

## Multiple schwannomas in the oral floor : Case report

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## **Multiple schwannomas in the oral floor: A case report**

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## **Abstract**

We herein present a rare case of multiple schwannomas in the oral floor that occurred in a 62-year-old male patient. No café-au-lait spots on the skin, cutaneous tumors, the eighth nerve tumors, or lens opacity was detected. There was no history of neurofibromatosis 2 or definite schwannomatosis in any first-degree relative. A CT scan and MR imaging demonstrated the presence of two isolated and well-bordered tumors in the oral floor. Both tumors were histopathologically diagnosed as schwannoma. Immunohistochemically, the tumor cells were positive for the S-100 protein and CD34, while the cells were negative for epithelial membrane antigen (EMA) and Factor XIIIa. This case, therefore, met the diagnostic criteria for schwannomatosis.

**Keywords:** multiple shwannomas; oral floor; schwannomatosis

## **Introduction**

Schwannoma is a relatively uncommon benign nerve sheath tumor in the oral cavity and usually occurs as a solitary tumor.<sup>1,2</sup> Multiple schwannomas are often detected in neurofibromatosis (NF) 2 which is an autosomal dominantly inherited disorder.<sup>3</sup> Recently, schwannomatosis is recognized as the third major form of NF.<sup>4</sup> Schwannomatosis is characterized by multiple non-intradermal or non-vestibular schwannomas. The presence of multiple schwannomas in a single patient thus suggests a possible association with NF2 or schwannomatosis. In this paper, we present a rare case

of multiple schwannomas in the oral floor which met the diagnostic criteria for schwannomatosis.

### **Case report**

A 62-year-old male was referred to Kyushu University Hospital for a painless swelling of the oral floor on the left side in May 2009. The patient had noticed the swelling about 3 months prior to visiting the hospital. Clinical examination revealed the presence of a tumor which was well-bordered, non-tender, and non-compressible. The overlying oral mucosa of the tumor was normal (Figure 1). No café-au-lait spots on the skin, cutaneous tumors, the eighth nerve tumors, or lens opacity was detected. There was no history of NF2 or definite schwannomatosis in any first-degree relative.

Computed tomography (CT) examination demonstrated the presence of large (40 x 30 mm) and small (10 x 10 mm) low-density tumors in the oral floor. Both tumors were well-bordered and no communication was detected between the two tumors. T1-weighted magnetic resonance (MR) image showed that both tumors were homogeneously hypointensive, while T2-weighted MR image and Gadolinium-enhanced T1-weighted MR image showed that both tumors were heterogeneously hyperintensive (Figure 2). No vestibular tumor was detected.

Schwannoma or neurofibroma was suggested by an incisional biopsy of the large tumor. The large and small tumors were completely enucleated through an intraoral approach under general anaesthesia. Both tumors were encapsulated and were completely distinguished from each other. There was no apparent communication

between the tumors and nerve bundles. Postoperative healing was uneventful.

Histopathologically, the specimens of the large and small tumors revealed a characteristic encapsulated schwannoma composed of Antoni-A and Antoni-B patterns (Figure 3). Immunohistochemical staining revealed that the tumor cells were positive for the S-100 protein but negative for epithelial membrane antigen (EMA) and Factor XIIIa. The tumor cells in the Antoni-B region were positive for CD34.

## **Discussion**

We herein present a rare case of multiple schwannomas in the oral floor. Histopathologically, the Antoni-B structure of schwannoma is sometimes difficult to distinguish from neurofibroma. Recently, nerve sheath tumors with hybrid features of neurofibromas and schwannomas have been reported in the literature.<sup>5</sup> Immunohistochemically, schwannomas are positive for S-100 protein and CD34, but negative for Factor XIIIa.<sup>6-9,10</sup> On the other hand, neurofibromas are positive for S-100 protein, CD34, and Factor XIIIa.<sup>6,8,9</sup> Therefore, the staining intensities for Factor XIIIa are clearly different between schwannoma and neurofibroma. The tumor cells in our case were negative for Factor XIIIa, supporting the diagnosis of the tumors in this case as being schwannomas.

The diagnostic criteria for schwannomatosis consist of a two-tiered paradigm for “definite” and “possible” schwannomatosis (Table 1). Possible schwannomatosis is permitted when a patient is older than 45 years of age, does not have symptoms of an eighth nerve dysfunction, and the tumors are multiple non-intradermal schwannomas.

Therefore, our case may meet the diagnostic criteria for possible schwannomatosis.

A surgical excision is the treatment of choice for schwannoma. Malignant change is extremely rare in a benign schwannoma and no reports of such change occurring in the cases of multiple schwannomas. In our case, no recurrence of the tumor was observed during the 1-year follow-up period.

### **Acknowledgments**

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### **Conflict of interest**

The authors do not have any financial and personal relationships with other people or organizations that inappropriately influence this work.

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### **Legends**

**Figure 1.** Intraoral view. A large tumor that presented in the oral floor on the left side was well-bordered, firm, and non-compressible.

