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Case Report

An intraosseous malignant peripheral nerve sheath tumor in the mandible of a patient with neurofibromatosis type 1

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas that originate in peripheral nerves or neurilemma cells. Here, we report an extremely rare case of an intraosseous MPNST in the mandible of a patient with neurofibromatosis type 1 (NF1). A 57-year-old woman with a history of NF1 was referred to our hospital because she had abnormal sensations at her left mandible. She was diagnosed with MPNST and underwent radical resection, although local recurrence and multiple metastases were detected during follow-up. Despite receiving palliative radiotherapy, the patient died at 13 months after the initial diagnosis.

1. Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas that originate in peripheral nerves or neurilemma cells, and these tumors are characterized by rapid tumor expansion, early invasion of the surrounding tissue, and distant metastasis [1]. MPNST accounts for approximately 10%–12% of soft tissue sarcomas, and approximately one-half of the patients with MPNST also have neurofibromatosis type 1 (NF1) [2].

As MPNST generally arises in the soft tissue and rarely in the bone, intraosseous MPNST in the jaw is extremely rare, with only 20 cases reported in the English literature [3]. Furthermore, most cases of intraosseous MPNST in the jaw are not associated with NF1. To the best of our knowledge, only one case report has described an intraosseous MPNST in the maxilla of a patient with NF1 [3]. Thus, we report an extremely rare case of an intraosseous MPNST in the mandible of a patient with NF1.

2. Case report

A 57-year-old woman experienced abnormal sensations at her left mandible, and she was referred to our clinic. During the year before her visit, she had been noticing a slightly abnormal sensation at her left mandible, which gradually progressed to numbness. The patient initially visited a general dental practitioner and underwent a radiographic examination, which suggested an odontogenic tumor with a widespread osteolytic lesion in her left mandible. Based on a diagnosis of NF1, the patient had also previously undergone 14 local excisions of neurofibromas at our dermatology clinic.

An initial assessment in our clinic revealed no systemic symptoms, although countless cutaneous neurofibromas had manifested as raised bumps throughout her skin. An extraoral examination revealed no remarkable findings (other than the cutaneous neurofibromas) (Fig. 1A), and her regional lymph nodes were not palpable. An intraoral examination revealed diffuse swelling that extended from the second premolar to the retromolar area of her left mandible (Fig. 1B). The mucosal surface of the mass was smooth, and no ulceration was observed. Panoramic radiography revealed a poorly defined radiolucent lesion in the left mandible (Fig. 1C), and computed tomography (CT) revealed a poorly defined homogeneous enhancement of the mass with destruction of the cortical bone in the left mandible and the inferior alveolar canal was not identified (Fig. 1D and E). No other specific findings were detected on the abdominal and thoracic CT images. A biopsy specimen was submitted for histopathological testing, which revealed spindle-shaped cells with atypia and polymorphism. Immunohistochemistry revealed that the specimen was positive for S-100. These findings supported the diagnosis of high-grade spindle cell

