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CASE REPORT

Adamantinoma of Pelvis: A Rare Tumor at an Uncommon Site, with Diagnostic Implications

Jagandeep Singh Virk[1] · Manish Pruthi[2] · Bharat Rekhi[3] · Sunil Pasricha[4]

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Abstract

Adamantinoma is a low-grade malignant bone tumor with metastatic potential in the range of 15–20%, commonly affecting mid-diaphyseal tibial region and jaw. Numerous cases of adamantinoma affecting the appendicular skeleton have been reported but only three in the pelvis till date. We present the case of a 24-year-old male, who was initially reported as fibrous dysplasia for a lesion in pelvis. On review, a cellular tumor was noted, including areas resembling fibrous dysplasia, along with cords and nests of polygonal cells, which displayed positive immunoexpression with cytokeratin and p40. Subsequently, he underwent wide local resection (internal hemipelvectomy), along with mesh pseudoarthrosis was done. Diagnosis of adamantinoma was further confirmed, with clear resection margins. Currently, the patient is on a regular clinical and radiological follow-up. Careful assessment of key histomorphologic features, coupled with immunohistochemical stains and clinico-radiological correlation, is helpful in identifying this uncommon tumor at a rather rare site.

Keywords Pelvis · Biopsy · Pathology · Resection · Pseudoarthrosis

Introduction

The unusual location of the adamantinoma makes this case of particular interest. Invariably, an adamantinoma occurs in the long bones (97% of cases), especially the tibia (80–85% of cases) [1]. Only three cases of adamantinomas occurring in pelvic bones have been reported in literature till date [2–4].

Case History

A 24-year-old male patient presented to us with a history of intermittent pain in his left gluteal region, of 18 months duration, leading to difficulty in weight bearing on his left lower limb.

He consulted a local hospital where he underwent plain radiographs and magnetic resonance imaging (MRI) of the pelvis. The radiographs demonstrated multiple, confluent lytic regions with sclerotic borders in the supra-acetabular region of left ilium, without significant cortical breech or soft tissue component (Fig. 1). MRI of the pelvis demonstrated an expansile, lytic lesion in his left ilium, involving posterior acetabular pillar, with extension up to the articular margins of the left hip joint space along with minor lateral

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✉ Manish Pruthi
manishpruthi@gmail.com

Jagandeep Singh Virk
jaganvirk_09@yahoo.co.in

Bharat Rekhi
rekhi.bharat@gmail.com

Sunil Pasricha
drsunilpasricha@yahoo.com

[1] Department of Orthopaedic Oncosurgery, Paras Hospitals, Panchkula, Haryana, India

[2] Department of Orthopaedic Surgical Oncology, Rajiv Gandhi Cancer Institute, Rohini, Delhi, India

[3] Department of Surgical Pathology, Tata Memorial Centre, Parel, Mumbai, India

[4] Department of Surgical Pathology, Rajiv Gandhi Cancer Institute, Rohini, Delhi, India

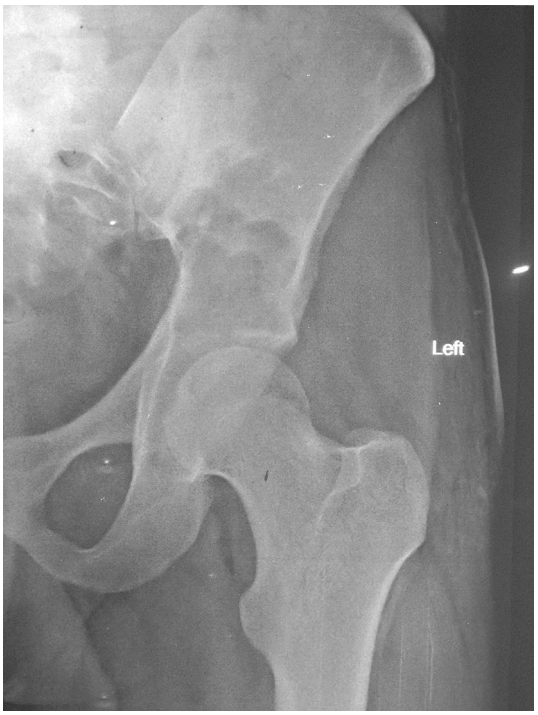

Fig. 1 Radiograph showing multiple lytic regions with surrounding sclerosis in the supra-acetabular region of ilium

