Primary Intraosseous Schwannoma

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Abstract

Schwannomas are benign tumors arising from the peripheral nerve sheath. Neurogenic tumors of bone are extremely uncommon and they compose less than 1% of all benign tumors [1]. We present a case of intraosseous schwannoma in a 15 year old girl who presented with pain and pathological fracture of tibia. The radiology revealed an expansile and lytic lesion in the diaphysis. Histopathology confirmed the diagnosis of intraosseous schwannoma. The tumor cells were immunoreactive for S100protein. We present this case as tibial schwannoma is extremely rare and its diaphyseal location in the bone is virtually unknown.

Key words: Schwannoma, tibia, diaphysis

Introduction

In 1908, Verocay first described a nerve tumor arising from the nerve sheath and termed it neurinoma. Stout, later on, went to describe tumors from nerve sheath and neuroectodermal origin [1]. Schwannoma almost always manifests as a solitary neoplasm most commonly occurring at the flexor surfaces of the extremities, posterior spinal roots, and cerebellopontine angle [2]. In the bone, it is generally found in the mandible and sacrum. It constitutes <1% of all benign bone tumors [3]. The rarity of osseous involvement leads to omission of schwannoma from the initial differential diagnosis in the majority of cases [4].

Case Report

A 15-year-old female presented with a history of pain and swelling over the middle one-third of the left leg. On examination, the swelling was of 4 cm × 3 cm × 2 cm. The radiological examination done in the form of an X-ray revealed an expansile and lytic lesion measuring 1.8 cm × 1.4 cm in the diaphyseal region of the left tibia, along with a pathological fracture (Fig. 1). The differential diagnosis based on radiology, comprised benign bone tumors including bone cyst, aneurysmal bone cyst, giant-cell tumor, and fibrous dysplasia. The patient was operated and some part of tumor tissue was sent for histopathological evaluation. We received the gross specimen as multiple bits of gray-white tissue pieces aggregating to 1.5 cm × 1 cm × 0.5 cm. The entire tissue was embedded for paraffin block. The routine H and E sections revealed a schwannoma with cells arranged in two different patterns. Antoni A area was cellular and made of spindle cells arranged in palisading fashion (Verocay bodies). Antoni B area had tumor cells separated by edematous fluid forming cystic spaces (Fig. 1c). Vascular spaces were noted along with the areas of hemorrhage. Mitotic activity was extremely scanty. The periphery of the tumor showed native bone. Due to the presence of such a rare tumor in the diaphysis, immunohistochemical studies were carried to confirm the obvious diagnosis of schwannoma. As expected, the tumor cells diffusely expressed S100p (Fig. 1a), Vimentin (Fig. 1d) and were negative for SMA (Fig. 1b).

Discussions

Primary neurogenic tumors are a part of uncommonly rare tumors of bone [5]. Benign peripheral nerve sheath tumors can be classified into schwannomas and neurofibromas [6]. Neurofibromas should be distinguished from schwannomas because the latter have practically no potential for malignant transformation [5]. These tumors are shown a peak prevalence in the second decade of life and are known to have a slight female predilection with a ratio of 1.6:1 [7]. Schwannoma is associated closely with sensory nerves that are of low density within the bone. Therefore, their occurrence in the bone is exceedingly low [8]. There are three mechanisms by which schwannomas may involve bone: (1) A tumor may arise centrally within the bone, (2) a tumor may arise within the nutrient canal and produce canal enlargement, or (3) a soft tissue or periosteal tumor may cause secondary erosion and penetration into bone [9]. In our case, although it may be difficult to decipher the exact pathogenesis, looking at the radiology and circumscription of the tumor, we propose that the tumor may have risen centrally within the bone. Although, Suzuki et al. have discussed in their article that, as intraosseous nerves are typically associated with arterial vessels in the nutrient canal, an intraosseous schwannoma involving a long bone may be associated with the second mechanism.; they concur that the most common location of intraosseous schwannomas in long bones remains to be elucidated [10].

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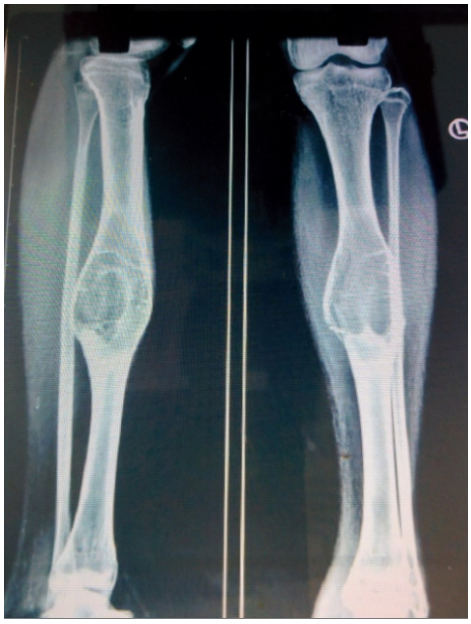

Fig. : 1- X-ray revealed an expansile and lytic lesion measuring 1.8 cm × 1.4 cm. In the diaphyseal region of the left tibia, along with a pathological fracture

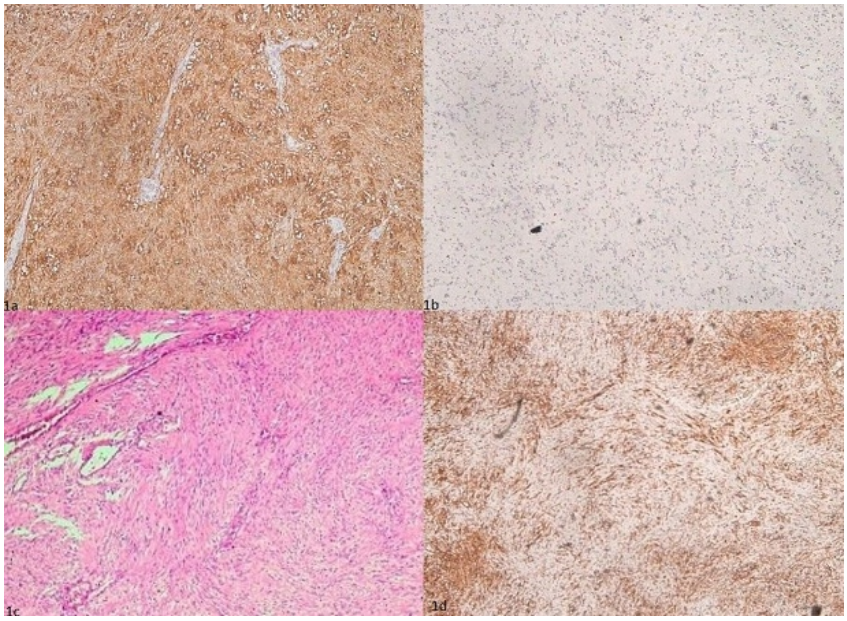

Fig. : 2 The routine H and E sections revealed a schwannoma with cells arranged in two different patterns. Antoni A area was cellular and made of spindle cells arranged in palisading fashion (Verocay bodies). Antoni B area had tumor cells separated by edematous fluid forming cystic spaces (Fig. 1c). immunohistochemical studies showed tumor cells diffusely expressed S100p (Fig. 1a), Vimentin (Fig. 1d) and were negative for SMA (Fig. 1b).

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