CASE REPORT

Below-knee amputation through a joint-sparing proximal tibial replacement for recurrent tumour


We report a case which highlights the progression of osteofibrous dysplasia to adamantinoma and questions whether intralesional curettage is the appropriate treatment. The role of a joint-sparing massive endoprosthesis using cortical fixation is demonstrated and we describe a unique biomedical design which resulted in the manufacture of an end cap to allow amputation through a custom-made proximal tibial replacement, rather than an above-knee amputation following recurrence.

The term osteofibrous dysplasia of the tibia and fibula was first used by Campanacci and Laus.1 They highlighted the histological similarity of this condition to that of fibrous dysplasia and its predilection for the tibia. It occurs rarely outside the tibia.2 We report a case of osteofibrous dysplasia of the tibia which progressed to adamantinoma with osteosarcomatous change, which was treated by joint-sparing proximal tibial replacement. Subsequent below-knee amputation was required for recurrence.

Case report

A 37-year-old woman presented with a pathological fracture of her left tibia as a result of a low-energy skiing injury. The diagnosis of fibrous dysplasia was made following a biopsy of the site of the fracture. She was treated in a cast for two months and subsequently underwent intralesional curettage, and bone grafting. She remained asymptomatic for three years and then developed further pain; plain radiography showed recurrence of the disease. Two subsequent percutaneous biopsies confirmed the diagnosis of fibrous dysplasia and she was treated by a second curettage and cementation with wire reinforcement.

After this procedure she remained free from pain for a further year before presenting for a third time with pain. She received a course of monthly intravenous infusions of pamidronate over a period of eight months. This had little effect and she was referred to the Royal National Orthopaedic Hospital. The lesion had enlarged and imaging suggested the diagnosis of adamantinoma (Fig. 1). A biopsy revealed a high-grade malignant bone-forming neoplasm which was extensively positive for the cytokeratins MNF 116 and AE1/AE3, but not CAM5.2. The biopsy specimens taken at the initial presentation and three years later from both curettings were reviewed and a diagnosis of osteofibrous rather than fibrous dysplasia was made. The features of a bone-forming fibrous lesion in which there were numerous scattered cytokeratin cells positive for MNF 116 + and AE1/AE3 but not for CAM5.2 in the absence of well-formed epithelial structures, supported this diagnosis (Fig. 2). There was no evidence of histological progression over the three years. The diagnosis of osteofibrous as opposed to fibular dysplasia was also supported by the failure to detect a GNAS1 mutation.3 In light of the recent imaging and previous pathology a diagnosis of dedifferentiated adamantinoma showing osteosarcomatous differentiation was then made (Fig. 3). This was confirmed on the fully excised specimen when a joint-sparing proximal tibial replacement (Stanmore Implants Worldwide, Stanmore, United Kingdom) was performed in February 2007 (Fig. 4). Although imaging showed no metastatic disease, the patient received adjuvant PAM chemotherapy (cisplatin, doxorubicin and methotrexate) for six months.

One year after surgery she had nearly full function at the knee with a range of movement of 0° to 100° and could cycle, swim and regularly attend the gymnasium.

In April 2008, she presented with a recent history of a tender swelling on the medial aspect of the calf. Ultrasonography and MRI demonstrated a soft-tissue mass (19 cm × 5 cm) extending from the distal prosthesis-bone interface.
(Fig. 5). Percutaneous biopsy showed a recurrence of the dedifferentiated adamantinoma with osteosarcomatous change.

Further limb salvage was considered to be impracticable and in May 2008 a below-knee amputation was performed. The proximal tibial device was exposed and uncoupled leaving a residual 7 cm implant in situ. A small end cap was attached to the remaining prosthesis and a myocutaneous flap fashioned over it (Fig. 6). The post-operative course was uncomplicated. The wound healed well and the patient was discharged five days after the operation. She had a further five cycles of adjuvant chemotherapy with ifosfamide and etoposide. At follow-up at six months she was walking freely with her below-knee patellar-tendon-bearing prosthesis with a supracondylar suspension and laminated socket, which spread the load across the patella and decreased direct distal contact with the stump. The foot component was an Endolite (Endolite, Basingstoke, United Kingdom) multiflex foot and ankle providing multiaxial movement at these two joints. Gait analysis demonstrated a walking velocity of 61 m/min at a self-selected speed and 84 m/min at a purposely-fast speed. She had a pain-free range of knee movement from 0° to 100°.