cortical breech of the iliac bone. The lesion was heterogeneously hyperintense on T2 and STIR images, while heterogeneously isointense on T1-weighted images (Fig. 2). Whole body PET-CT with F-18 FDG revealed an FDG avid lytic lesion in left iliac bone and left acetabulum (SUV max 10.9). There was no other lesion identified. Differential diagnoses offered on radiologic imaging were fibrous dysplasia, giant cell tumor, desmoplastic fibroma, and chondromyxoid fibroma.

Subsequently, the patient underwent computed tomogram (CT)-guided core needle biopsies, at two occasions, from the lesion, which were initially reported as benign fibro-osseous lesion, elsewhere. Thereafter, the patient consulted another hospital, where he underwent curettage and cementing, which was reported as fibrous dysplasia. The patient remained symptom-free for 3 months, following which he developed similar symptoms of more severe pain in his hip, leading to limping.

At this point, he visited our institution, wherein he underwent repeat radiologic imaging that revealed a pathological fracture and cortical destruction of his ilium with cement in situ in the supra-acetabular region (Fig. 3). Repeat MRI showed diffuse, expansile enhancing altered marrow signal intensity lesion in the adjacent part of his left iliac bone with extensions indicative of residual/recurrent lesion (Fig. 4). Next, he underwent an image-guided core needle biopsy from the lesion. His recent biopsy and paraffin blocks of

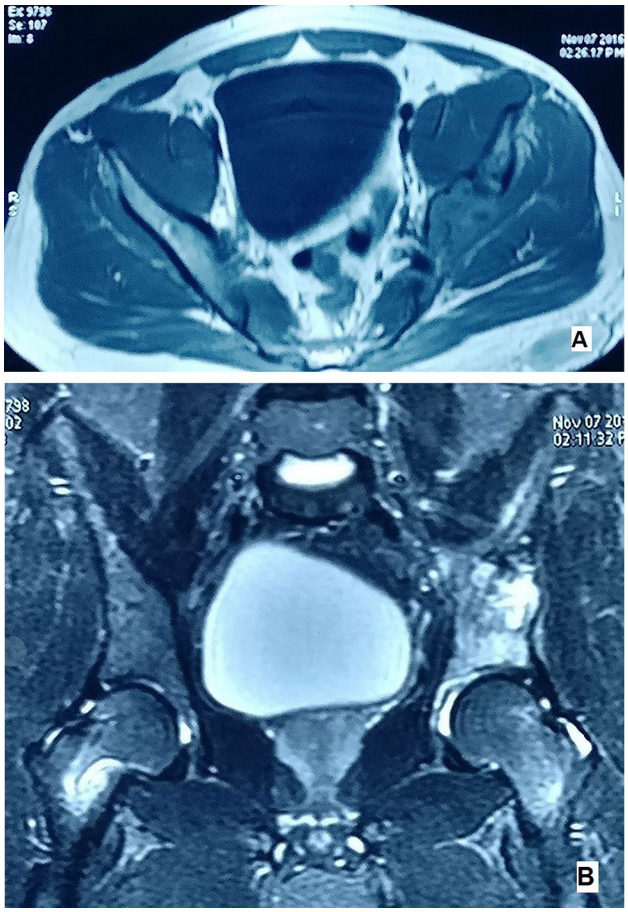

Fig. 2 **a** T1 axial MRI image showing a lesion in his left ilium, involving posterior acetabular pillar. **b** T2-contrast image of the pelvis showing extension up to the articular margins of the left hip joint space

