spectrum^[8]. It appeared that OFD/LA might have a malignant potential.

Conclusion (S)

A young male had a painful swelling lower forearm. X-ray examination showed a osteolytic lesion in middle ulna. Histopathological examination showed bony trabeculae in fibrous stromal tissue. Osteoblastic rimming of trabeculae was seen. Several diffusely distributed keratin positive cells were detected. There was no evidence of a malignant change. Long-term observation/follow-up of the case was advised for a dysplastic or malignant change to occur.

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