Case Report

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First-ever intraosseous ancient schwannoma of the proximal ulna successfully treated using the cement technique

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Abstract

Schwannoma or neurilemmoma is a common soft tissue neoplasm arising from the neural sheath of Schwann cells. However, intraosseous schwannoma is rare, accounting for less than 0.2% of primary bone tumours. Several variants of schwannoma have been reported; among them, intraosseous schwannoma with ancient change is extremely rare. This current report presents an extremely rare case of ancient intraosseous neurilemmoma. The patient presented with right elbow pain and disability. A radiolucent, well-defined, lobulated lesion with a thin sclerotic rim in the proximal ulnar metaphysis that had caused a pathological fracture was noted. The mass was surgically excised using marginal resection and bone curettage was undertaken. The bone deficit was grafted with hydroxyapatite and β-tricalcium phosphate and augmented with bone cement. There were no signs of any recurrence after 3 years. This is the first case of an ancient intraosseous schwannoma of the proximal ulna. Although rare, intraosseous schwannoma should be considered in the differential diagnosis of radiographically benign-appearing osseous tumours in the bone. The cement technique is recommended for the treatment of intraosseous schwannoma.

Keywords

Ancient type, cement technique, intraosseous schwannoma, ulna

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Introduction

Schwannomas or neurilemmomas are common benign nerve sheath neoplasms derived from Schwann cells and are often located in the soft tissue. However,

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intraosseous schwannomas are rare, accounting for less than 0.2% of primary bone tumours.[1] Intraosseous schwannomas are usually observed in the mandible, maxilla, spine and sacrum; involvement of the long bones is particularly rare.[1]

Based on their histological characteristics, schwannomas can be classified into several variants, including classical, cellular, ancient, epithelioid, melanotic and plexiform.[2] Among them, the ancient type is extremely uncommon and intraosseous schwannoma with ancient change is even rarer. An ancient intraosseous schwannoma is typically a slow-growing, degenerative lesion that tends to grow to a large size and is commonly observed in the elderly.[3]

This current report presents an extremely rare case of ancient intraosseous neurilemmoma of the proximal ulna that caused a pathological fracture. Surgical resection, curettage and bone grafting have been widely used to treat intraosseous schwannoma.[1] However, recurrence has been noted in cases with incomplete resection and early detection is difficult with current treatment methods.[4] This current case was treated with the cement filling technique as part of the traditional treatment method of intraosseous schwannoma and the result was satisfactory.

Case report

A 77-year-old woman was brought to the emergency department of Far Eastern Memorial Hospital, New Taipei City in June 2017 due to right elbow pain and disability after falling from a standing height. Upon arrival, she was conscious and haemodynamically stable. On physical examination, her right elbow was moderately swollen and tender when palpated and she reported a visual analogue scale pain score of 7/10. No local heat, erythema or palpable mass was observed, and the overlying skin was intact. Her distal pulse was palpable and her sensory response was symmetrical.

Radiography indicated a right ulnar olecranon fracture. Additionally, a radiolucent, well-defined, lobulated lesion with a thin sclerotic rim was accidentally observed at the proximal ulnar metaphysis. No significant calcification or periosteal reaction was noted (Figure 1). Magnetic resonance imaging showed a $7.8 \times 4.4 \times 3.6$ cm expansile intraosseous mass invading into the adjacent muscle and subcutaneous fat tissue (Figure 2). She was diagnosed with a right ulnar olecranon pathological fracture and the initial differential diagnosis of the lesion included mesenchymal soft tissue neoplasm (e.g. low-grade sarcoma), giant cell tumour at an atypical age or low-grade intraosseous lymphoma.

An open incisional biopsy was performed and a pathological review of the specimen indicated spindle cells in fascicles with focal nuclear palisading. A special arrangement of spindle cells formed Antoni A and Antoni B tissue patterns. There was a diffuse strong positive immunoreactivity for S-100 protein. Based on these findings, intraosseous schwannoma was diagnosed. Further, degenerative change in the thrombosed vessels, cystic change and nuclear atypia without mitotic activity supported the diagnosis of the ancient histopathological variant (Figure 3).

