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Intraosseous neurilemmoma of the proximal ulna

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ABSTRACT

INTRODUCTION: Neurilemmoma is a benign nerve sheath neoplasm commonly located in the soft tissue. Intraosseous neurilemmoma is rare, constituting less than 1% of primary bone tumors.
PRESENTATION OF CASE: A 21 year-old woman was presented with left elbow pain of 1-month duration. Plain radiographs showed a well-defined, lytic and expansile lesion of the proximal ulna. Computed tomography revealed cortical destruction and soft tissue extension. Because the tissue of origin for the tumor was uncertain, an open biopsy was performed. The specimens demonstrated a benign spindle cell tumor suggestive of a neurilemmoma, similar to a soft tissue neurilemmoma. The diagnosis of intraosseous neurilemmoma was established. Marginal excision of the soft tissue component and curettage of the lesion in the bone were performed. After 3.5 years of follow up, there is no clinical or radiographic finding to suggest any recurrence.
DISCUSSION: The major site of intraosseous neurilemmoma is the mandible. Occurrence in the long bone is particularly rare. Only two cases of intraosseous neurilemmoma involving the bones around the elbow have been reported to our knowledge; these cases arose in the distal humerus. We describe the first case of intraosseous neurilemmoma of the proximal ulna of the left elbow. The recommended treatment is conservative resection and bone grafting, as malignant change is extremely rare.
CONCLUSION: Although very rare, intraosseous neurilemmoma should be taken under consideration in the differential diagnosis of painful, radiographically benign-appearing osseous tumor around the elbow.

1. Introduction

Neurilemmoma (schwannoma) is a benign neoplasm that arises from the myelinating schwann cells of the nerve sheath.[1–3] It is a relatively common tumor, comprising approximately 5% of all benign soft tissue tumors, and shows a predilection to affect the sensory nerves.[3] Intraosseous neurilemmoma, however, is very rare, accounting for less than 1% of benign bone tumors.[4,5] It shows no sex-, race-, or age-dependant predilections.[6] The mandible is the most frequently affected site, followed by the sacrum; this tumor rarely arises in the bones of the extremities.[5,7,8] Only 2 cases of intraosseous neurilemmoma around the elbow have been described in literature[6,9]; these cases arose in the distal humerus. We present the first case report of an intraosseous neurilemmoma affecting the proximal ulna.

2. Presentation of case

A 21-year-old woman was referred to our orthopedic unit with a 1-month history of pain in the left elbow. The initiating factor, such as trauma, was not clear. On physical examination, the overlying skin was intact, and there was no evidence of warmth, erythema, or induration. No other masses were palpable. The elbows and forearms had a normal range of motion (ROM). No evidence of lymphadenopathy or neurovascular involvement was found.

Plain radiographs revealed a well-defined, lytic, and expansile lesion, with thin marginal sclerosis and trabeculation in the proximal ulna (Fig. 1). Computed tomography (CT) showed considerable destruction of the cortex of the ulna, which connected with a mass of soft tissue. At the edge of the destructed cortex overhung a soft tissue mass (Fig. 2a), which had invaded into the cortex, and no periosteal reaction was seen (Fig. 2b). Magnetic resonance imaging (MRI) showed the well-defined and lobulated lesion to be isointense to skeletal muscle on T1-weighted images, and heterogeneous and hyperintense on T2-weighted images. The lesion revealed uniform enhancement following Gd-DTPA administration.

