Case Report

Metastatic or metachronous adamantinoma: An Enigma

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Abstract

Adamantinoma is an uncommon tumour of low grade malignancy in terms of biologic aggressiveness; metastasis occurring many years after amputation. A young male underwent an above knee amputation of left leg for adamantinoma of tibia. He returned with pain and swelling in his right thigh five years later. Investigations revealed an adamantinoma in the diaphysis of the right femur. The unusual presentation of this adamantinoma at an uncommon site threw up interesting insights into the nature of this lesion. In particular, the metachronous origin of the adamantinoma in the femur is discussed in this report.

Key words: Adamantinoma, metastases, metachronous

Introduction

The bone tumour commonly referred to as adamantinoma of long bones remains controversial. It is a low grade malignant tumour of mixed epithelial and mesenchymal nature which develops in the diaphyses and metaphyses of the long bones, predominantly of the tibia. The histomorphological appearance of these lesions is typical but varies between cases, making their embryological origin uncertain.[1,2] Numerous papers have been published addressing the radiological and histological spectrum of adamantinoma.[2-4] This locally aggressive neoplasm is also known to metastasize.[5] Although reports have documented the occurrence of adamantinoma in uncommon sites, none have addressed the occurrence of two adamantinomas occurring in the same patient at different sites and different time interval with discordant morphological appearances. An attempt has been made to highlight the concept of a metachronous adamantinoma in this case report.

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Case Report

A 21 year old male, presented with pain and swelling of the right thigh of six months duration. A definitive treatment of above knee amputation of left leg for adamantinoma of tibia had been performed five years back [Figure 1]. Whole body scan was done before amputation and no other lesions were detected anywhere else in the body. X rays showed presently a multilocular, osteolytic, expansile lesion in diaphysis of right femur with cortical thinning. There was no evidence of cortical breach or a pathological fracture [Figure 2]. A radiological diagnosis of aneurysmal bone cyst was rendered. There was no regional lymphadenopathy. Fine needle aspiration of the swelling was inconclusive. Hence a diagnostic biopsy was performed by curetting the osteolytic lesion. Microscopically typical epithelial islands of basaloid cells with peripheral palisading, separated by fibrous septa were seen [Figure 3]. The relative amounts varied from predominantly epithelial islands with little intervening stroma to predominantly fibrous with small islands of cells. Unusual, rarely described feature was presence of multinucleate osteoclastic giant cells abutting the islands and in the septae [Figure 4]. On review, they were not seen in the initial tibial lesion [Figure 5].

Discussion

Adamantinoma is an extremely rare bone tumour constituting <1% of all malignant tumours of bone. Males in the third decade are commonly affected. The tibial diaphysis leads the list as the most common site of occurrence followed by fibula,

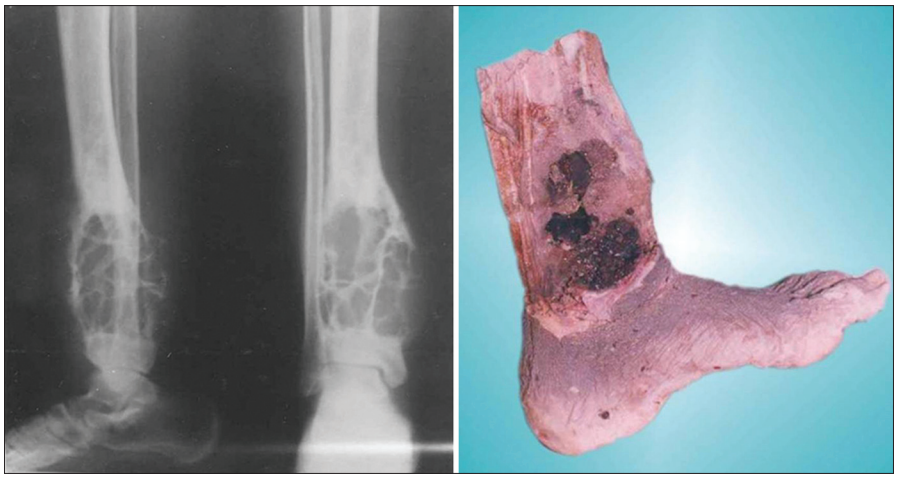

Figure 1: Classical radiological appearance of left tibial adamantinoma (left) along with the amputated left lower limb showing the tumor (right)

