

Adamantinoma arising in the distal end of the fibula

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Abstract

Adamantinoma is a rare, low-grade, malignant bone tumor. It frequently occurs in the tibia but rarely arise in the distal end of the fibula. This study reported a case of adamantinoma arising in the distal end of the fibula, resulting in good prognosis. A 38-year old female felt left ankle pain, and was suspected as having a bone tumor at the distal end of the fibula by X-ray. She was diagnosed as the classical adamantinoma of the fibula by open biopsy. En bloc wide resection of the tumor, primary arthrodesis of the ankle was performed. During the follow-up period of 7 years after the surgery, she has lived without any metastasis and local recurrence. A wide resection and arthrodesis of the ankle joint can provide a good outcome for adamantinoma arising in the end of the fibula.

Introduction

Adamantinoma is a rare, low-grade, malignant bone tumor accounting for approximately 0.4% of all primary bone tumors.¹ It frequently occurs in young to middle-aged adults (20 to 40 years of age) and slightly common in males (male-to-female ratio is 1.3:1).² Anterior metaphysis or diaphysis of the tibia is involved in 85-90% of cases, but the other sites, including the fibula, ulna, femur, humerus, and radius have been reported.^{1,3} Its histopathology shows biphasic patterns of epithelial cells and osteofibrous components. It is thought that there are two types of adamantinoma: the *classical*, more aggressive, type and the *differentiated*, more benign form. The latter one resembles osteofibrous dysplasia (osteofibrous dysplasia like adamantinoma). The former one is characterized by epithelial

and osteofibrous components, which may be intermingled in various proportions and differentiation patterns. The four main differentiation patterns of classical adamantinoma are basaloid, tubular, spindle cell, and squamous. In immunostaining, it shows expression for keratin, cytokeratin (CK) 5, 14, 19, epithelial membrane antigen (EMA) and vimentin.¹

Though there have been no definitive guidelines for a treatment of adamantinoma, a wide excision with clear margin is the most effective treatment, but radiotherapy and chemotherapy are not efficient.²⁻⁴ Because of the frequency, there are many reports about surgical treatments for adamantinoma arising in the tibia.⁵⁻⁷

Adamantinoma arising in the distal end of the fibula is extremely rare. To the best of our knowledge, there are only two reports about adamantinoma arising in this area. This study reported a case of adamantinoma arising in the distal end of the fibula, resulting in good prognosis after a wide resection.

Case Report

A 38-year old female felt pain in the left ankle after sprain. Two months after the injury, she visited a local hospital because the pain had gotten worse. She was suspected of having a bone tumor at the distal end of the fibula by X-ray, and referred to our hospital. At the initial visit, the pain decreased while resting and walking. Physical examinations revealed a tenderness on the distal end of the fibula, but there was no swelling and restriction in range of motion of the ankle joint. She had no family history of this type of injury. Plain radiographs showed multilocular osteolytic lesions in the distal end of the fibula with thinning of the cortex (Figure 1a). On magnetic resonance imaging (MRI), the lesion showed intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The septum was enhanced by an injection of gadolinium (Figure 1b-d). Although the tumor extended surrounding soft tissue, distal tibiofibular joint was not involved (Figure 1e). Osteofibrous dysplasia, fibrous dysplasia, giant cell tumor, metastasis, abscess or adamantinoma were suspected from the image analyses.

An open biopsy was performed under local anesthesia. Histology showed biphasic pattern of epithelial cells and fibrous components with a tubular epithelial pattern (Figure 1f, g). Strong immunoreactivity of CK5 and vimentin were observed in the epithelial cells (Figure 1h, i). From these findings, she was diagnosed as the classical

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adamantinoma of the fibula.

One month after the open biopsy, we planned en bloc wide resection of the tumor. An oval shaped skin incision was made around the linear scar of the open biopsy. The distance between the oval skin incision and linear scar was kept 3 cm. At the proximal side, the fibula was cut at the same level of the oval shaped skin incision. At the distal side, lateral ligaments of the ankle and the tendon of the peroneal muscle were resected together with the distal end of the fibula. In the anterior and posterior sides, posterior part of the extensor digitorum longus muscle (EDL) and anterior part of the flexor hallucis longus muscle (FHL) were resected with fibula to cover the tumor, respectively. In the medial side, the interosseous membrane was resected with the fibula at the margin of the tibia. Finally, distal half of the fibula, lateral ligaments of the ankle, EDL, peroneal muscle, FHL and the interosseous membrane were resected. Because instability of the ankle joint was expected after the resection, arthrodesis of the ankle with intramedullary Nail with Fins (Smith and Nephew, London, UK) were performed. Reconstruction of soft tissues was performed to cover the defect by using free muscle flap of the Latissimus dorsi

muscle (Figure 2). The surgical margin of the resected specimen was evaluated according to the evaluation system of the Japanese Orthopaedic Association⁸ and a 1cm wide margin was achieved. From the surgical specimen, histology showed the same findings to the biopsy specimens. Her left ankle had been immobilized for 4 weeks using splints. She was allowed partial weight bearing (one-third of her body weight) gait at 4 weeks after the surgery. Weight bearing was increased gradually after 4 weeks, and she back to full weight bearing gait at 8 weeks. During the follow-up period of 7 years after the surgery, she has lived without any metastasis and local recurrence, and joined social activities without any prosthesis.