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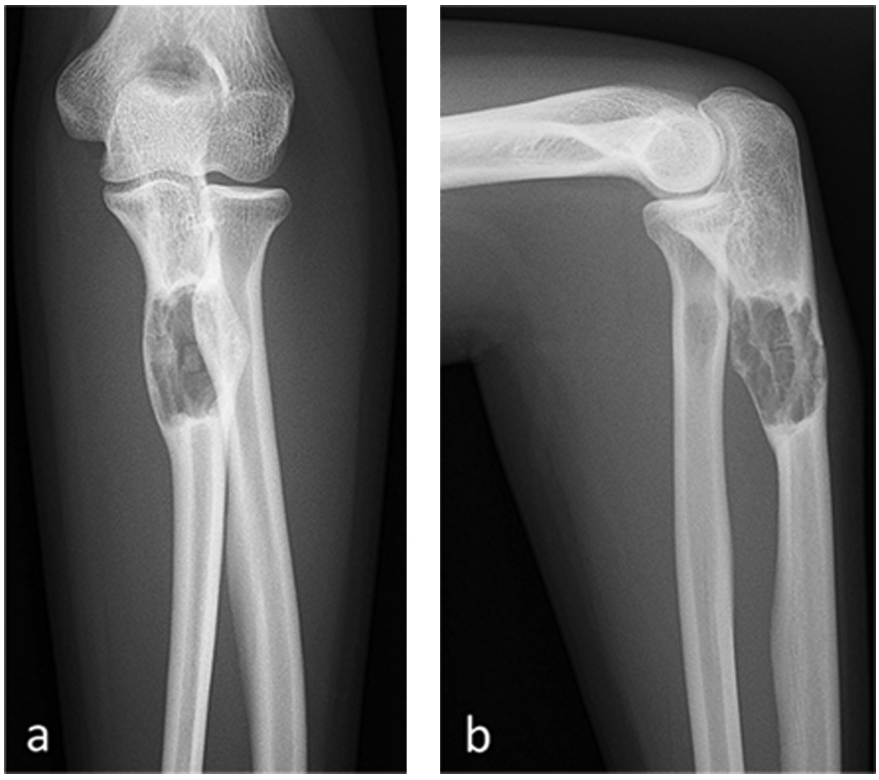

Fig. 1. Anteroposterior (a) and lateral (b) plain radiographs, showing a well-defined, lytic, and expansile lesion, with marginal thin sclerosis and trabeculation in the proximal ulna.

Because of uncertainty regarding the histological origin of the tumor, we performed an open biopsy. The pathologic specimen consisted of hypercellular regions with palisading nuclei (Antoni type A), and hypocellularity regions with myxoid background (Antoni type B). Almost all of the area was Antoni type A (Fig. 3). We could not observe nuclear atypia, necrosis, or mitosis. Immunohistochemical studies showed strong, diffuse reactivity for S-100 protein within the lesional cells. Microscopic findings were consistent with a diagnosis of neurilemmoma.

The diagnosis of intraosseous neurilemmoma was established. We surgically excised the soft tissue component of the mass by marginal resection. Curettage was performed on the bone lesion, and the resulting deficit was grafted with beta-tricalcium