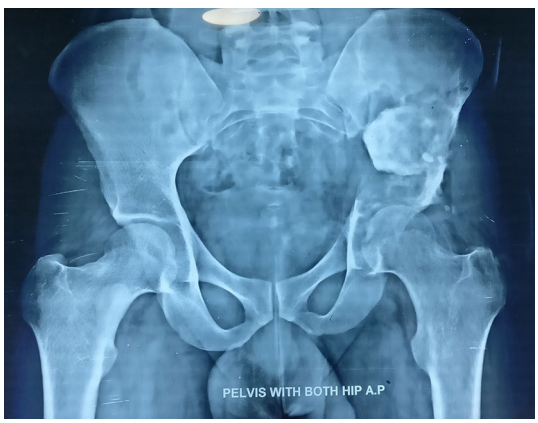

Fig. 3 Radiograph pelvis—3 months following his index surgery (curettage + cementing) showing pathological fracture and cortical destruction of his ilium with cement in situ in the supra-acetabular region

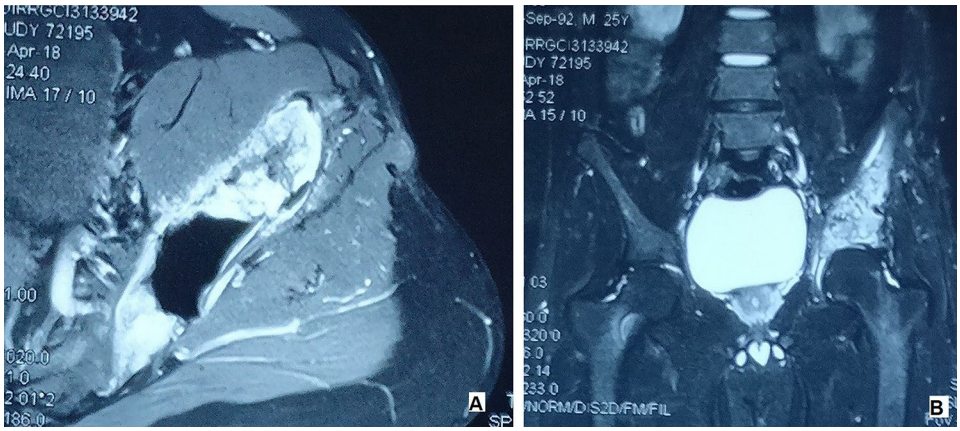

Fig. 4 **a** MRI of the pelvis following index surgery (curettage + cementing). T2 axial image showing cement in situ along with the disease. **b** T2-contrast coronal images—showing diffuse, expansile enhancing altered marrow signal intensity lesion in the adjacent part of his left iliac bone with extensions indicative of residual/recurrent lesion

earlier tumor resection were sent to a sarcoma pathologist for review.

Multiple sections revealed a cellular tumor showing distinct areas reminiscent of fibrous dysplasia. In addition, there were polygonal cells arranged in interconnecting cords and few clusters and nests. By immunohistochemistry, the polygonal cells were distinctly positive for pan cytokeratin (AE1/AE3) and p40 (Fig. 5). Diagnosis of osteofibrous dysplasia-like adamantinoma was offered. Staging investigations in the form of bone scan and non-contrast CT scan of the chest were performed, which did not reveal any metastatic lesion.

In order to facilitate complete surgical excision, patient received two injections of zoledronic acid at a gap of 4 weeks which helped in supra-acetabular ossification (Fig. 6). Subsequently, patient underwent wide local resection of the tumor in the form of internal hemipelvilectomy (Type 1 + Type 2 + part of Type 3) and mesh pseudoarthrosis (Fig. 7). The final histopathological examination of the resected specimen further confirmed the diagnosis of an adamantinoma with osteofibrous dysplasia-like areas. All the resected margins were free of tumor. Our patient has completed 2 years of follow-up and is disease free and able to ambulate independently with a shoe raise.