Discussion

There are many reports about surgical treatments for adamantinoma arising in the tibia.⁵⁻⁷ After en block wide resection, the limb reconstruction can be performed with distraction osteogenesis, allografts, vascularized fibular autografts, non-vascularized autogenous bone grafts and metallic segmental replacement.⁹ Adamantinoma arising in the fibula is very rare and distal end of the fibular cases are more. To the best of our knowledge, reports about adamantinoma arising in the area were less than 20 cases. Among them, 5 cases were reported with distal fibulectomy for the initial treatment like ours. Borbas *et al.* reported a case was in good outcome from both oncological and functional point of views after wide resection without surgical reconstruction.¹⁰ Mohler *et al.* reported a patient who got good functional outcomes after marginal resection without surgical reconstruction, but tumor metastasized to the lung.¹¹ The patient reported by Papagelopoulos *et al.* had a second adamantinoma at the ipsilateral distal tibia at 17 years after marginal resection, and below-knee amputation was performed.¹² Baker *et al.* and Beabout reported similar cases needed to below-knee amputation after initial treatment (Table 1).^{13,14}

Because of the rareness in adamantinoma, there are no definitive guidelines for the treatment. However, some reported about the excisional margins of it.^{1-4,9,11,12,15} Curettage was not recommended because of the high frequency of recurrence.³ Marginal surgery is one of the risk factors for recurrence in adamantinoma. Recurrence rate after non-radical surgery rose up to 90%.¹ The most effective treatment is recognized as a wide excision with clear margins.^{2,4} Qureshi *et al.* reported in a review of 70 patients, en bloc tumor resection with wide

margins and limb salvage were resulted in a 10 years survival rate of 87.2%. The patients who had wide operative margins had a significantly lower prevalence of local recurrence than those who had less-than-wide margins.¹⁵ Several authors advised amputation, emphasizing the multicentric nature of adamantinoma.³ Jain *et al.* suggested that amputation may be advisable if local recurrence occur or if en bloc resection and limb salvage are not an option.⁹ However, amputation for adamantinoma has not been proved to improve survival rate when compared with the limb preserving surgery.⁹ It is difficult to keep surgical margins around the ankle joint for limb sal-

vage surgery. In the case of a tumor arising in the distal end of the fibula, a wide margin can be achieved with the followings guidelines: only intraosseous tumor without involving the distal tibiofibular joint and the intraarticular resection. If the tumor involves the distal tibiofibular joint, it is necessary for *en bloc* resection of the joint with a resecting lateral part of the tibia to achieve a wide margin. Papagelopoulos *et al.* reported a case with intrarticular marginal resection, and the patient presented local recurrence. They did not report about expansion of the tumor.¹² Mohler *et al.* reported a case with an intrarticular marginal resection, and the patient did not present

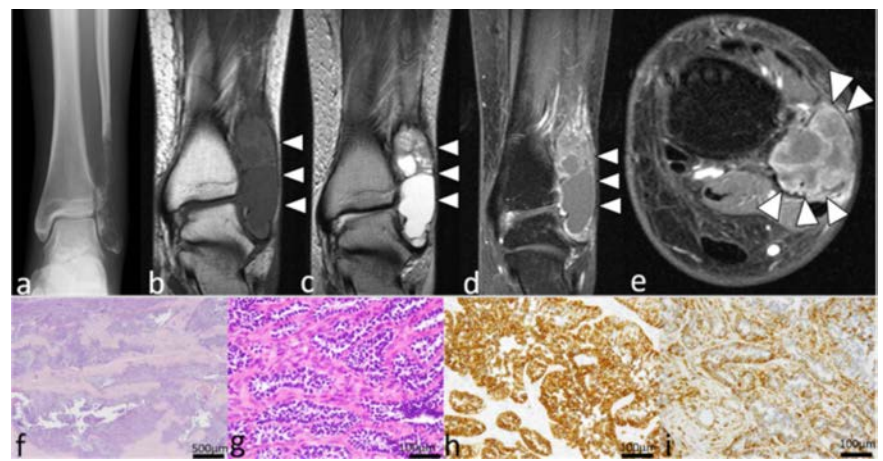


Figure 1. Plain radiograph of anteroposterior view (a). Magnetic resonance imaging (MRI) images (b-e). T1 weighted coronal image (b). T2-weighted coronal image (c). T1 fat suppression with an injection of gadolinium coronal (d) and axial (e) images. Arrow heads indicate the tumor. Hematoxylin and eosin stained biopsy specimen demonstrates biphasic pattern of epithelial cells and fibrous component (f: $\times 40$). The epithelioid cells make tubular structure (g: $\times 200$). Immunostaining for CK5 (h: $\times 200$) and vimentin (i: $\times 200$).