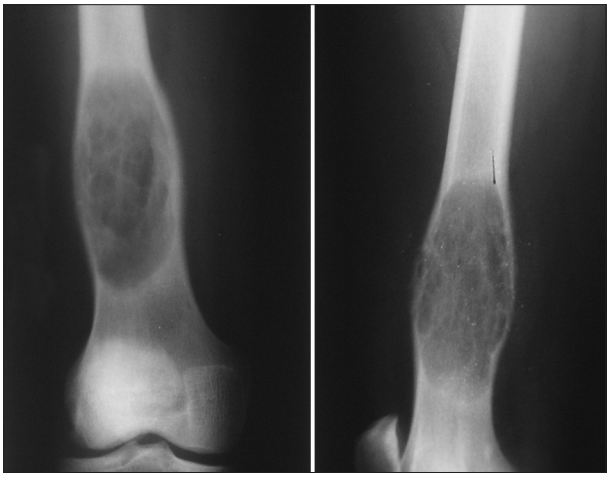

Figure 2: X-ray showing a multilocular, osteolytic, expansile lesion in diaphysis of right femur with cortical thinning

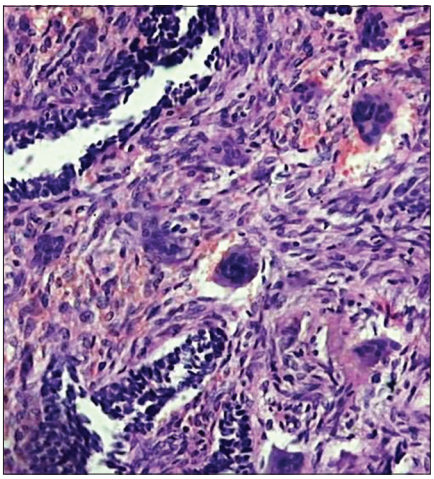

Figure 4: Multinucleate osteoclastic giant cells abutting the islands and in the septae (H and E ×10)

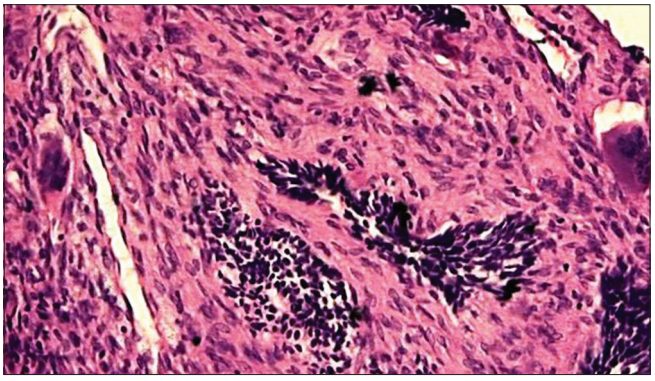

Figure 3: Epithelial islands of basaloid cells with peripheral palisading separated by fibrous septa (H and E ×10)

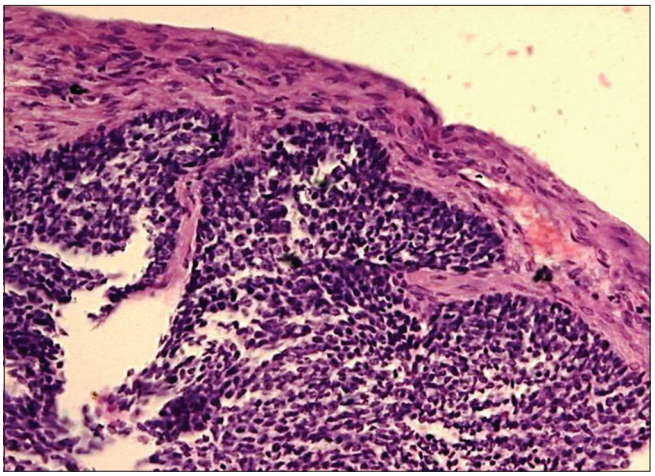

Figure 5: Islands of basaloid cells with peripheral palisading along with fibrous component and absence of giant cells (H and E×10)

femur, ulna and radius.[1,3] Exotic reports of adamantinomas arising exclusively from the pretibial soft tissues without any bony involvement are also documented in literature.[6]

Non-specific symptoms like swelling with or without pain may be the only complaint in many patients initially.[1]

