TIBIAL ADAMANTINOMA:
Limb sparing surgery with reconstruction using allograft bone

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Abstract
We present our experience in the management of four patients with adamantinoma of the tibia. All patients underwent limb-sparing surgery with wide margins being obtained in three patients. The gap was reconstructed using fresh-frozen allograft bone. Vascularised fibular autograft was used in addition to the allograft in two patients. The median duration of follow-up was 106 months.
Material and Methods

A review of the records of the Queensland Bone Tumour Registry between 1991 and 2002 revealed four cases of tibial adamantinoma. Medical records including clinical notes, histology slides and roentgenograms were reviewed. Follow-up information regarding local recurrence, systemic metastases, and patient outcome was obtained in all cases. Mean follow-up was 106 (74-135) months. There were 2 male and 2 female patients. The average age at the time of diagnosis was 28 years (range 18-35 years).

Table 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs.)/Sex</th>
<th>Treatment</th>
<th>Complications</th>
<th>Followup (months)</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>33/F</td>
<td>Wide excision &amp; Allograft</td>
<td>None</td>
<td>135</td>
<td>NED</td>
</tr>
<tr>
<td>2</td>
<td>27/M</td>
<td>Wide excision &amp; Allograft + Vascularised fibula</td>
<td>None</td>
<td>88</td>
<td>NED</td>
</tr>
<tr>
<td>3</td>
<td>18/M</td>
<td>Wide excision &amp; Allograft + Vascularised fibula</td>
<td>Allograft infection – 3 months. Pulmonary metastases – 62 months.</td>
<td>74</td>
<td>AWD</td>
</tr>
<tr>
<td>4</td>
<td>35/F</td>
<td>Marginal excision &amp; Allograft</td>
<td>Local recurrence-72 months Sacral metastases – 120 months</td>
<td>128</td>
<td>AWD</td>
</tr>
</tbody>
</table>

NED: No evidence of disease, AWD: Alive with disease

Case 1: A 33-year-old lady presented with anterior leg pain that had been present for three months. Roentgenograms (Fig. 1) revealed an adamantinoma in the proximal one-third of the tibial diaphysis, which was subsequently confirmed on biopsy. This was managed with wide resection of the lesion and the defect reconstructed with allograft. The patient declined the use
of a vascularised fibular graft. Radiographs 11-years postoperatively showed good healing at both ends (Fig. 2). The patient is asymptomatic and pursuing an active life-style.

**Fig. 1:**
*Pre-operative radiographs.*

**Fig. 2:**
*Post-operative radiographs*

**Case 2:** A 27-year-old male sustained a pathological fracture of the proximal one-third of the tibial diaphysis. Biopsy confirmed the lesion to be an adamantinoma. It was treated with wide resection and the intercalary gap filled with tibial allograft along with the contra-lateral vascularised fibula. Follow-up X-rays four years postoperatively revealed a new lesion distal to the previous resection. Biopsy revealed this new lesion to be fibrous dysplasia, which was treated by curettage. At a seven year follow-up the patient is asymptomatic and there is no evidence of recurrence.

**Case 3:** An eight-year-old boy sustained a pathological fracture of the tibial shaft. Roentgenograms revealed a diagnosis of fibrous dysplasia, which was confirmed on biopsy. The lesion was curetted and grafted and the limb immobilised in a cast. The boy fractured the tibia once again after two years and was managed conservatively in a cast. At the age of 16 years, the boy had another fracture of the same tibia and was treated with intramedullary nail
fixation. Two years later, when the boy was 18 years old, he was referred to our institution, for what was suspected on roentgenograms to be an adamantinoma at the site of the previous lesion. Biopsy confirmed the diagnosis and the lesion was widely resected. This required a complex osteotomy to resect almost the whole of tibia as it was considered contaminated following the nail fixation. The gap was filled with tibial allograft and vascularised fibular graft.

Unfortunately, three months later, the wound became infected with Methicillin-resistant Staphylococcus aureus and Vancomycin-resistant Enterobacter. The patient required multiple debridements and eventually had to undergo a below-knee amputation four years after the resection and allograft procedure. The patient developed bilateral pulmonary metastases 62 months after resection of the adamantinoma. At the time of writing (74 months follow-up) the patient is alive with disease and the chest metastases have remained unchanged over a period of 12 months.

**Case 4:** A 35–year lady was diagnosed with adamantinoma of the distal third of the tibial diaphysis. This was treated elsewhere with a marginal excision keeping the opposite cortex of the tibia intact. The diaphysis was reinforced with a strut allograft. After a disease free interval of six years the patient presented with a recurrence and a pathological fracture of the strut allograft. The patient was referred to our institution for further management and a below knee amputation was performed. She developed biopsy-proven sacral metastases, ten years following excision of her tibial lesion. The patient refused treatment for these. The lesion has not progressed and the patient is alive with disease at the last follow-up.
Discussion

Adamantinoma of the long bones is a rare tumour which constitutes less than 1% of all malignant bone tumours [2,7,17,20]. More than 80% occur in the tibia [9,14,16,19,21,22], typically in the anterior mid-diaphysis. Adamantinomas have characteristic radiographic appearances with sharply outlined lucencies containing sclerotic foci that sometimes surround the lesion [4,8,10]. MR imaging characteristics correlate well with the histologic features and biological behaviour [3,19].

