A Rare Primary Intraosseous Schwannoma of the Tibia

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Case Report

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Abstract

**Background:** Primary intraosseous schwannoma is a rare benign tumor derived from schwann cells that cover myelinated nerve fibers, accounting for less than 0.2% of all primary bone tumors. Because of its rarity, the new World Health Organization classification of bone tumors has deleted it.

**Case presentation:** A 29-year-old woman presented with a history of intermittent pain on her right ankle over three years. Radiological examination revealed a multilocular cystic and lytic lesion in the distal tibia. Histologically, the tumor was composed of spindle shaped cells, similar to soft tissue neurilemmoma. The patient underwent a surgical resection of the intraosseous lesion. After a 38-month follow-up, the symptoms were significantly relieved and there was no clinical or radiological recurrence.

**Conclusion:** Primary intraosseous schwannoma is a rare benign bone tumor. The pathologists should be aware of the incidence of the tumor, although WHO has deleted it.

Introduction

Primary intraosseous schwannoma is a rare benign bone tumor, accounting for less than 0.2% of all primary bone tumors[1]. It is an infrequent entity, especially occurring in long or short bones, such as femur, metatarsal, calcaneus and hamate[2–6]. In this article, we reported a case of a intraosseous schwannoma in the tibia.

Case Presentation

A 29-year-old woman presented with a history of intermittent pain on her right ankle over three years. The pain had increased gradually in the past one year. There were no obvious initiating factors, such as trauma or inflammation. Physical examination revealed tapping pain on the right lower extremity. The overlying skin was intact and there was no evidence of fistula, induration or hyperemia. No evidence of lymphadenopathy was found.

Plain radiographs indicated a multilocular cystic and lytic lesion in the distal tibia. The inner partial cortex was disrupted without apparent periosteal reaction. Computerized tomography (CT) scans showed that the lesion was expansive with cortical interruption and mild sclerotic borders (Fig. 1). Magnetic resonance imaging (MRI) in other hospital demonstrated a well-defined lesion with hypo-intense signal on T1-weighted images and hyper-intense on T2-weighted images. Edema of periosteum and soft tissue was observed.

Based on examinations, benign or low-grade malignant tumor was suspected by clinicians. In the operation, hemorrhagic efflux was visible from the marrow cavity. Intramedullary tissue was scrapped for biopsy. Microscopically, the histological features of the tumor were consistent with schwannoma in soft tissues, which was mainly composed of spindle shaped cells. The tumor contained hypercellular regions
with nuclear palisading in fibrous background (Antoni A pattern) and hypocellular regions with myxoid matrix (Antoni B pattern) (Fig. 2a). In addition, bone interruption could be seen in the tumor (Fig. 2b). There was no evidence of mitotic activity and necrosis. Immunohistochemistry showed that the tumor cells were diffusely positive for S-100 and SOX-10 protein (Fig. 3). Ki-67 proliferation index was less than 5%.

Based on these evidences, the diagnosis of primary intraosseous schwannoma occurring in tibia was established.

**Discussion**

Schwannoma is a benign nerve sheath tumor deriving from Schwann cells which cover the peripheral myelinated sensory nerves. Schwannoma usually occurs in soft tissues while primary intraosseous schwannoma is exceptionally uncommon, accounting for less than 0.2% of all primary bone tumors. By reviewing the literature, there are three underlying hypotheses of the development of intraosseous schwannoma: (1) the tumor arises from the periosteum lesion which invades into the bone by consequent erosion. (2) the tumor originates from the nerves which enter bone through nutrient canals. (3) the tumor occurs directly within the medullary cavity from minute non-myelinated nerves associating with vessels. In the literatures, intraosseous schwannoma is commonly found in the mandible, maxilla, sacrum and vertebrae\[7–11\]. These bones contain nerve foramina with peripheral nerves pass through it. The tumor may arise from the nerves traversing intraosseous canals or from sensorimotor nerves passing through sacral foramina. Strictly, they are not primary intraosseous schwannoma. Primary intraosseous schwannoma is limited to those arising from the substance of the bone, such as minute nerves accompanying vessels that nourish bone on the medullary cavity. The rarity of primary intraosseous schwannoma may be due to the low density of sensory nerves in bone. Munehisa et al.\[12\] has suggested that the apoptosis of sensory nerve fibers within bone tissue may lead to the rare occurrence of intraosseous schwannoma. Most intraosseous nerves are non-myelinated nerves travelling with the nutrient vessels and participate in vasomotor functions. Primary intraosseous schwannoma probably arises from Schwann cells of these paravascular nerves.

In our case, the tumor occurs in the medullary cavity of long bone and has the typical morphological characteristics of schwannoma, so it’s a real primary intraosseous schwannoma.

**Conclusion**

The purpose of our paper is to attract attention to the fact that intraosseous schwannoma does exist as a primary bone tumor. Its rarity, especially long bone involvement, makes it easily to be ignored in the initiate differential diagnosis.

**Declarations**
Statements

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication was obtained from the patient.

Availability of data and materials

All data generated or analysed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

Each author is expected to have made substantial contributions to the conception. Xueqing Liu analyzed the patient data and wrote the manuscript; Yunlong Ding and Yanling Shen performed the histological examination; Wensheng Yang have drafted the work and substantively revised it. All authors read and approved the final manuscript.

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