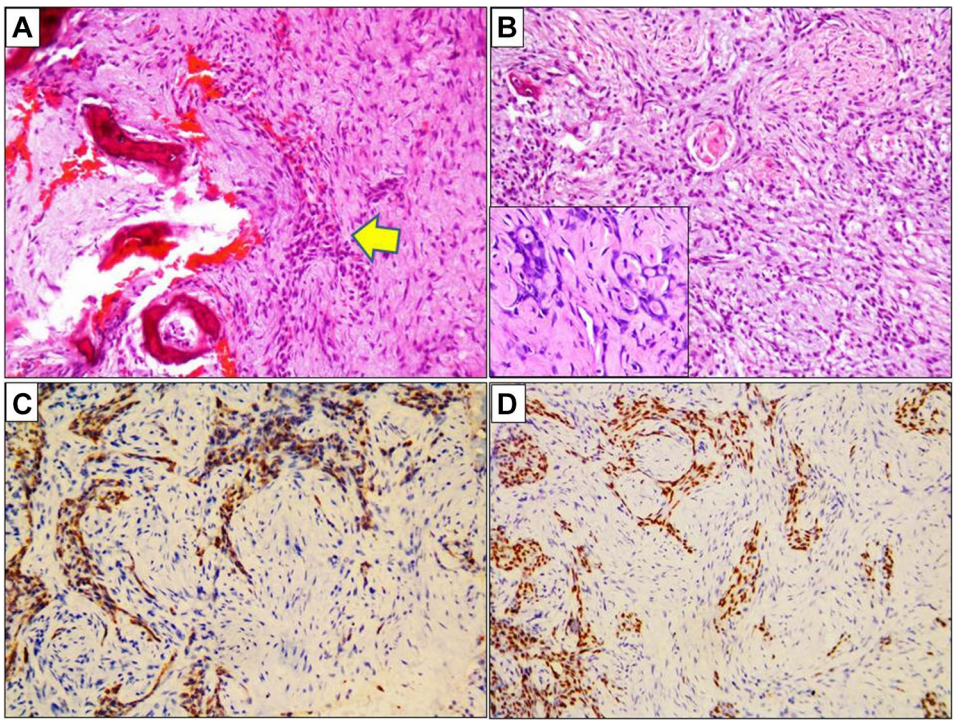

Fig. 5 Microscopic findings. **a** Cellular tumor showing fibro-osseous differentiation reminiscent of a fibrous dysplasia, along with clusters of polygonal/epithelioid cells. Hematoxylin and Eosin, × 200. B. Areas showing prominent cords of epithelioid cells. H and E, × 200. Inset: higher magnification showing focal areas showing clusters and nests of epithelioid cells. *H* and *E* × 400. Immunohistochemical staining (**c**, **d**). **c** Pancytokeratin (AE1/AE3) distinctly highlighting the cords of epithelioid cells. Diaminobenzidine (DAB) × 200. **d** Positive p40 immunoexpression in the epithelioid cells

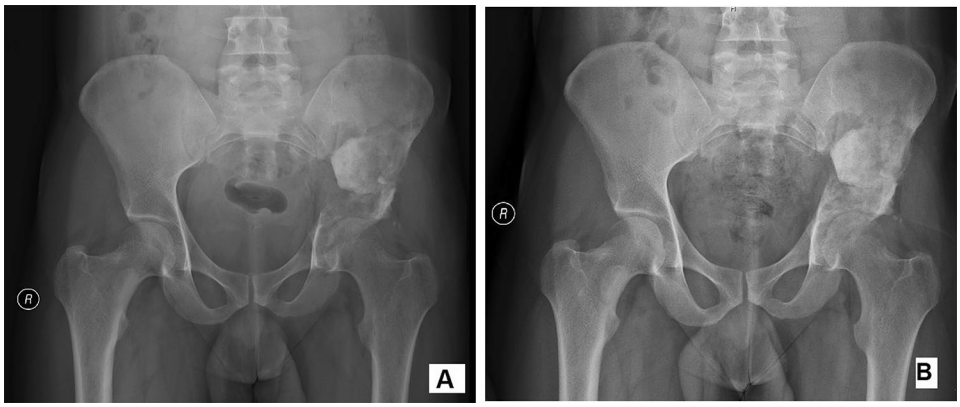

Fig. 6 **a** Radiograph of the pelvis before administration of zoledronic acid showing extensive lysis in the supra-acetabular region. **b** Radiograph following administration of two doses of zoledronic acid at four weekly interval showing ossification in the supra-acetabular region

