Intracortical Osteosarcoma: Report of a Case

Shu-Hua Yang, Chen-Tu Wu, Chung-Jen Wang, Ming-Shin Kuo, and Rong-Sen Yang

Abstract: Intracortical osteosarcoma is the rarest anatomic variation of osteosarcoma. There have been only 12 cases reported in the English-language literature. We present a case of osteosarcoma in an 18-year-old Taiwanese man that originated within the cortex of the tibial diaphysis. The initial radiograph revealed a lytic mass confined to the cortex, mimicking a benign bone lesion. Histopathologic examination of the biopsy specimen showed an osteoblastic osteosarcoma mingled with some fibroblastic foci. He underwent en bloc resection, and a metallic prosthetic intercalary stem was used to replace the larger bone defect. Adjuvant chemotherapy was administered before and after the operation. He was free of disease during 40 months of follow-up. A review of all reported cases of intracortical osteosarcoma revealed that the initial method of treatment plays an important role in local recurrence and distant metastasis. Local excision and curettage leads to the worst results. The outcomes of more recently reported cases have improved because of early awareness of the possibility of malignancy and advances in chemotherapy. However, whether patients with intracortical osteosarcoma have a different prognosis from those with conventional osteosarcoma cannot be determined, because of the small number of intracortical osteosarcoma cases available for analysis.

Case Report

An 18-year-old Taiwanese man had sustained an open fracture of the right tibia during a traffic accident in 1985, when he was 8 years old. The fracture was treated with closed reduction, cast immobilization, and antibiotics. The fracture healed well and no specific problems were noted during the following 10 years. In June 1995, he suffered a contusion over the anterior aspect of the right lower leg as a result of a motorcycle accident. That minor trauma resulted in a small bulging mass and mild local tenderness over the contused area. The pain became aggravated and the mass enlarged progressively during the following 5 months. He sought medical help at a local hospital in early November 1995.

Physical examination revealed a 6 x 4 x 1-cm bulging mass over the anterior aspect of the middle third of his right tibia. Plain radiographs revealed a well-defined, radiolucent area confined to the cortex of the diaphysis of the right tibia (Fig. 1). Initially, he underwent intralesional curettage of the tumor, and the lesion was filled with bone cement. The inner cortex of the lesion had ruptured during the surgical procedure. Histopathologic examination of the excised specimen revealed areas of pleomorphic, hyperchromatic mesenchymal cells with malignant osteoid formation (Fig. 2), which was compatible with osteosarcoma. He was then referred to National Taiwan University Hospital in early December 1995.

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In this report, we present a patient with ICOS and also summarize previously reported cases, with special attention to the clinical presentations, radiologic and pathologic findings, methods of treatment, and final outcomes.
Fig. 1. Radiograph of the right tibia shows a well-defined, radiolucent area confined to the cortex of the diaphysis. The cortex, in conjunction with the normal area, is thickened and the inner cortex seems to be intact. Neither periosteal reaction nor medullary involvement is noted in this film.

The surgical procedure was delayed until April 1996, when an en bloc excision of the diaphysis and the tract of the previous biopsy surgery was performed. A 14-cm length of tibial shaft was resected and the section margin was at least 5 cm away from the lesion site. A metallic prosthetic intercalary stem was used to replace this defect (Fig. 3). The resected tibial diaphysis was sent for pathologic examination. On gross inspection of the specimen, the inner cortex of the tumor mass was ruptured. Bone cement had filled in the area of the lesion and penetrated into the medullary canal (Fig. 4). Neither tumor tissues (during gross inspection) nor malignant cells (during microscopic examination) were noted in any part of the specimen. After the surgical wound healed, the patient received three further courses of adjuvant chemotherapy. He recovered well after the operation and chemotherapy.

During 40 months of follow-up, there was no evidence of local recurrence or distant metastasis, and no evidence of loosening or subsidence noted around the implanted prosthesis on radiographs (Fig. 5). His functional outcome score evaluated using the Musculoskeletal Tumor Society Scoring System [13] was excellent.

ICOS is a distinct tumor entity from conventional osteosarcoma and is the rarest anatomic variation of osteosarcoma. It was first reported by Jaffe in 1960 [1]. The clinical characteristics, methods of treatment, and final outcomes of the 12 previously reported cases as well as the present case are listed in the Table.