The mass was surgically excised by marginal resection and a bone curettage was undertaken. The bone deficit was grafted with hydroxyapatite and β-tricalcium phosphate and augmented with bone cement. The ulna was fixed with a locking plate. The right elbow was immobilized with a long arm splint for 2 weeks, after which the patient underwent early active range of motions.

Follow-up radiography indicated the union of the fracture (Figure 4) and the Mayo Elbow Performance Score was 100 after 1 year. There were no clinical or

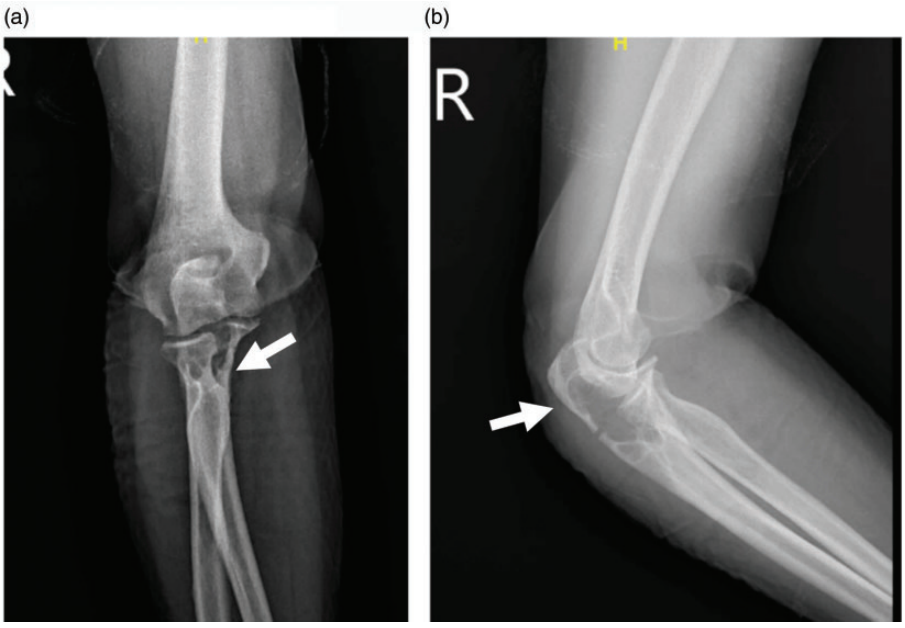

Figure 1. (a) Anteroposterior radiography view of the right elbow. (b) Lateral radiography view of the right elbow. Right elbow radiography images showing a pathological fracture (arrows) of the right olecranon. A lobulated, well-defined radiolucent lesion is observed at the proximal ulnar metaphysis.