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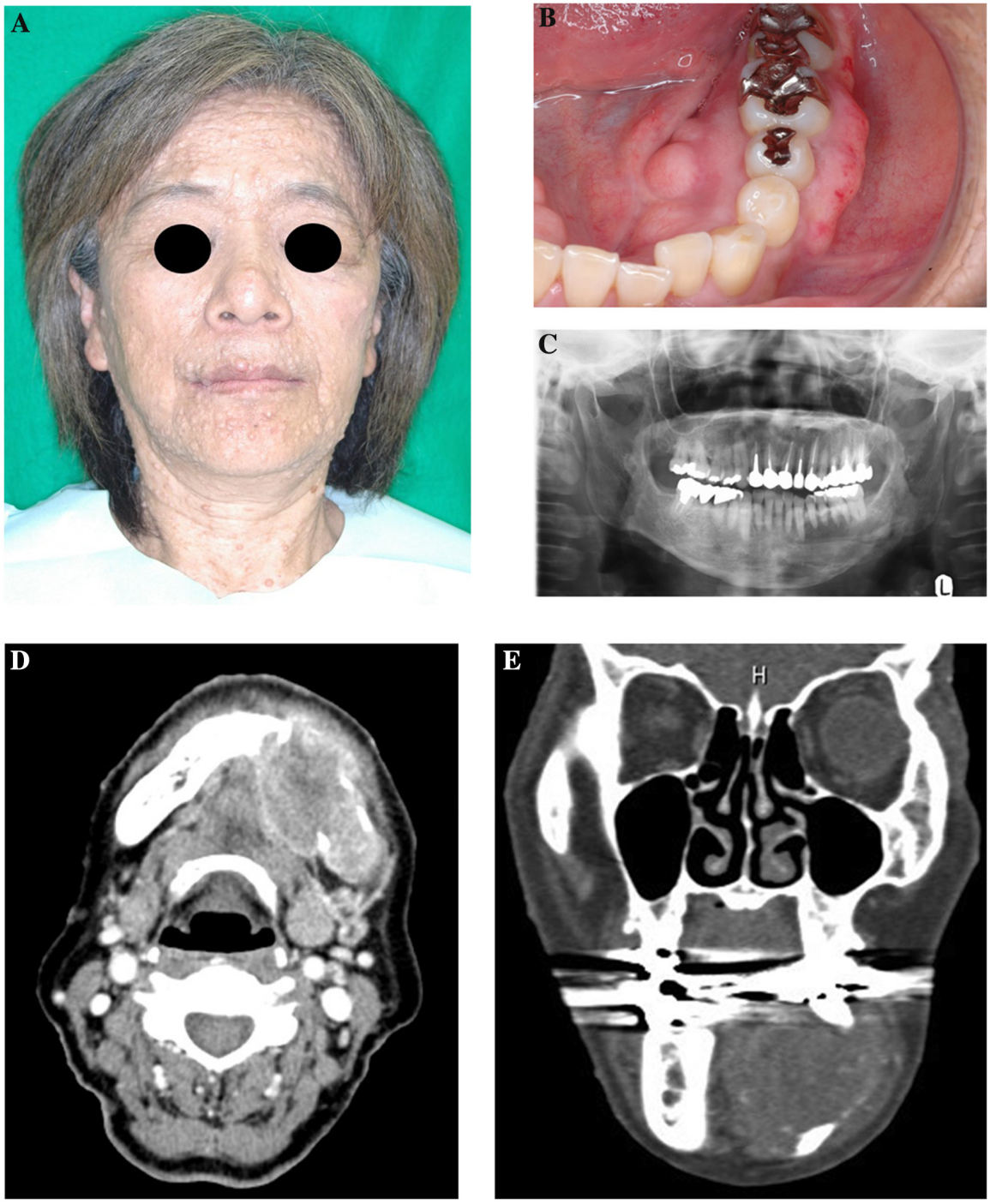

Fig. 1. Preliminary findings. (A) The extraoral examination revealed numerous cutaneous neurofibromas that manifested as small bumps on the patient's skin. (B) The intraoral examination revealed diffuse swelling of the left mandible. (C) Panoramic radiography revealed a poorly defined radiolucent lesion in the left mandible. (D, E) Computed tomography revealed a poorly defined homogeneous enhancement of the mass with destruction of the cortical bone in the left mandible.

sarcoma, and the mass was specifically diagnosed to be MPNST on the basis of the patient's history of NF1.

The patient was treated by radical mandibulectomy with modified radical neck dissection and reconstruction using a free vascularized latissimus dorsi muscle flap and a rigid titanium plate (Fig. 2A and B).

Microscopically, the tumor cells were mostly atypical spindle-shaped with cellular polymorphism and high mitotic activity (Fig. 3A). The immunohistochemical examination of the tumor is summarized in Table 1. The tumor cells were positive for S-100 (Fig. 3B), and focally positive for calponin, but negative for AE1/AE3, epithelial membrane antigen (EMA), alpha-smooth muscle actin (α-SMA), desmin, myogenin, CD34, and c-kit. On the basis of these histopathological features, the tumor was diagnosed to be MPNST.

However, at the 4-month follow-up, local recurrence was detected at the premolar region of the right side of the mandible. Moreover, lung and multiple bone metastases were detected (Fig. 4). The patient underwent palliative radiotherapy for recurrence of the mandibular tumor (30 Gy in 10 fractions to the mandible, vertebral bones, and lumbus). The recurrent tumor pro-