**Figure 2.** MR image. MR image demonstrated the presence of well-bordered hyperintensive large and small tumors (white arrows) in the oral floor. The small tumor was located inferior to the large one.

**Figure 3.** Histopathological finding of the large tumor. Hematoxylin-eosin (HE) stain in Antoni-A region composed of interwoven bundles of long bipolar spindle cells (100X magnification).



**Fig. 1**



Fig. 2

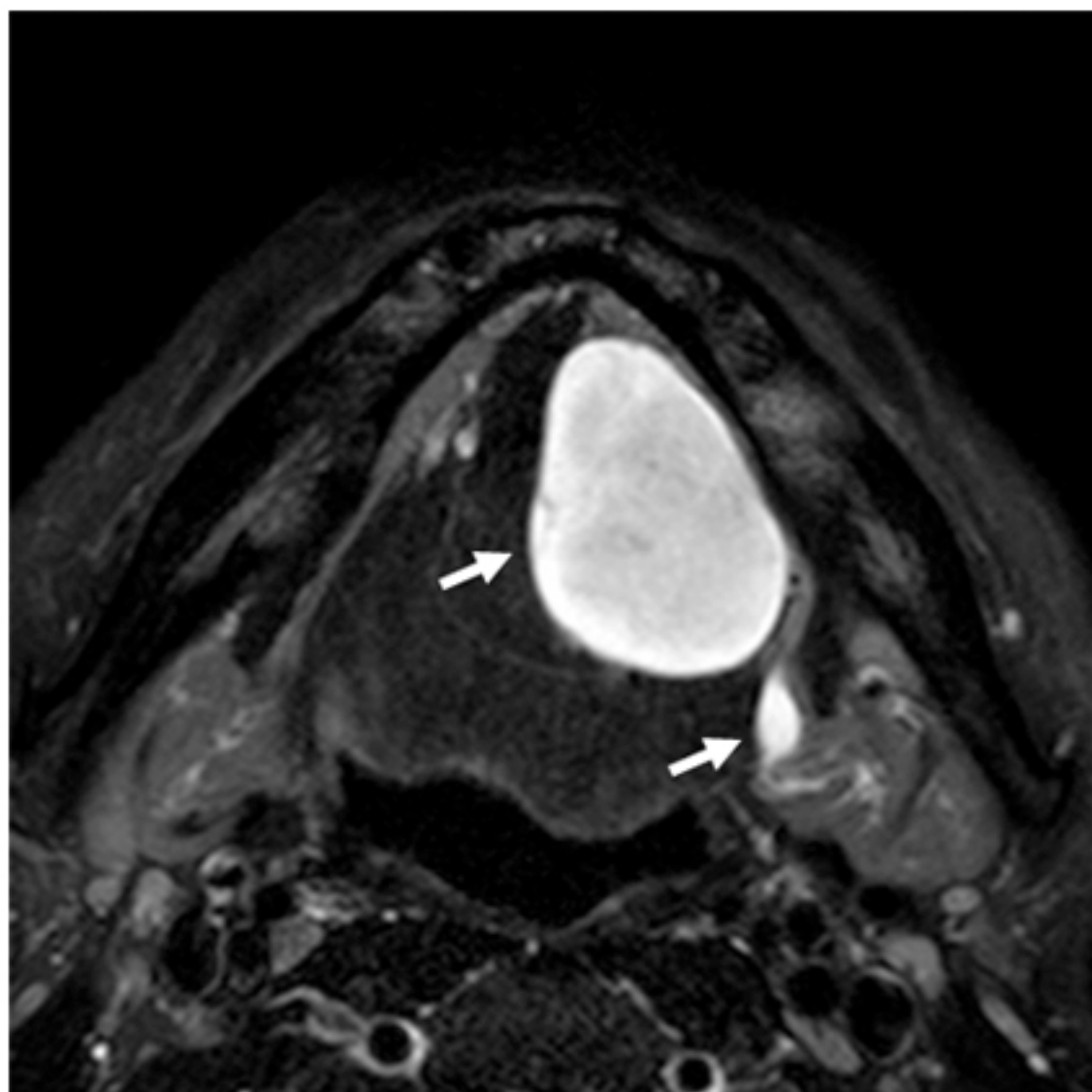


Fig.3

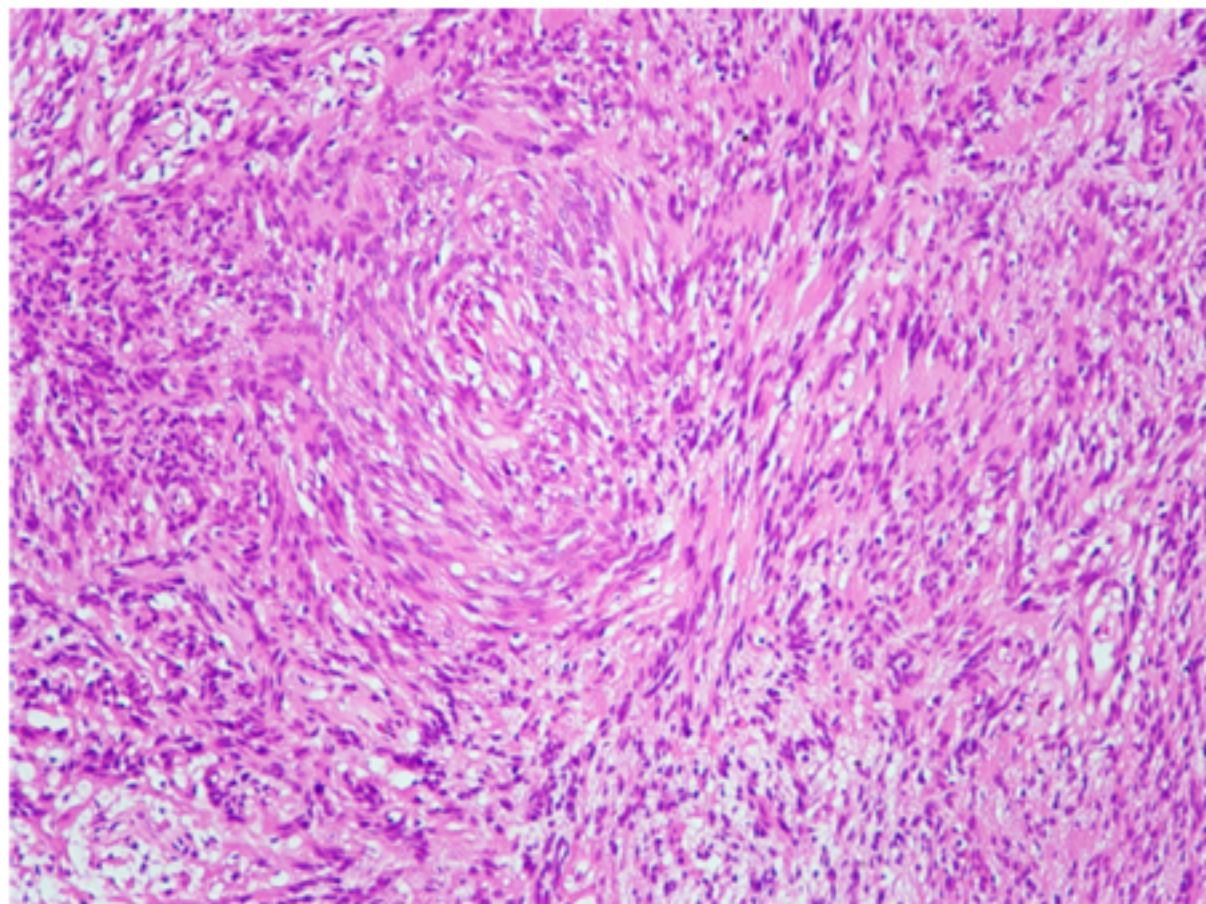


Table 1. Diagnostic criteria for NF1, NF2, and schwannomatosis

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I. NF1 (NIH criteria<sup>3</sup>: 1987); autosomal dominantly inherited disorder (chromosome 17q)

Two or more of the below criteria

- i) Café-au-lait spots (6 or more) (>5 mm in children or >15 mm in adults)
- ii) Cutaneous or subcutaneous neurofibromas (2 or more) or one plexiform neurofibroma
- iii) Axillary or groin freckling
- iv) Optic glioma
- v) Lisch nodules (2 or more)
- vi) Bone lesion with sphenoid wing dysplasia or bowing of the long bones with/without pseudoarthrosis
- vii) First-degree relative with NF1