In these 13 cases, tumor masses originated in the diaphysis of either the tibia (7 cases) or femur (6 cases). All tumors were confined to the cortex. The ages of these patients ranged from 10 to 43 years (mean, 21.8 yr).
Intracortical Osteosarcoma

Fig. 4. Surgical specimen (3.2 cm in length) bisected along the longitudinal axis. Gross inspection reveals a segment of the tibia with thickened cortex at the center. The inner cortex of the tumor mass is ruptured. Bone cement that was used to fill in the lesion area has penetrated into the medullary canal. There is no obvious tumor tissue around the bone cement.

There were eight male and four female patients; the sex of one patient was not reported. There was no side predominance in these patients. Interestingly, eight patients (cases 1, 5, 7, 9–13) reported a history of minor trauma to the lesion side months or years before presentation with the tumor. Most diagnoses were made within 1 month to 1 year after the occurrence of the trauma.

Imaging studies provide important information for diagnosis and treatment. The typical radiographic features included 1) a well-defined margin of the osteolytic lesion; 2) surrounding dense, thickened cortical bone with a smooth border; and 3) some patchy densities throughout the central portion of the thickened cortex [4]. The sizes of the tumors ranged from 1 to 7 cm (mean, 3.5 cm) on radiographs at the time of diagnosis. Early findings during radiographic examination simulated a variety of benign conditions such as fibrous cortical defects, solitary focus of osteomyelitis (Brodies abscess), adamantinoma, osteoid osteoma, and osteoblastoma [8, 12]. Furthermore, the typical findings on computed tomography (CT) scans included marked eccentric thickening of the cortex, a lytic area within the area of cortical thickening, and minimal or no involvement of the medullary canal [10, 14].

The histologic findings of ICOS included well-defined osteoblastic lesions. The stromal cells showed some pleomorphism, a moderate mitotic rate, occasional atypical mitoses, and the epithelial appearance of osteoblasts [9]. Including the present case, results from microscopic examination of specimens were described in 12 cases. Seven patients (cases 1, 5–7, 9, 11, 13) had spindle-cell fibroblastic or fibrosarcomatous foci. Malignant chondroblastic foci were found in three patients (cases 1, 5, 6), and multinucleated osteoclast-type cells in four patients (cases 3, 5, 7, 10).

Review of the literature suggests that the initial method of treatment played an important role in patient prognosis. Local recurrence and metastasis tended to occur in patients who were initially treated with local excision and curettage, without awareness of malignancy. Five patients had local recurrence (cases 1–4, 6), four of whom were initially treated with local excision and curettage. The possibility of malignancy may have been overlooked and/or the surrounding tissue might have been contaminated during the first operation in these patients, two of whom later died of widespread metastasis (cases 2, 6). One patient died of another type of cancer 26 years and 5 months after the initial treatment for ICOS (case 1).

The oncologic outcomes of more recently reported cases have improved as a result of early awareness of the possibility of malignancy and advances in chemotherapy. In our patient, complete necrosis of the tumor was achieved after chemotherapy. However, the prognosis for patients with ICOS is still uncertain. It cannot be determined if there are any prognostic differences between patients with these lesions and those with conventional osteosarcomas, because only a small number of cases have been reported.

Another challenge in treating these patients is the replacement of the large bone defect after en bloc resection. In previous reports, Kyriakos et al used banked allograft [11]; Anderson et al used ipsilateral fibular strut autograft for the tibial lesion [8]; and Lopez-Barea et al used contralateral fibular strut autograft for the femoral lesion [9]. However, there are other options to replace the defect, such as vascular-
## Table: Clinical characteristics, treatment, and outcome in 13 reported cases of intracortical osteosarcoma

