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CASE REPORT

Primary meningioma of the mandible

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KEYWORDS

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Summary We reported a case of primary extracranial meningioma in the mandible of a 10 year-old-boy with basal cell nevus syndrome. The tumor had a well-delineated large round shaped radiolucency including an impacted canine in the mandible. Microscopic examination revealed a fibrous tumor composed of uniform spindle-shaped cells and fine collagen bundles. The spindle-shaped cells were arranged in whorls and interconnecting fascicles, and some nuclear pseudoinclusion and psammoma bodies were detected. Immunohistochemically, the tumor cells were stained for epithelial membrane antigen, vimentin and desmoplakin, but not for S-100 protein. No recurrence of the tumor was detected for 4 years.

Introduction

Meningiomas are benign tumors that are derived from the aracnoid villous structures of the meninges, and are common in the central nervous system, especially of the parasagittal meninges, falx cerebri, and sphenoid ridge.[1] On the other hand, primary extracranial meningiomas are uncommon, and the most cases occurred in glabella, bridge of the nose, skin, temporal bone, paranasal sinus, and pterygopalatine fossa.[1] Extracranial meningiomas in the jaws are extremely rare. To the best of our knowledge, five cases of primary meningiomas in the jaws have been reported in the English literature.[2–5] All of the reported cases in the jaws were present in adult women aged from 26 to 74 years.[2–5] We present an additional case of primary

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extracranial meningioma which occurred in the mandible of a 10 year-old boy with basal cell nevus syndrome.

A case report

A 10 year-old boy was referred to the Kyushu University Dental Hospital for a painless swelling of the mandible in the left molar region. Just before visiting our hospital, his left mandibular first molar deciduous tooth had been extracted by his dentist. The mandible in the left molar region was expanded to the buccal and lingual sides. No tenderness, fluctuation of the mandible or hypoesthesia of the lower lip was detected. Gingival mucosa looked normal. Radiographic examinations showed the presence of a large round and well-defined radiolucency in the mandible including an impacted left canine. The cortex of the mandible was

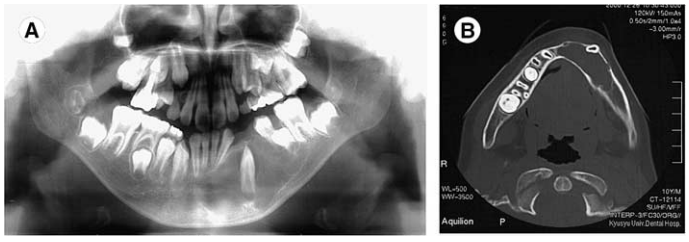

Figure 1 Panoramic radiogram and computed tomographic. A panoramic radiogram (A) and a computed tomographic (B) revealed the presence of a large and well-delineated radiolucency in the mandible. The radiolucency extended from the right second incisor to the left second premolar, and included an impacted left canine.

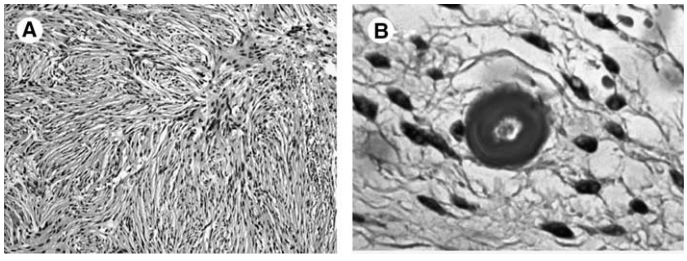

Figure 2 Histopathological findings of the tumor. (A) The tumor was composed fibrous spindle-shaped cells and fine collagen bundles. The uniform spindle-shaped cells were arranged in whorls and interconnecting fascicles (HE; ×100). (B) A psammoma body was identified in the tumor (HE; ×200).

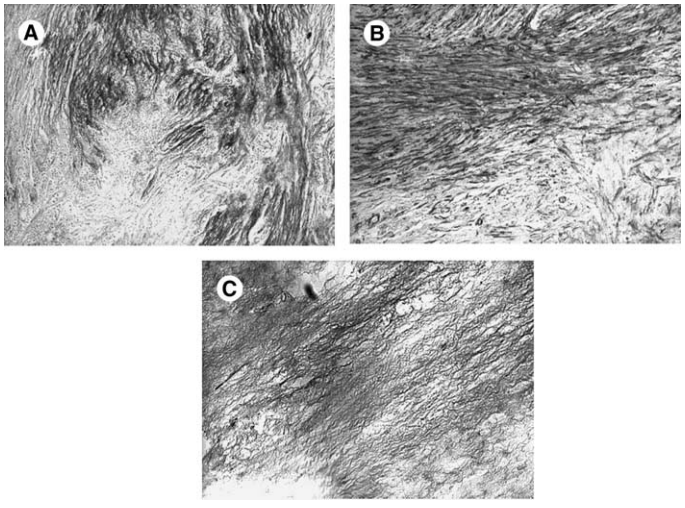

Figure 3 Immunohistochemical staining. The tumor cells were stained with anti-epithelial membrane antigen (A), anti-vimentin (B), and anti-desmoplakin (C) antibodies (magnification ×100).

