Intraosseous Schwannoma of the Second Metacarpal: Case Report

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Osseous involvement in schwannoma (neurilemmoma) is rare. This report presents a case of intraosseous schwannoma in a child. The lesion was located in the second metacarpal of his right hand and had a soft tissue extension into the first web space. (*J Hand Surg 2010;35A: 776–779. Copyright* © 2010 by the American Society for Surgery of the Hand. All rights reserved.)

Key words Intraosseous schwannoma, intraosseous neurilemmoma, hand tumor.

S CHWANNOMA OR NEURILEMMOMA that originates from the myelinating schwann cells of the nerve sheath is a benign tumor.¹ Osseous involvement in schwannoma is rare because schwannomas are usually located in the soft tissue. Intraosseous schwannomas most commonly involve the mandible.^{2,3} Only 16 cases of intraosseous schwannomas have been reported in the upper extremity.^{1,4–11} To our knowledge, 6 were in small bones (metacarpals and phalanges) of the hand.^{1,6–11} Because of the rarity of the lesion, preoperative diagnosis of the intraosseous schwannoma is difficult, if not impossible. We were able to find only one report that described an intraosseous schwannoma in a child. The lesion was located in the fibula.¹²

This report presents a case of intraosseous schwannoma in a child, involving the second metacarpal of his right hand. The tumor had a large soft tissue component that extended into the first web space.

CASE REPORT

A 12-year-old boy presented with a painless tumor in the first web space of his right hand. He first noticed the slow-growing mass 18 months before seeking medical

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0363-5023/10/35A05-0012\$36.00/0 doi:10.1016/j.jhsa.2010.02.006 treatment. The neurovascular structures were intact. There was no involvement of the digital nerves. The tendon functions of the right hand were normal. X-rays showed a well-defined lytic and expansile lesion with cortical disruption at the base of second metacarpal (Fig. 1). Slow-growing benign tumors such as aneurys-mal bone cyst and enchondroma were considered in the differential diagnosis.

Magnetic resonance imaging (MRI) showed that the tumor originated in the bone. Cortical disruption and bone marrow involvement at the base of the second metacarpal were noted. There was a large, well-defined, heterogeneous (iso-hypointense and hyperintense to adjacent muscle) lesion on T1-weighted images in the soft tissue of the volar aspect of the second metacarpal bone (Fig. 2). The lesion was hyperintense on T2-weighted images (fat saturation) and proton density–weighted images.

Because of uncertainty about the tissue origin of the tumor, we performed an incisional biopsy through the dorsum of the first web. At surgery, there was white tissue with firm consistency, measuring about $3 \times 3 \times 3$ in the first web space.

On microscopic study, sections revealed encapsulated tumoral tissue composed of spindle cells arranged in short bundles or interlacing fascicles in hypercellular areas, which was Antoni A–type tissue. Edematous, hypocellular areas known as Antoni B–type tissue were also seen (Fig. 3.) Hyalinization, hemorrhage, thickened wall blood vessels, and a few giant cells were also seen. We did not note nuclear atypia, necrosis, or mitosis. Immunostaining for the S-100 antigen was diffusely positive (Fig. 4).

No benefits in any form have been received or will received related directly or indirectly to the subject of this article.



FIGURE 1: Anteroposterior x-ray of the right hand depicting an osteolytic expansile lesion.

Microscopic findings of schwannoma are specific. Schwannoma depicts 2 types of cell arrangement; Antoni A and Antoni B. The Antoni A pattern is composed of compact spindle cells arranged in bundles and cords. The nuclei of the Antoni A cells are arranged in palisade rows forming the so-called Verocay bodies. The Antoni B pattern is composed of scattered and irregular Schwann cells separated by loose myxoid stroma.^{4,5} On immunohistochemistry, S-100 immunoreactivity confirms the origin of the spindle cells from the schwann cell.^{5,6}

The diagnosis of intraosseous schwannoma was established. We removed the soft tissue component of the tumor in the web space by marginal excision. The lesion in the bone was curettaged and the cavity filled with autogenous bone grafts. The tumor originated in the bone. There was no dumbbell nature to the lesion. Follow-up x-rays after 6 months showed no sign of recurrence; incorporation of the bone grafts into the metacarpal had occurred (Fig. 5).