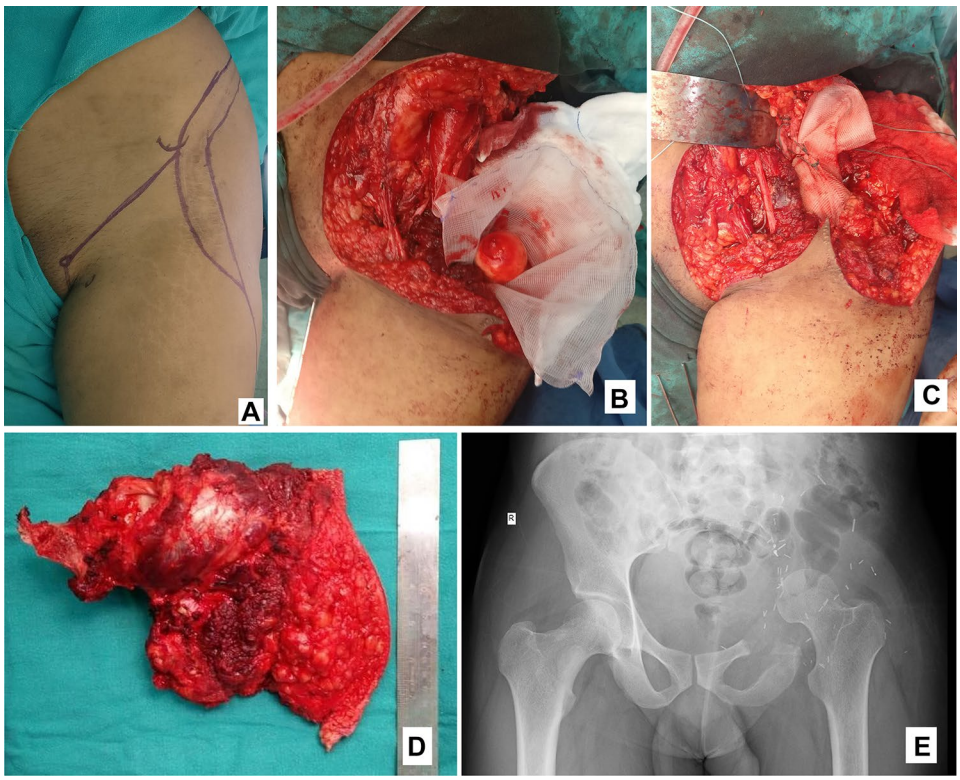

Fig. 7 **a** Skin marking showing the utilitarian incision to pelvis incorporating the scar mark of previous surgery. **b** Prolene mesh reconstruction. **c** Prolene mesh reconstruction completed. **d** Resected gross specimen. **e** Post-op radiograph showing Type 1 + Type 2 + partial Type 3 internal hemipelvectomy

Discussion

On radiologic examination, an adamantinoma displays characteristic features, which are useful in establishment of a presumptive diagnosis, especially when it occurs in its classical location. In the tibia, it usually appears as a central or an eccentric, slightly expansile, sharply or poorly delineated osteolytic, multilocular lesion, as a result of the sclerotic margins and overlapping radiolucencies. The lesion is mostly intra-cortical, but can be destructive, leading to invasion of the extracortical soft tissues in about 15% of cases [5]. The periosteal reaction is variable from minimal to prominent [6]. In addition, complete involvement of the medullary cavity on MRI is suggestive of adamantinoma and therefore imaging can assist in diagnosis [7]. Similar radiological features can be extrapolated to adamantinomas when present at unusual locations, as in the present case, wherein final diagnosis was achieved on histopathologic examination.