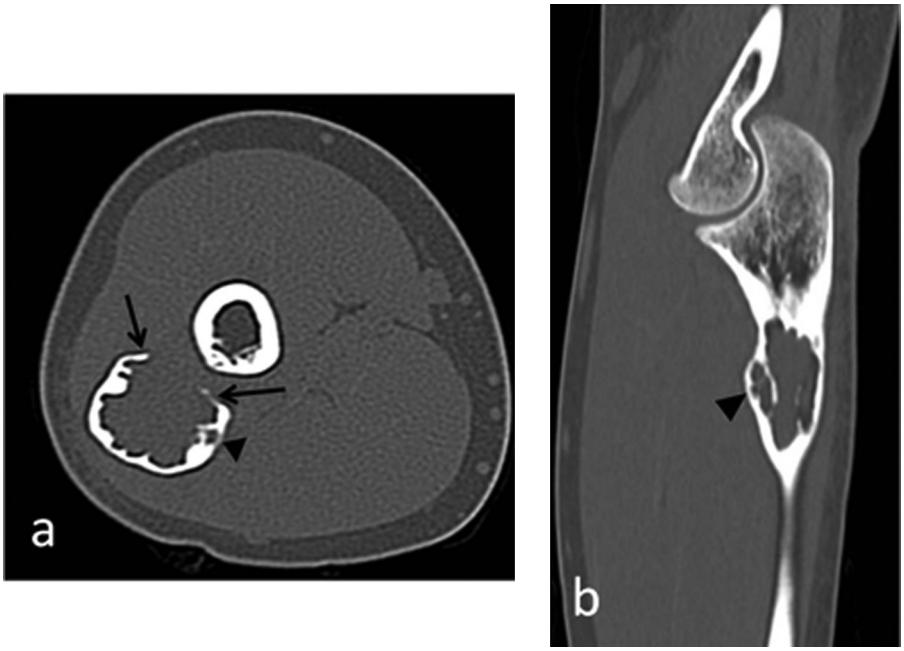

Fig. 2. Axial computed tomography scan (a) and sagittal reconstruction (b) of the ulna, demonstrating destruction of the cortex of the ulna, with extension into the adjacent soft tissue. At the edge of the destructed cortex overhung a soft tissue mass (arrows), which had invaded into the cortex (arrowhead).

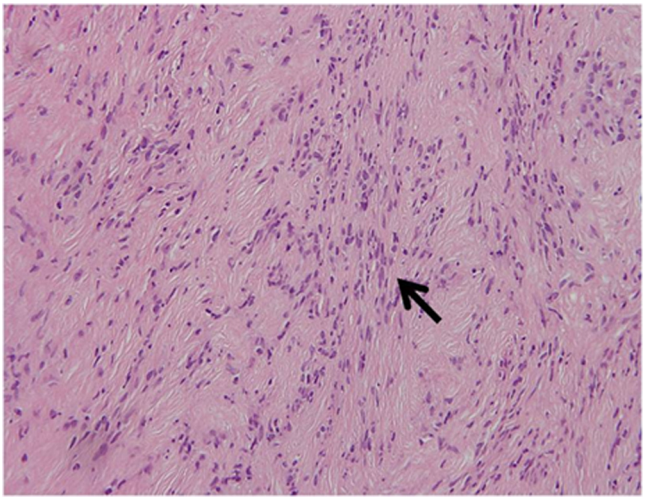

Fig. 3. Photomicrograph of the tumor specimen, showing the highly cellular Antoni type A pattern of growth. Nuclear palisading is noted (arrows) (hematoxylin–eosin staining; magnification, 200×).

phosphate (Fig. 4). The pathologic tissue that was removed was yellow-to-brownish. After 3.5 years of follow up, the patient is clinically asymptomatic, and the radiographs show complete graft incorporation (Fig. 5). There is no clinical or radiographic finding to suggest any recurrence.

The patient was informed that the case would be submitted for publication and her consent was obtained.

3. Discussion

We present the case of an intraosseous neurilemmoma affecting the proximal ulna. This tumor rarely arises in the bones of the extremities.[5,7,8] Only two cases have previously been reported around the elbow. These cases arose in the distal humerus.[6,9] This is the first report of its origin in the proximal ulna.

Neurilemmomas may involve bone tissue via 3 mechanisms: (1) they may be intramedullary, producing rarefaction of the bone; (2) they may be located within the nutrient canal, with the formation of a dumbbell-shaped tumor; or (3) they may be extraosseous, eroding into the bone.[5,7,10] With regard to the comparatively high incidence of intraosseous neurilemmoma in the mandible, it has been speculated that the long intraosseous path of the mandibular nerve may predispose to metaplasia of the schwann cells in its nerve sheath.[5] However, some authors have refuted this theory, noting that the nerves innervating the long bones through the nutrient foramina are longer than the mandibular nerve; nevertheless, the frequency of intraosseous neurilemmoma in the long bones is several times lower than that in the mandible.[7,8] Neurilemmoma shows a predilection to the myelinated nerves, especially the sensory nerves. However, most intraosseous nerves are non-myelinated and participate in vasomotor functions.[11] de la Monte and colleagues have suggested that the death of sensory nerve fibers within bone tissue may account for the exceedingly rare occurrence of intraosseous neurilemmoma.[8] We think that the high frequency of neurilemmoma in the mandible is not because of the long intraosseous course, but because the mandibular nerve consists of sensory nerves of the trigeminal nerve origin.