DISCUSSION

Osseous involvement in intraosseous schwannoma occurs through one of 3 possible mechanisms. The tumor may arise originally within the bone^{2-5,13} or it may invade the bone through a nutrient foramen and produce a dumbbell-shaped lesion as it enlarges.^{2-5,13} In the third mechanism an extraosseous schwannoma that ini-



FIGURE 2: The MRI sagittal cut shows a large heterogeneous lesion on T1-weighted image.



FIGURE 3: Antoni A tissue (large arrow) and Antoni B tissue (small arrow) (hematoxylin-eosin stain; magnification ×100).

tially arises in the soft tissue erodes and penetrates into the adjacent bone.^{4,5,12,13} In this case, as the MRI sections showed, the tumor originated in the bone. The bone was not involved through a nutrient foramen and there was no dumbbell shape to the lesion. Wirth and Bray reviewed intraosseous schwannomas.¹ They found 3 hand lesions: a left fourth metacarpal lesion described by Jones,⁹ a right ring finger lesion described



FIGURE 4: Immunoreactions of the spindle cells are positive for S-100 (magnification \times 400).



FIGURE 5: Follow up x-ray after 6 months.

by Morton and Vasser,¹⁰ and a distal phalanx of right index finger reported by Seth et al.⁸ Vora et al.⁶ reported a case of intraosseous schwannoma in the long finger metacarpal. Flugel et al. described a case of intraosseous schwannoma in the proximal phalanx of the right thumb.¹¹ Kujala et al. presented a case of intraosseous schwannoma in the middle phalanx of a right ring finger.⁷ In the mentioned literature, the exact mechanisms of intraosseous schwannomas in the hand were not mentioned. Osseous involvement has occurred mostly in the mandible.^{1,3,5,6} It had been suggested that the long course of the alveolar nerve in the mandible may predispose the development of the tumor. This theory has been challenged because nerves with a longer course innervating the long bones have a lower incidence of schwannoma.^{5,6} Manor et al. analyzed the cells of an intraosseous schwannoma involved in the mandible of a 57-year-old woman. Of 25 cells, 10 (40%) were found to carry choromosomal aberrations. The importance of their finding in diagnosis and prognosis needs further examination by karyotyping of more schwannoma cells.¹⁴

The x-ray features of intraosseous schwannoma include a well-defined lytic lesion, scelerotic margins, lobulated or trabeculated contour, cortical expansion and erosion, and absence of central calcification.^{4,15,16} However, the x-ray findings are nonspecific.^{1,6,16} The MRI appearance of the tumor tends to be isointense to skeletal muscles on T1-weighted images and markedly hyperintense to fat on T2-weighted images.^{4,16,17}

Although most schwannomas contain both Antoni A and Antoni B arrangements, further subclassification of these lesions by predominate tissue has been considered. Verocay type is the lesion in which Antoni A tissue is dominant, and Antoni type is the lesion in which Antoni B tissue is dominant.³ Antoni A and Antoni B have no bearing on the biologic behavior of the tumor,¹ but some authors think that Antoni B tissue may indicate aggressive behavior.^{18,19}

Because malignant change and recurrence are extremely rare, curettage and bone grafting is the most recommended treatment for intraosseous schwannomas.^{4–6,12,16}

Owing to the rarity of the lesion, the diagnosis is often not even suspected until histologic study after a biopsy.¹⁷ The aim of this report was to bring attention to the possibility of intraosseous schwannoma in the differential diagnosis of well-defined, slow-growing, and benign-appearing osseous tumors of the hand.

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