Histopathologically, adamantinoma is classified into two distinct types: classic and OFD-like adamantinoma. Classic adamantinoma is characterized by admixture of both epithelial and osteofibrous components. OFD-like adamantinoma is characterized by predominance of osteofibrous tissues, in which small groups of epithelial cells are detected on a careful search or immunohistochemistry. In the present case, while part of biopsy material revealed OFD-like

adamantinoma, there were otherwise substantial areas indicative of adamantinoma, including on sections from the resected specimen. The diagnosis was reinforced by positive immunohistochemical expression of antibody markers, such as cytokeratin and p40.

There are usually overlapping features (both radiologically and pathologically) between fibrous dysplasia, osteofibrous dysplasia, and adamantinoma, which usually lead to misdiagnosis. Springfield et al., in their review of 32 cases (with initial diagnoses of fibrous dysplasia, osteofibrous dysplasia, or adamantinoma of the tibia) highlighted similar problem. Six of 9 patients with classic adamantinoma and 6/10 patients with OFD-like adamantinoma had been previously diagnosed as fibrous dysplasia and OFD [8]. There is also a possibility of progression of OFD into OFD-like adamantinoma or a classic adamantinoma. In the study of Hazelbag et al. (32 patients of adamantinoma), there were 7 cases of OFD-like adamantinoma, of which 3 progressed to classic adamantinoma with 1 developing metastasis [9].

In a recent study by Scholfield et al., five of the eight patients initially treated as OFD actually harbored OFD-like adamantinomas, to begin with, who subsequently developed progressive disease and required surgery [10]. This was observed in another study too and thus highlights the combined importance of imaging and pathology in the diagnosis of adamantinoma [11].

The treatment of a conventional adamantinoma is wide resection with no proven role of any adjuvant therapy. Various reconstruction options can be employed, such as allografts, autografts, or endoprosthesis. The treatment for OFD-like adamantinoma is not well established due to the scarcity of cases in literature. There is a trend towards undertaking wide resection of the tumor rather than curettage, in cases of OFD-like adamantinomas [7, 9].The aggressiveness of OFD-like adamantinoma, compared with OFD, is seen not only on radiological appearance, but also clinically, with more than 80% cases presenting with pain requiring treatment. We carried out internal hemipelvectomy (Type 1 + Type 2 + part of Type 3), based on the patient's clinical features of unrelenting pain, disease extension as gauged on MRI, and histological confirmation of an adamantinoma. Principles of limb salvage surgeries in this condition remain same as in other primary malignant bone tumors, which is ability of achieve tumor free margins and an expected function superior to amputation. As there is no proven adjuvant treatment in adamantinoma, stress should be given to achieve wide margins.

Although adamantinoma is a malignant tumor with a previously reported ten-year survival of 87.2% and given the incidence of late local recurrences and distant metastases, a long-term follow-up is usually recommended [10, 11]. Accordingly, we have also recommended a long-term follow-up to our patient.

The present case constitutes the 4th reported case of adamantinoma in the pelvic bones, including the first such case reported from the Indian subcontinent. Careful assessment of key histomorphologic features, coupled with immunohistochemical stains and clinico-radiological correlation, are helpful in identifying an adamantinoma at a rare location, as in the present case. A review by a sarcoma pathologist in such diagnostic challenge cannot be overemphasized. A correct diagnosis has significant implications as it could have avoided an intralesional surgery in our case. Wide resection with negative margins forms the mainstay of the treatment of adamantinoma of the bone.

Patient Declaration Statement

"The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed."

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Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical standard statement This article does not contain any studies with human or animal subjects performed by the any of the authors.

Informed consent For this type of study informed consent is not required.

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