<table>
<thead>
<tr>
<th>Case/ Author</th>
<th>Age/ Sex</th>
<th>Site*</th>
<th>Therapy</th>
<th>Course</th>
<th>Follow-up observation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/ Jaffe [1]</td>
<td>14/M</td>
<td>Left tibia</td>
<td>En bloc resection; R/T (Oct ‘59)</td>
<td>LR, A-K amputation (May ‘60); curettage, osteoid osteoma left humerus (Oct ‘74)</td>
<td>Metastatic angiosarcoma to bones, lung, and liver (Nov. ‘84); dead (T) 26 yr 5 mo after diagnosis (Mar ‘86) Dead (T) with widespread metastasis (May ‘61)</td>
</tr>
<tr>
<td>2/ Jaffe [1]</td>
<td>25/F</td>
<td>Left femur</td>
<td>Bx; R/T; hip disarticulation (May ‘60)</td>
<td>Skin metastasis (Feb ‘61)</td>
<td></td>
</tr>
<tr>
<td>3/ Scranton et al [2]</td>
<td>21/F</td>
<td>Left femur</td>
<td>Local excision (Feb ‘59)</td>
<td>LR, en bloc resection ’59, ’60; femoral shaft resection (Dec. ’60); LR bone and soft tissue,’62, ’64, ’67, ’70; locally excised</td>
<td>Alive and well (Nov ‘89)</td>
</tr>
<tr>
<td>4/ Lichtenstein [3]</td>
<td>NS</td>
<td>Tibia</td>
<td>Excision (type not stated)</td>
<td>LR 6 months</td>
<td></td>
</tr>
<tr>
<td>5/ Kyriakos [4]</td>
<td>24/M</td>
<td>Left tibia</td>
<td>En bloc resection; knee disarticulation; C/T (Mar ’78)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6/ Picci et al [5]</td>
<td>30/M</td>
<td>Right femur</td>
<td>Bx and curettage (July ‘81)</td>
<td>LR 2 months; hip disarticulation; C/T (Oct ‘81); bilateral lung metastasis (Dec ’82)</td>
<td>Dead (T) (Nov ‘83)</td>
</tr>
<tr>
<td>8/ Schwinn</td>
<td>27/F</td>
<td>Right femur</td>
<td>Bx; en bloc resection; C/T (Jul ’83)</td>
<td>No recurrence</td>
<td>Alive and well 3.5 years after diagnosis</td>
</tr>
<tr>
<td>9/ Anderson et al [8]</td>
<td>10/F</td>
<td>Left tibia</td>
<td>Bx; en bloc resection (Jun ‘85)</td>
<td>No recurrence</td>
<td>Alive and well (Oct ‘89)</td>
</tr>
<tr>
<td>10/ Lopez-Barea et al [9]</td>
<td>19/M</td>
<td>Right femur</td>
<td>Bx (Jul ‘88); en bloc resection (Nov ‘88); C/T (Oct ‘91)</td>
<td>No recurrence</td>
<td>Alive and well (May ‘91)</td>
</tr>
<tr>
<td>11/ Mirra et al [10]</td>
<td>43/M</td>
<td>Right femur</td>
<td>Bx (Jun ‘89); wide resection of femur (Aug ‘89)</td>
<td>No recurrence</td>
<td>Alive and well (May ‘91)</td>
</tr>
<tr>
<td>12/ Kyriakos et al [11]</td>
<td>15/M</td>
<td>Right femur</td>
<td>Needle Bx; en bloc resection (Apr ‘87); C/T</td>
<td>No recurrence</td>
<td>Alive and well (Jan ‘92)</td>
</tr>
<tr>
<td>13/ Yang et al (this case)</td>
<td>19/M</td>
<td>Right tibia</td>
<td>Excision; Bx (Nov ‘95); C/T; en bloc resection (Apr ‘96)</td>
<td>No recurrence</td>
<td>Alive and well (Aug ‘99)</td>
</tr>
</tbody>
</table>

*All in the diaphysis; †probable case; ‡ summarized by Mirra [7]; R/T = radiotherapy; LR = local recurrence; A-K = above knee; Bx = biopsy; C/T = chemotherapy; Dead (T) = died of tumor; NS = not stated.
ized fibular graft [15, 16], and gradual lengthening of the remaining bone using the Ilizarov method [17, 18]. We used a metallic prosthetic intercalary stem in our patient and achieved a very good result. There was no significant subsidence or loosening that could be detected on radiographs, and the involved limb functioned very well during the follow-up period. This new method for the replacement of the shaft of long bones has the advantage of not sacrificing other intact structures.

In conclusion, early detection of ICOS is crucial for a good prognosis. Even if the lesion is confined to the cortex of the diaphysis and looks benign on imaging studies, ICOS should be considered as a differential diagnosis. Proper initial treatment plays the most important role in local recurrence and distant metastasis, and should include en bloc resection of the tumor and adjuvant chemotherapy. This type of management has achieved favorable results in more recently reported cases. However, because data from only a small number of cases are currently available, whether prognostic differences exist between ICOS and conventional osteosarcoma is difficult to determine.

References