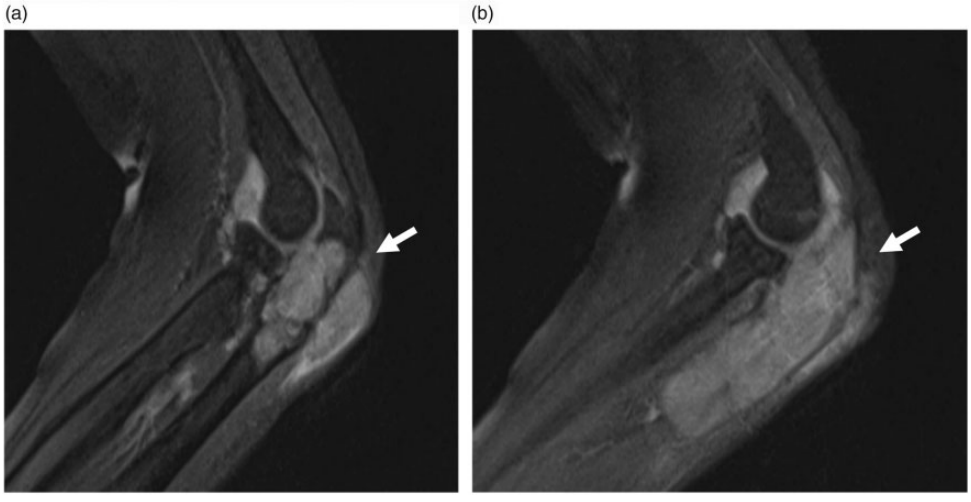

Figure 2. Sagittal proton density, turbo spin-echo, fat suppression sequence magnetic resonance imaging of the right elbow showing a 7.8 × 4.4 × 3.6 cm expansile intraosseous mass (arrows) invading into the adjacent muscle and subcutaneous fat tissue.

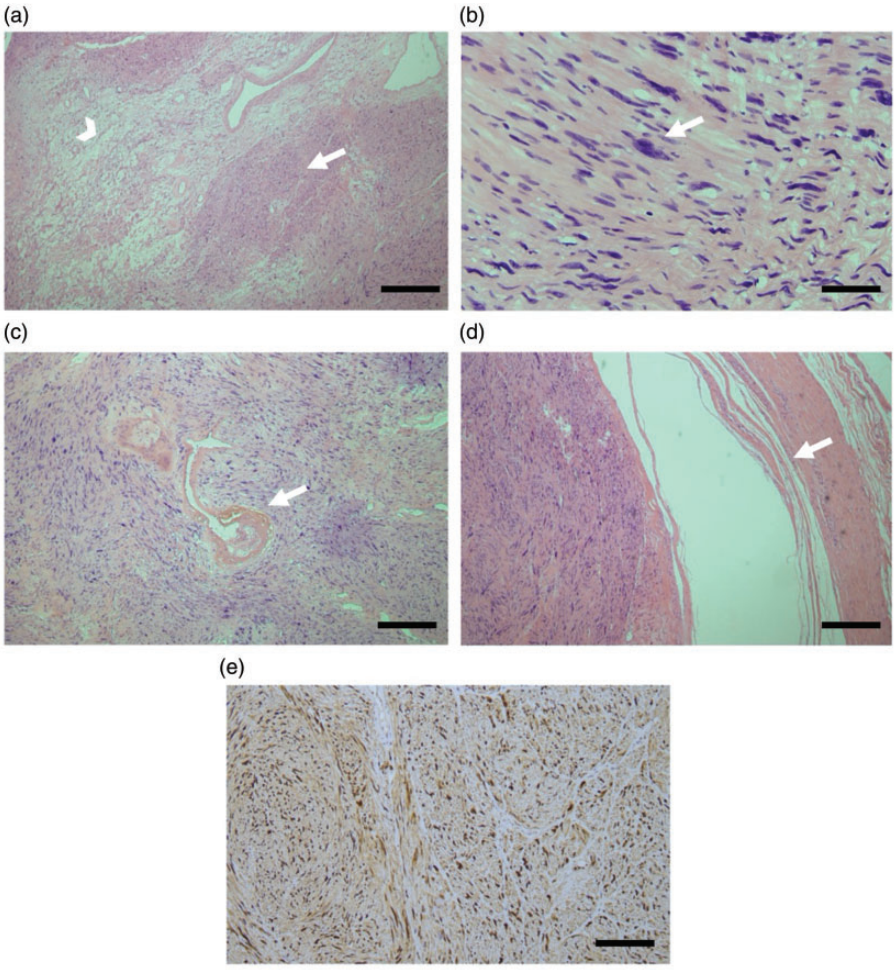

Figure 3. Pathological and immunohistochemical analysis of the biopsy specimen. (a) Haematoxylin & eosin (H&E) staining of Antoni A (arrow) and Antoni B (arrowhead) tissue patterns (scale bar 200 μm). (b) H&E staining of spindle cells (arrow) with focal nuclear palisading and nuclear atypia (scale bar 50 μm). (c) H&E staining showing thrombosed vessels (arrow) (scale bar 200 μm). (d) H&E staining showing cystic degenerative change (arrow) (scale bar 50 μm). (e) Diffuse strong positive immunoreactivity for S-100 (scale bar 200 μm). The colour version of this figure is available at: http://imr.sagepub.com.