irregularly absorbed (Fig. 1). An incisional biopsy of the lesion revealed the presence of spindle cells and collagen bundle without atypical nor mitotic figures in the specimen, but the final diagnosis was elusive. The tumor, including the impacted canine and the left first premolar, was enucleated in a lump from the mandible under general anesthesia. The tumor was apparently dissociated from the mandibular nerve. Six months after the surgery, new bone formation in the defect of the mandible was detected in radiograms. Odontogenic keratocysts appeared in the lower right third molar region and the upper right impacted canine region 2 years after the tumor enucleation. He also had palmar pits in both hands, and was diagnosed as basal cell nevus syndrome. No recurrence of the meningioma or odontogenic keratocyst was observed during a 4-year follow-up period.

Pathologic findings

The enucleated tumor was composed of thin spindle-shaped cells and fine collagen bundles. The uniform spindle-shaped cells were arranged in whorls and interconnecting fascicles, and some nuclear pseudoinclusion and psammoma bodies were detected (Fig. 2).

An immunohistochemical examination with monoclonal antibodies for epithelial membrane antigen (EMA) (CN Bioscience Inc.; 1:250) and human vimentin (ICN Pharmaceuticals Inc.; 1:250), and polyclonal antibody for S-100 protein (Ylem S.R.L.; 1:500) on the paraffin sections, and with monoclonal antibody for desmoplakin (PROGEN Biotechnik GmbH; 1:10) on the frozen sections revealed that the tumor cells were stained positively for EMA, vimentin, and desmoplakin, while S-100 protein was not detected in the sections (Fig. 3).

Discussion

In this paper, we have reported a case of primary extracranial meningioma of the mandible in a 10 year-old boy with basal cell nevus syndrome. Extracranial meningiomas in the jaws are extremely rare, and only five cases in the jaws have been reported in the English literature.[2–5] Extracranial meningiomas are thought to arise from displaced arachnoid cell rests during embryogenesis or neural sheeth with arachnoid cell rests. Nerve injury such as extraction of teeth or chronic apical inflammation might be a cause for the acceleration of ectopic arachnoid cells.[3,5] The tumor in this case, however, was completely separated from the inferior alveolar nerve, and neither the history of a mandibular injury nor an infection of the left mandibular molar region was present. An interesting finding is that the patient was diagnosed as basal cell nevus syndrome by multiple odontogenic keratocysts in the jaws and palmar pits according to a diagnostic criteria of the syndrome.[6] The syndrome shows a high incidence of tumors such as multiple basal cell carcinomas, medulloblastomas, and meningiomas. Most cases of meningiomas in the syndrome occurred after radiation therapy for the medulloblastomas.[6] In our case, the patient has not had an experience of radiation therapy. Recently, it has been shown that the mutations of the *patched* gene, the human homolog of Drosophila segment polarity gene *patched*, are associated with odontogenic keratocysts and meningiomas in the syndrome.[7,8] The *patched* gene mutation, therefore, might be involved in this case.

Meningiomas are divided into some subtypes e.g. meningotheliomatous, fibrous, transitional, psammomatous, angiomatous, haemangioblastic, haemangiopericytic, papillary, and anaplastic types. Histopathological findings suggest that the tumor in our case is of fibrous type. The tumor cells were stained with EMA, vimentin, and desmoplakin antibodies immunohistochemically, while no immunoreactivety was detected for S-100 protein. Although there are some different diagnoses for meningiomas-such as Schwannoma, neurofibroma, paraganglioma, and perineurioma-Schwannoma, neurofibroma, and paraganglioma can be distinguished from meningioma for their different immunophenotype (positive for S-100 protein). On the other hand, the histopathological structure of perineurioma quite resembles meningioma. The whorl-like structures and psammoma bodies, which are a well-known structure in meningioma, are frequently found in perineurioma.[9] Furthermore, the tumor cells in both tumors are stained positively for EMA and vimentin, but not for S-100 protein immunohistochemically. Desmoplakin is one of the desmosomal plaque components. It has been reported that desmoplakin is a suitable marker for distinguishing meningeal tumors from perineurioma, because desmoplakin is expressed in the cells of arachnoidal tissue both of diverse meningioma subtypes and of a meningioma-derived cell line, while perineuriomas lack immunoreactivity for desmoplakin.[10]

The prognosis of meningiomas in the jaw is reported to be good relatively when complete surgical excision or mandibular resection is performed.[3,4] However, a case of recurrent extracranial meningioma presenting 27 years after

successful surgical eradication of the primary lesion has been reported.[11] Thus, long term follow-up is recommended in this case, although no recurrent sign was observed in a 4-year follow-up period.

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