An unusual localization of intraosseous schwannoma: the hamate bone

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Abstract

Intraosseous schwannoma of the hamate bone presented in this case is a very rare benign tumor, and its diagnosis combined with clinical, imaging and needle biopsy is important to guide further therapy. The diagnosis of schwannoma of the hamate was proved histologically following its surgical treatment by curettage.

Introduction

Schwannomas (neurilemmomas) are the most common tumors derived from schwann cells of nerve fibers. It is a relatively common tumor, approximately 5% of all benign soft tissue tumors. Intraosseous schwannoma, however, is very rare, accounting for less than 1% of all benign bone tumors.1-8 They are generally asymptomatic. Radiologically, schwannomas present as well-circumscribed lytic, expansile intramedullary lesions with sclerotic borders. There is no predilection of sex, age and location. There is no case of intraosseous schwannoma around the wrist has been described in literature. We present the first case report of an intraosseous schwannoma of hamate bone of the wrist.

Case Report

A 34-year-old woman presented with 2-year history of the right wrist pain. The pain generally occurred in the morning and increased with daily activations. There was mildly significant swelling on the wrist. Physical examination showed a 2×2-cm firm, round and hard mass on the dorsal aspect of the right wrist. The wrist had almost full range of motion. The overlying skin was intact, and there was no evidence of warmth, erythema, or induration.

Plain radiographs revealed a lytic lesion with well-defined sclerotic borders in the hamate bone. It was multilocular, with no inner calcifications; the dorsal cortical part of the bone was disrupted without periosteal reaction (Figure 1A). Computerized tomography (CT) scans showed the lesion was expansile with cortical break-through in its its dorsal aspect. It has sclerotic, well-defined borders with narrow transition zone (Figure 1B). Magnetic resonance imaging (MRI) revealed again cortical disruption of the dorsal cortex and extraosseous soft tissue extension of the lesion to the dorsal side of the hand (Figure 2A). The lesion was hypointense on PD-weighted sequences, and demonstrated solid and homogenous enhancement on contrast-enhanced T1-weighted sequences. Despite its soft tissue extension, it has still well-defined borders within bone and soft tissues. Immunohistochemistry studies of the tissue obtained by a needle biopsy showed S-100 protein positive spindle cells, addressing a benign neurogenic tumor. Then, we performed totally resection of the lesion by curettage via dorsal longitudinal incision. After curettage, bone defect was reconstructed by autograft harvested from the iliac crest. The gross specimen showed a solid lobular tumor with clear boundary, partly located within the bone. Histologically, the tumor was composed of multiple nodules of spindled Schwann cells within in the bone parenchyma (Figure 3A and B). Antoni A pattern was predominant. Nuclear palisading and Verocay bodies were present (Figure 3C). Immunohistochemically, tumoral cells were strongly positive for S-100 protein (Figure 3D). The final diagnosis was plexiform schwannoma of the bone. The patient did not receive any adjuvant therapy after the surgery. She had been followed up for seven months with no sign of recurrence and pain (Figure 2B).

Discussion

Schwannomas are the most common tumors derived from schwann cells of nerve fibers. Intraosseous schwannoma, however, is very rare, accounting for less than one per-cent of all benign bone tumors.1-8 In addition to the sacrum, iliac wing and mandible, intraosseous schwannomas also occur in the long bones, vertebra, fibula and frontal bone.1-3,8 However, there is no case in literature intraosseous schwannoma of hamate bone. Schwannomas may involve bone tissue via three mechanisms: i) they may be intramedullary, producing rarefaction of the bone; ii) they may be located within the nutrient canal, with the formation of a dumbbell-shaped tumor; or iii) they may be extraosseous, eroding into the bone.1-6 On the base of imaging, intraoperative findings of dumbbell-shaped tumor inside and outside the hamate bone and gross examination, this case demonstrates an example of intraosseous schwannoma.

Intraosseous schwannomas have characteristic radiographic features such as a well-defined lytic lesion, sclerotic margins, narrow transition zone, lobulated contours, cortical expansions and absence of central calcification. However, all these radiographic findings are non-specific and does not help clinicians in differential diagnosis. On the basis of CT and direct X-ray, it is difficult to differentiate the intraosseous schwannoma from other lytic benign bone lesions of such simple bone cyst, aneurysmal bone cyst, and giant cell tumor of bone, fibrous dysplasia, enchondroma and non-ossifying fibroma. On MRI, schwannomas are solid lesions contrary to simple bone cysts and aneurysmal bone cysts. T2 hyperintensity of the schwannomas can be bright as much as that of enchondroma. However, enchondromas may contain calcifications with typical contrast enhancing views on MRI. Giant cell tumors, fibrous dysplasia and non-ossifying fibromas are heterogeneous tumors having both T2 hyper and hypointense areas. On CT, giant cell tumors have lytic borders due to its local agressive-
Ground-glass appearance with wide transition zone. CT and MRI scans in our case showed that the lesion was expansile with cortical break-through and extra osseous extension but well defined borders within both bone and soft tissues. The sclerotic, well-defined borders have narrow transition zone on CT and plain radiography. These findings may be the characteristic for intraosseous schwannomas. The final diagnosis of intraosseous schwannoma was made by histologic examination of tumor. The histologic features of this lesion are similar to those of soft tissue schwannoma.

Conclusions

Making diagnosis of an intraosseous schwannoma pre-operatively with clinical and radiological findings, and needle biopsy was beneficial to the patient to avoid unnecessary adjuvant therapy. Surgical excision is the preferred treatment option. If significant bony defect occurs, bone grafting should be considered. These tumors are generally well encapsulated without invasion of the surrounding structures, enabling complete resection possible. Recurrence is rare after complete resection.

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Figure 1. A) AP-plain x ray through wrist shows a lytic expansile lesion in hamate bone. It has benign appearance with well-defined border and narrow transition zone. Cortical break through is noted in its dorsal aspect. B) On coronal CT scan through carpal bone, a lytic lesion with well defined, sclerotic borders and narrow transition zone is seen in hamate bone. The lesion is expansile with extrasosseous soft tissue extension from its dorsal aspect.

Figure 2. A) On coronal PD-fat sat MRI scan through carpal bone, the lesion in hamate bone is T2 hyperintense, expansile and multilobulated with well-defined borders despite its soft tissue extension. On postcontrast images (not shown here), the lesion is solid and homogeneously enhancing. B) The patient did not receive any adjuvant therapy after curettage and autograft. She had been followed up for 7 months with no sign of recurrence and pain.

Figure 3. Microscopic appearance of the case. A) Plexiform growth pattern of tumor with multiple nodules, HEx40; B) The tumor within bone parenchyma (arrow indicates the calcified bone trabeculae), HEx40; C) Antoni A areas illustrating prominent nuclear palisading (Verocay bodies), HEx100; D) Strong staining of Schwann cells with S100-protein, but not intervening stroma.