Discussion

Although fibrous and osteofibrous dysplasia are fibro-osseous lesions they are recognised as different diseases. The former results from a GNAS1 mutation which has never been demonstrated in osteofibrous dysplasia and cytokeratins present in osteofibrous dysplasia and adamantinoma are absent in fibrous dysplasia. Furthermore, antibodies which recognise cellular molecules have been used successfully to subclassify the disease. Adamantinoma is an important differential diagnosis but is difficult to separate radiologically and histologically from osteofibrous dysplasia. There is extensive evidence that osteofibrous dysplasia is the precursor of adamantinoma and Hazelbag et al. in a study of 32 cases showed that both tumours express the same class of cytokeratins. The epithelial/cytokeratin-positive cells represent a minor component in osteofibrous dysplasia, but become the dominant cellular element in adamantinoma.
noma. Recognition of the potential of osteofibrous dysplasia to transform into adamantinoma is important since the risk of metastatic adamantinoma is reported to be as high as 20%. In addition, osteofibrous dysplasia occurs in children, occasionally in newborn infants and rarely above the age of 15 years whereas adamantinoma generally presents after this age.

Our case report highlights the histological overlap between fibrous and osteofibrous dysplasia and the importance of performing immunohistochemistry for cytokeratins.
on tibial and fibular fibro-osseous lesions which appear to be benign. The current tumour was widely sampled twice and showed a ‘bland’ lesion without obvious epithelial elements. We believe that a genetic mutation occurred in the residual tumour, resulting in transformation to adamantinoma. Dedifferentiation of an adamantinoma is rare with few cases hitherto reported. However, our case highlights the risk of the transformation of residual osteofibrous dysplasia into an aggressive neoplasm with a high metastatic potential.

In view of the early recurrence after an en bloc resection, the existence of residual disease was almost certain despite a histological report of clear margins. One explanation for this is that the two previous curettage procedures resulted in seeding of tumour cells outside the resection field. This is impossible to prove but in our experience, there is a considerable risk of local recurrence of osteofibrous dysplasia after curettage, and our practice is to perform excision rather than curettage.

Since the adamantinoma and its local recurrence did not involve the proximal tibial metaphysis, a joint-sparing prothetic procedure was possible. Such prostheses have been particularly successful in the distal femur but hitherto there have been no reports on joint-sparing proximal tibial replacements. However, joint-sacrificing cemented proximal tibial replacements have a high incidence of aseptic loosening, which would be lessened by preserving the knee and thereby reducing stress of the prosthesis-bone interface.

Our joint-sparing proximal tibial prosthesis is a modular device comprising a stem, a hydroxyapatite collar and a titanium shaft which slides into the proximal part and is locked in place by way of male-female integrated ridges and two locking bolts (Fig. 7). For strong and stable fixation at the knee the proximal and distal components are fixed separately and then locked together. The distal part is first cemented into the medullary canal, the proximal part is fixed to the remaining tibial metaphysis through the cortical screws, and the two components are locked together by two locking bolts.

The fashioning of an end cap to fit the proximal part of the prosthesis has not been previously reported. The cap is bolted to the prosthesis (Fig. 8) and leaves a rounded end which allows for the stump to be closed and reduces the risk of pressure necrosis in the overlying soft tissues.

Six months after the amputation procedure the patient was walking freely, and her kinematics were similar to those of a conventional below-knee amputation. The walking velocities at a self-selected and purposely-fast speed were also comparable with those of below-knee amputation previously published. Waters et al showed that walking velocity was greater in below-knee than above-knee amputations (71 m/min and 52 m/min, respectively). The relative energy cost is markedly increased in above-knee amputees whose lifestyle is thereby less active. We consider that by salvaging the proximal part of the prosthesis we have enabled this patient to have a better quality of life and functional outcome.
We wish to thank Dr J. Johnston, for providing the histology slides from the patient's original presentation in USA and Dr M. Thornton for gait analysis. UCL is a partner of the Eurobonet consortium, a European commission granted network of excellence for studying the pathology and genetics of bone tumours. We obtained full informed consent from the patient involved to discuss her case and informed her that the case would be submitted for publication. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References