The pathogenesis of adamantinoma has engendered considerable interest. Much of the controversy has centered on the nature of the cell nests in these tumours. While

immunohistochemistry has convincingly put an end to this by demonstrating the epithelial nature of these cells, the basic question of how an epithelial neoplasm developed within a bone is yet to be answered.[3,7]

Plain radiographs are often helpful in diagnosis because of the tumour's classic location and appearance. An expansile, eccentric mass with variable amount of periosteal reaction and cortical thinning, especially involving the diaphyseal region of the tibia is the most common radiological finding. Medullary and soft tissue involvement is also known.[3] Attempts to diagnose the lesion on cytology can be difficult but can be fruitful if the clinico-pathological presentations of the lesion are known well.[8]

The morphology of a classic adamantinoma is an admixture of epithelial and osteofibrous component in various proportions. Four main differentiation patterns have been described for a classic adamantinoma viz., basaloid, tubular, spindle cell and squamous. Though the first two patterns are commonly encountered, an admixture of all these patterns may also be seen. There are reports suggesting a more aggressive behaviour for the basaloid and spindled variants. The spindle cell component is more often observed in recurrences and in metastases.[1,3,6] It is interesting to note that our case had both the basaloid and spindle pattern in both the lesions. The osteofibrous component is composed of storiform oriented spindle cells. Woven bone trabeculae prominently rimmed by osteoblasts are usually present.

A fifth histological pattern, the so called osteofibrous dysplasia like variant, in which the osteofibrous tissue dominates with the epithelial cells being limited to small clusters, is also described.[1,4] Several studies have proposed the concept that osteofibrous dysplasia and adamantinoma form a continuum of pathological processes where osteofibrous dysplasia like adamantinoma is the intermediate step.[7,9]

Multinucleate osteoclastic giant cells, like the one seen in the present case, foam cells and mast cells are rarely detected in an adamantinoma.[1] Adamantinomas are known for their recurrences and metastases to sites like lymph nodes, lungs, skeleton, liver and brain. Recurrences are common after a non-radical surgery. Besides male gender, pain at presentation, short duration of symptoms, age less than 20 years and lack of squamous differentiation of the tumour have been associated with increased rates of recurrence or metastasis.[1] The differentiated or the osteofibrous dysplasia like adamantinoma usually has no metastatic potential. But it is important to note that histologic subtypes have not correlated with biologic behaviour and no consistent method of grading to predict its aggressiveness has been developed.[1-5]

Since adamantinoma is a rare lesion, there is insufficient data regarding the safest and most effective treatment modality. Wide tumour excision and limb salvage reconstruction surgery, or an amputation, are the currently preferred surgical treatment options. Radiotherapy and chemotherapy have not shown any encouraging results.[10] In the present case, the patient underwent an above knee amputation which is considered a definitive treatment. The surgical margins at the time of surgery were negative for any tumour.

The biologic behaviour of an adamantinoma is highly unpredictable as seen in this case. In some cases, metastases may appear in the lungs or other parts of the body along with a primary bone lesion. These are considered as *synchronous* lesions (Greek: *syn,* "together"; *chronos,* "time"). In contrast, lesions may appear, after definitive treatment of the primary bone lesion. These lesions are named as *metachronous* tumors (Greek: *meta,* "occurring later"; *chromos,* "time"). This implies that metachronous tumors arise independent of the primary tumour and there is no evidence of any metastases after successful treatment of the primary tumour. Such single or multiple metachronous tumors have been described by Jaffe *et al*. in patients who underwent treatment for osteosarcoma.[11]

In the present case, there was no regional lymph node metastasis or lung involvement when the left tibial adamantinoma was treated. The whole body scan done at that time too didn't reveal any additional lesions. The present lesion arose in a side opposite to the initial lesion. Also conspicuously osteoclastic giant cells were noted in the femoral adamantinoma. These weren't seen in the tibial lesion. Since metastatic deposits usually recapitulate the histomorphology of the primary, we excluded the possibility of metastases and considered the lesion to be a metachronous tumour. To the best of our knowledge, such a concept of a metachronous adamantinoma has been never described in literature.

The patient refused surgical intervention and was lost to follow up. As more cases are being reported, it is very clear that a concept of five or ten year survival rates in adamantinoma will only lead to false optimism. With improved surgical treatment modalities and cure rates, metachronous adamantinoma should be recognised as an important sequel in long term survivors.

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