The characteristic radiographic features of intraosseous neurilemmoma include the following: (1) a well-defined lytic

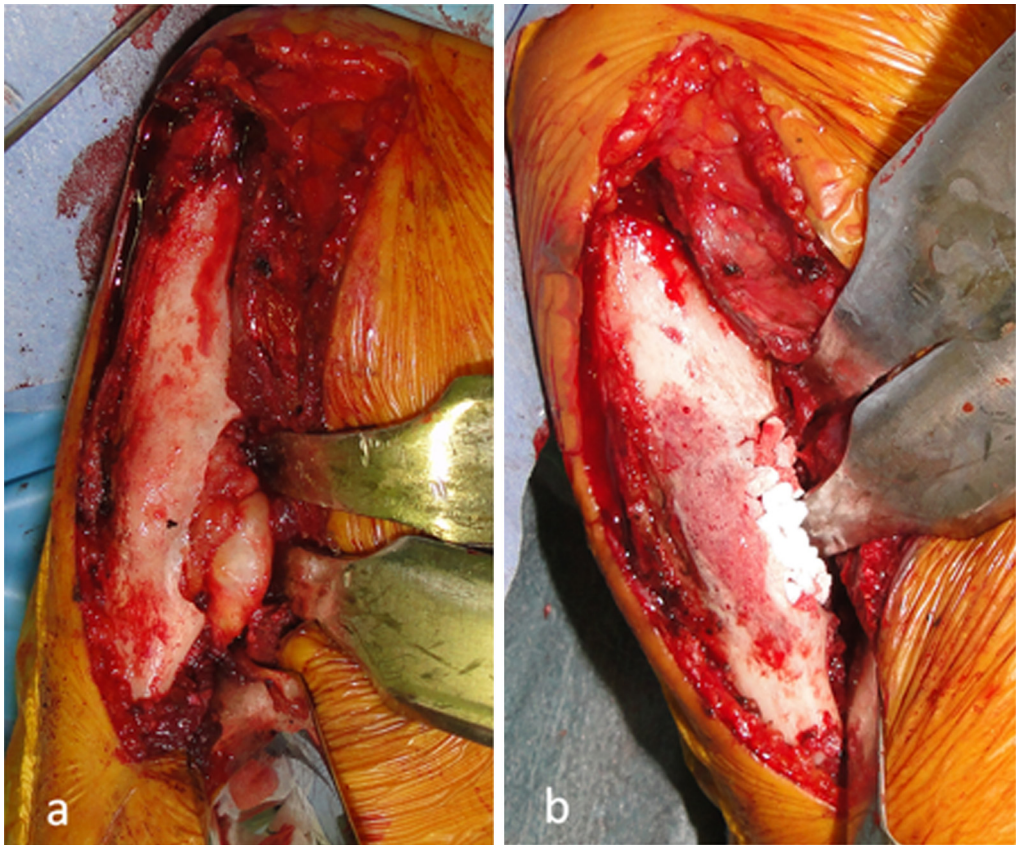

Fig. 4. (a) Intraoperative photograph shows that the tumor destructed the cortex of the ulna. (b) Curettage was performed on the bone lesion, and the defect was filled with beta-tricalcium phosphate.