radiographic findings to suggest any recurrence at the 3-year follow-up.

Written informed consent was obtained from the patient and she clearly understood that her data and information were being collected for publication. The Research Ethics Review Committee of Far Eastern Memorial Hospital waives the requirement for ethical approval for case reports.

Discussion

This current report presented the case of a 77-year-old woman with intraosseous

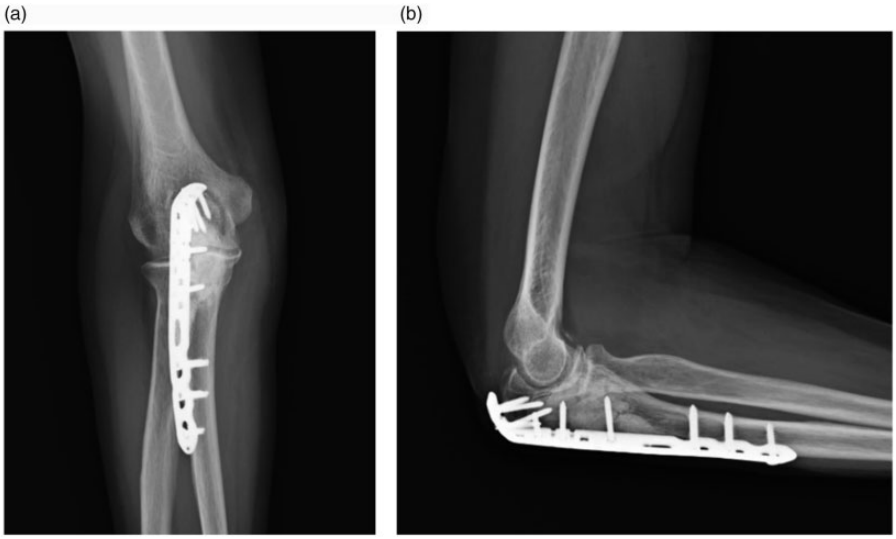

Figure 4. (a) Anteroposterior radiography view of the right elbow. (b) Lateral radiography view of the right elbow. Radiography images at 38 months post-operation showing union of the fracture without absorption of the bone graft.

schwannoma located at the metaphysis of the proximal ulna, which caused a pathological fracture. In addition to tumour excision, curettage and bone substitute grafting, the cement filling technique was also used to treat this current patient with a satisfactory outcome.

The occurrence of intraosseous schwannoma in the long bones is particularly rare; only two cases of intraosseous schwannoma of the ulna have been published.[1,5] To the best of our knowledge, this current report may be the third case of intraosseous schwannoma located in the ulnar bone.

Intraosseous schwannomas seem to be more predominant in females than in males in a ratio of 2:1.[6] Likewise, they have histological characteristics similar to those of soft-tissue schwannomas, including Antoni type A areas, formed by compactly arranged palisaded spindle cells; and Antoni type B areas, which are disorganized hypocellular areas.[2] There are several subtypes of schwannoma based on histopathological features, including classical, cellular, ancient, epithelioid, melanotic and plexiform.[2] The first ancient variant of schwannoma was described by Ackerman and Taylor in 1951.[7]Ancient schwannomas are relatively rare compared with other variants and fewer of them are intraosseous lesions.[8] Nuclear atypia, calcification, hyalinization, haemorrhage, cystic degeneration and haemosiderin deposition may be observed in ancient schwannomas.[9] These degenerative changes result from the long duration of tumour growth. Degenerative changes, including nuclear pleomorphism, cystic degeneration and thrombosed vessels, were observed in this current case, suggesting the diagnosis of ancient intraosseous schwannoma. This may be the first case of ancient intraosseous schwannoma observed in the long bones.