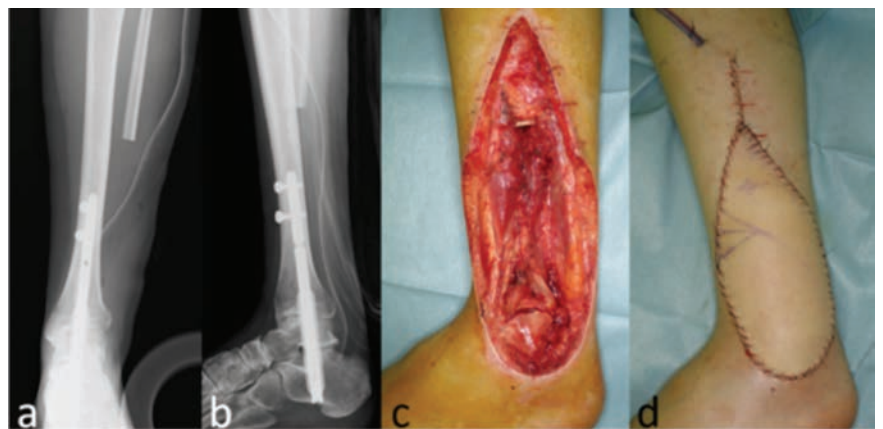


Figure 2. Plain radiograph of anteroposterior view (a) and lateral view (b) at immediately after operation. Perioperative photograph after en bloc wide resection (c) and reconstruction of soft tissue by using free muscle flap of Latissimus dorsi (d).

Table 1. Literature review of adamantinoma arising in the distal end of the fibula. Distal fibulectomy was performed for initial treatment in these cases.

Authors	Age, gender	Resection type	Adjuvant therapy	Reconstruction	Follow-up (years)	Oncological result	Functional result
Baker <i>et al.</i> 1954	35, M	Uncertain →BKA	None	No reconstruction	19	Uncertain	Uncertain
Mohler & Cunningham, 1997	13, M	Marginal	None	No reconstruction, cast (4 weeks)	3	Alive, lung metastasis	No pain, brace, deformity
Beabout, 1997	35, M	Uncertain →BKA	None	No reconstruction	21	Uncertain	Uncertain
Papagelopoulos <i>et al.</i> , 2005	35, M	Marginal →BKA	None	No reconstruction, cast (4 weeks)	22	Alive, NED	BKP
Borbas <i>et al.</i> , 2012	24, M	Wide	None	No reconstruction	5	Alive, NED	No complaint
Our case, 2016	38, F	Wide	None	Ankle arthrodesis	7	Alive, NED	No pain, no brace

BKA, below-knee amputation; BKP, below-knee prosthesis; NED, no evidence of disease.

local recurrence with short follow-up period (Table 1).¹¹ In our case, tumor did not involve the distal tibiofibular joint. Therefore intraarticular resection was performed, and wide margin was achieved.

The biologic behavior of adamantinoma is unpredictable.³ Even if a wide margin was achieved, local recurrence and metastasis can occur. It is not uncommon to develop recurrence and metastasis even up to 10 years after detection of the primary occurrence. Szendrői *et al.* and Filippou *et al.* reported cases who developed recurrence at 36 and 24 years after the primary resections, respectively.^{16,17} Khémiri *et al.* reported a case who developed pulmonary metastasis at 1 year after a complete resection of the primary lesion.⁴ In our case, a wide margin was achieved and any metastasis and local recurrence has never seen for 7 years after the primary surgery. However, careful follow-up is necessary.

After distal fibulectomy, one of the main concerns is the ankle joint instability. The stabilizing role of the lateral malleolus in the ankle joint has been reported.^{18,19} Absence of the lateral malleolus leads to lateral displacement of the talus. Such instability may predispose a patient to development of arthrosis in the ankle joint. In some cases, distal fibulectomy without reconstruction has shown in good results,^{12,20} however, progressive ankle valgus deformity, instability, and degenerative changes have been reported.¹² Several reconstructive options have been reported after fibulectomy. Ligamentoplasty, as lateral ankle ligament repair to the lateral tibia or as attachment of the peroneal tendons to the lateral tibia has been reported. Other reconstructive options include allograft transplantation, reversing the ipsilateral proximal fibula, using of a vascularized contralateral proximal fibular graft, primary ankle arthrodesis, or prosthetic ankle replace-

ment.¹² Papagelopoulos *et al.* analyzed the outcomes after distal fibulectomy for malignant bone tumors in 10 patients. They concluded that primary ankle arthrodesis achieved the most reliable result, therefore, it is preferred for adults. In children, repair of the lateral soft tissues and reconstruction of the tibiofibular mortise is necessary to avoid late ankle deformity or instability; and these patients may require a later arthrodesis.¹² In our case, the primary arthrodesis was performed after en bloc wide resection of the distal fibula.

Conclusions

Adamantinoma arising in the distal end of the fibula was successfully treated with en bloc wide resection and the primary arthrodesis of the ankle joint. A wide resection and adequate reconstruction of ankle can provide a good outcome for adamantinoma arising in the area.

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