II. NF2 (Manchester criteria<sup>3</sup>: 1992); autosomal dominantly inherited disorder (chromosome 22q)

- 1) Bilateral vestibular schwannomas
- 2) First-degree relative with NF2 *and* unilateral vestibular schwannoma or two of meningioma, schwannoma, glioma, neurofibroma, posterior subcapsular lens opacity
- 3) Unilateral vestibular schwannoma *and* two of meningioma, schwannoma, glioma, neurofibroma, posterior subcapsular lens opacity
- 4) Multiple meningiomas (2 or more) *and* unilateral vestibular schwannoma or two of schwannoma, glioma, neurofibroma, cataract

III. Schwannomatosis (Baser et al.<sup>4</sup>: 2006)

(1) Definite schwannomatosis

- 1) Age >30 years *and* non-intradermal schwannomas (2 or more)
- 2) One pathologically confirmed schwannoma *and* a first-degree relative who meets the above criteria

(2) Possible schwannomatosis

- 1) Age <30 years *and* non-intradermal schwannomas (2 or more)
  - 2) Age >45 years *and* no symptoms of eighth nerve dysfunction *and* non-intradermal schwannomas (2 or more)
  - 3) Radiographic evidence of a schwannoma *and* a first-degree relative with definite schwannomatosis
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