The most common symptoms of intraosseous schwannoma are pain and swelling, but some are asymptomatic.[9] They rarely cause bone fracture.[10] The ancient

intraosseous schwannoma in this current patient had special characteristics. Ancient schwannomas are slow-growing lesions and tend to grow to a large size over a long period.[3] Due to their distinctive features, they can grow gradually without eliciting any symptoms until they are large enough to destroy bony structures, as in this current case.

The radiological findings of intraosseous schwannoma are not always specific, but they include well-defined radiolucent lesions, sclerotic rims, lobulated or trabeculated contours, cortical expansion and a lack of internal calcification.[11] It is difficult for clinicians to differentiate intraosseous schwannoma from other benign osseous lesions.

The prognosis of intraosseous schwannoma is good because recurrence is uncommon and malignant change is extremely rare.[1] The recommended treatment of intraosseous schwannoma based on previous studies is surgical resection, curettage and bone grafting.[1] A previous report observed no recurrence in patients with complete tumour excision.[12] However, a recurrence rate of approximately 16% was observed in patients with incomplete tumour excision.[12] The current case underwent marginal resection, bone curettage and bone substitute grafting, followed by cementing with polymethyl methacrylate and fixation of the ulnar bone.

Complete resection and thorough curettage should be performed at any cost to minimize the recurrence rate. Autograft, especially iliac crest autograft, is most widely used for bone grafting, but it can be associated with significant morbidity.[13] Minor complications, including superficial infection and haematoma to major complications, including deep infection, vascular injury, iliac wing fracture and abdominal herniation, have been reported.[14] Moreover, it is difficult to harvest bone graft from the osteoporotic bone. Therefore, in this current case, most of the bone void and destructed bone cortex were filled with hydroxyapatite and β-tricalcium phosphate and augmented with cement. This is because the lesion was located at the metaphyseal area, which contains the trabecular bone and has a rich blood supply. Hydroxyapatite and β-tricalcium phosphate are osteoconductive materials that provide a sufficiently conducive environment for new bone formation while avoiding harvest site complications.

The cementation technique is strongly recommended because it has three major advantages. First, it improves dynamic stability and mechanical strength, which allow for a short period of immobilization and early range of motion, especially in patients with osteoporotic bones, pathological fractures or large bone windows created to allow complete resection of the intraosseous lesion. Secondly, it allows early detection of tumour recurrence.[15] The cement forms a dense radiopaque mass and the appearance of a progressive enlarged radiolucent lesion within the cement or between the cement–bone border generally indicates the recurrence of the tumour. This early identification allows for early management. Thirdly, the hyperthermia effect from the polymerization of the cement provides an antineoplastic effect on any residual tumour cells. Therefore, the use of the cement technique is recommended for patients with intraosseous schwannoma. Further studies need to be conducted to provide stronger evidence.

For the timing of cementation and fixation of the screws, it is best to place the screws in doughy cement as its pullout force is supposedly higher than when the screws are placed in hard or soft cement.[16] However, it is a race against time to place the screws in such a situation. In this current case, the screws were set when the cement was hard. Drilling and pre-tapping were performed before screw fixation to avoid cracking the bone cement.

In conclusion, this is the first case of ancient intraosseous schwannoma of the proximal ulna. Although very rare, intraosseous schwannoma should be considered in the differential diagnosis of radiographically benign-appearing osseous tumours in the bone. The cement technique was used in the treatment of this patient as it provided several advantages with no significant complications.

Declaration of conflicting interest

The authors declare that there are no conflicts of interest.

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