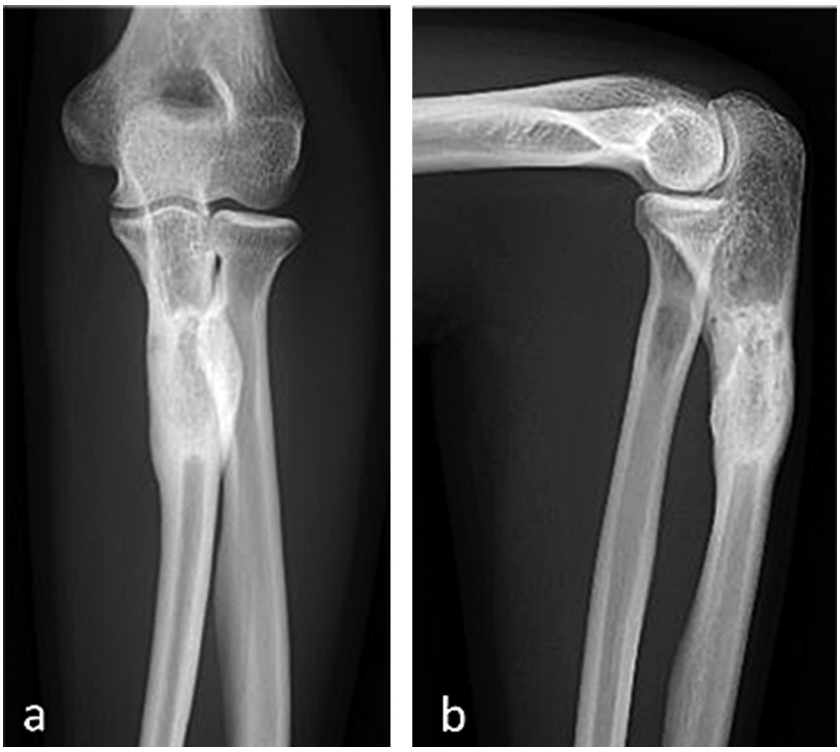

Fig. 5. Anteroposterior (a) and lateral (b) plain radiographs after 3.5 years of surgery, showing complete graft incorporation and no recurrence.

lesion, (2) sclerotic margins, (3) lobulated or trabeculated contours, (4) cortical expansion, and (5) absence of central calcification.[7–9] However, the radiographic findings are non-specific. It is difficult to differentiate intraosseous neurilemmoma from other benign neoplasms of such osseous lesions, including solitary bone cyst, aneurysmal bone cyst, giant cell tumor, non-ossifying fibroma, benign fibrous histiocytoma, desmoplastic fibroma, fibrous dysplasia, chondromyxoid fibroma, and enchondroma. On CT, we can confirm the origin of the tumor when there is soft tissue extension of the tumor. If the origin is intraosseous instead of periosteal, no periosteal reaction is seen, and the edge of the destructed cortex will be overhung by a soft tissue mass. Young and colleagues pointed out that the aspect of the overhanging edges of cortical bone can assist in distinguishing between an intraosseous desmoplastic fibroma versus a periosteal desmoplastic fibroma.[12] Similarly, intraosseous neurilemmoma is considered to originate from intraosseous nerves rather than from periosteal nerves, if CT shows overhanging edges of cortical bone. In our case and in others,[13–15] CT showed that the tumor had invaded into the cortex. This may be the characteristic finding of intraosseous neurilemmoma provided the tumor expands within the nutrient canal of the cortex.

The final diagnosis of intraosseous neurilemmoma was not made until after histologic examination of tissue obtained during excision. The histologic features of intraosseous neurilemmoma are similar to those of soft tissue neurilemmoma. Microscopically, neurilemmoma has identifiable Antoni A and Antoni B regions. The diffuse immunoreactivity for S-100 protein is indicative of a Schwann cell origin, and helps distinguish this tumor from other benign spindle cell lesions of similar histology.

The recommended treatment for intraosseous neurilemmoma is conservative resection and bone grafting, as malignant change is extremely rare. Recurrence also is rare following complete local excision. In previous reports, after performing only curettage, curettage and bone grafting, or en block resection, local recurrence only occurred in 2 cases.[16,17] These authors did not refer to the reason for local reference, but recurrence of the tumor is not evidence of malignant transformation. In our case, there was no evidence of recurrence.

4. Conclusion

Owing to the infrequency of an intraosseous neurilemmoma, the diagnosis is often not even suspected until histologic evaluation after a biopsy. Although very rare, intraosseous neurilemmoma should be taken under consideration in the differential diagnosis of painful, radiographically benign-appearing osseous tumor around the elbow.

Conflict of interest

The authors declare that they have no conflict of interest.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Case report writing, data collection, and discussion writing were done by Munehisa Kito and Yasuo Yoshimura. Discussion writing was carried by Ken'ichi Isobe, Kaoru Aoki, Takashige Momose, and Hiroyuki Kato.

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