The peak incidence of adamantinoma is in the second and third decade although the age range is reported as 10-62 years [9,21]. Adamantinomas have also been reported in children, but the histological pattern in this age group is different from that seen in adults and resembles osteofibrous dysplasia. The usual pattern of adamantinoma in children has been termed "differentiated adamantinoma" and follows a benign course [9].

Immunohistochemical and ultrastructural studies have proven beyond doubt the epithelial derivation of adamantinoma [9,18,22]. A probable histogenetic relationship exits between adamantinoma, osteofibrous dysplasia (ossifying fibroma) and differentiated adamantinoma or osteofibrous dysplasia-like adamantinoma [9]. Adamantinoma is composed of epithelial cells surrounded by fibrous stroma [9,10,16]. Several histologic variants have been described including tubular, basaloid, squamous, spindle cell, osteofibrous dysplasia-like and most recently Ewing’s-like adamantinoma or adamantinoma-like Ewing’s [5,9]. In our series, the four cases had varying proportions of the basaloid, squamous and spindle patterns.

Additionally, cases 2 and 3 had areas of fibrous dysplasia and osteofibrous dysplasia respectively. Gebhardt et al. [4], in their series of nine patients had a case in which there was an area of fibrous dysplasia in the proximal part of the tibia that was distinct from the adamantinoma of the shaft. Adamantinoma is a histologically diverse tumour and can mimic a
variety of lesions like fibrous dysplasia, vascular neoplasms, metastatic carcinoma, osteofibrous dysplasia and differentiated adamantinoma [6,10,16].

There has been extensive debate regarding the management of these tumours. Chemotherapy is ineffective and they have been found to be radioresistant [6,17]. Initially these tumours were often managed by intralesional or marginal excisions. The study by Moon and Mori [14] showed that local excision led to an unacceptably high rate of local recurrence. High recurrence rates with eventual metastases and mortality prompted several authors to advocate ablative surgery with amputation [22]. However, many studies have shown that wide excision with limb salvage is adequate treatment for adamantinoma [1,4,10,17,22].

Qureshi et al [17] showed in their series, that patients, who had wide operative margins, had a significantly lower prevalence of local recurrence than those who had less-than-wide margins. In the study by Hazelbag et al [6], of the eleven patients who had been managed with curettage or an excisional biopsy, nine had a local recurrence. In contrast, of the eight patients who had been managed with an en bloc resection, none had a local recurrence or metastases. In our study, three of the four patients were managed with wide resection and limb salvage. The fourth patient, who was referred to us after local recurrence, was managed elsewhere with a marginal excision.

The use of various methods including allografts, vascularized fibular transfers and metallic spacers have been described to bridge the intercalary gap resulting from excision of diaphyseal tumours [11,12]. Review of literature does not reveal as to which form of reconstruction has better results. The gap in the tibia, in all the four patients in our study, was reconstructed using an allograft. Two patients, in addition, received a vascularised fibular autograft from the contralateral limb.
Nonunion, fracture and infection are the oft-quoted complications associated with the use of allografts [15]. We encountered infection in one patient who subsequently developed a pathological fracture of the allograft and had to undergo an above knee amputation.

Rates of local recurrences have been reported to be about 30 percent [6,10]. Keeney et al [10] found a recurrence rate of 31% at an interval ranging from 3 months to 19.4 years. In our series, one of the four patients developed a local recurrence after a disease-free interval of six years. The rates of metastases have been reported to be about 15 to 20 percent [6,10,13,17]. Two of the four patients in our study developed distant metastases at 62 and 120 months respectively. One developed bilateral pulmonary metastases and the other developed metastases to the sacrum. Fillippou and others [2] reported a case of local recurrence of an adamantinoma localized in the tibia, along with the presence of lung metastases, 24 years after diagnosis and surgical treatment of the primary tumour. Qureshi et al [17] showed a survival rate of 87.2 percent at ten years. In our series all patients were alive at a median follow-up of 106 months.

Adamantinoma of long bones is an extremely rare primary malignant bone tumour. Whenever a diagnosis of adamantinoma is considered, a clinical and radio-pathological correlation should be made to ensure the correct diagnosis. The pathologist should examine the entire specimen and if necessary employ anticytokeratin immunohistochemistry [8]. This should help in correctly differentiating adamantinoma from histogenetically similar but benign lesions like osteofibrous dysplasia and differentiated adamantinoma.

The treatment of adamantinoma with limb salvage, which includes resection with wide margins, provides lower rates of local recurrence. Reconstruction with intercalary allograft provides good functional outcomes. However, long-term follow-up is essential to ensure that failure has not occurred.
References


