Case Report

Ancient Schwannoma Misdiagnosed as a Hemangioma in the Ventral Tongue

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Abstract

Schwannomas originate from the Schwann cells in the neural sheath of the peripheral nerves. Ancient schwannoma is one of five variants, and its characteristics include histopathological degeneration and diffuse hypocellular areas. Histopathological features show degenerative changes and atypical nuclei can easily be confused with malignant neoplasms. These cellular atypisms are caused solely by degenerative changes. Ancient schwannomas have been reported 17 cases in the oral cavity and five cases in the ventral tongue, including the floor of the mouth. We report a new case of an ancient schwannoma, misdiagnosed as a hemangioma with a 10-year evolution, located in the ventral tongue of a 29-year-old female.

Key words: Ancient schwannoma, Mouth floor, Hemangioma

Introduction

Schwannomas, known as neurilemmomas, are benign neural tumors, which develop rarely in the neural sheath of the Schwann cells of the peripheral, cranial, or autonomic nerves[1]. Approximately 25% to 45% of schwannomas develop in the head and the neck, and these mostly occur in people who are in their 40s. The incidence rate of the lesions in the oral cavity is only 1%;[2,3] Typically, schwannoma is attached to the nerve from which it originates, does not cause any pain, and grows slowly. It is difficult to diagnose this in the early stages because it lacks symptoms. Pain or neurological symptoms are mostly influenced by the mass itself[3]. Histopathologically, schwannoma is divided into the following variants: common, plexiform, cellular, epithelioid, and ancient schwannomas. An ancient schwannoma is a rare variant first described by Ackerman and Taylor[4] in 1951 and is characterized by degenerative changes and diffused hypocellular area. These changes were considered to be the result of a long-term degenerative alteration, and the term ‘ancient schwannoma’ was assigned. Schwannomas that show these degenerative changes have the potential to be misdiagnosed as sarcomas or other soft tissue neoplasms. In the present study, we report an ancient schwannoma misdiagnosed as a hemangioma which developed in the ventral tongue.

Case Report

A 29-year-old female patient visited our department due to discomfort caused by a mass in the lower left side of the tongue. The patient reported that the mass in the oral
cavity had appeared about 10 years earlier and had been slowly growing in size.

A 3 × 3 cm protruding mass was observed on the left side of the ventral tongue during an intraoral examination, This mass was firm, spherical, and had overlying normal mucosa (Fig. 1). The patient did not show any pain or dysphagia, and there was no impairment of tongue movement or paralysis.

![Fig. 1. Intraoral photograph showing the spherical movable mass in the left ventral tongue. The swelling was covered by normal mucosa. (A) On normal position. (B) Tongue elevation.](image1)

![Fig. 2. Magnetic resonance imaging shows an enhanced well-demarcated tumor in the left ventral tongue. (A) Well-encapsulated isointense lesion on the T1-weighted images. (B) Strong heterogeneous enhancement in the central mass on the T2-weighted image.](image2)
Magnetic resonance imaging (MRI) showed an ovoid-shaped 2.3×3.3×3.0 cm mass on the left side of the tongue, with a well-defined boundary. On the T2-weighted image, the inside of the lesion showed a heterogeneous signal intensity and a multifocal hypointense area considered to be indicative of the internal vascular structure. In addition, the peripheral portion of the mass exhibited a multifocal enhancement on contrast-enhanced imaging.

Based on the MRI finding, it was determined to be a benign hemorrhagic mass and was diagnosed as possibly being a benign hemangioma (Fig. 2). Needle aspiration was carried out, and fluid containing dark-colored blood was obtained from the lesion.

An angiography was conducted to determine whether the hemangioma contained an arterial component. The angiogram revealed tumor-feeding vessels in the left lingual artery, and embolization was performed in attempts to reduce the size of the lesion. Additionally, simple, direct puncture sclerotherapy with a sclerosing agent (Thromboject 3%; Omega Laboratories Ltd., Montreal, QC, Canada) was performed two times every week to shrink the mass (Fig. 3). The patient returned for a follow-up visit one month later, and no clinical changes were observed. The mass was considered to be a type of benign tumor other than a hemangioma, and an incisional biopsy was performed. The results revealed a benign fibroblastic proliferative lesion.

Excisional biopsy was then performed under general anesthesia. The mass was relatively easily separated from the surrounding tissue. The boundaries were clear, and neither high intraoperative bleeding nor the originating nerve was found. The size of the completely excised mass was 3.4×2.6×2.4 cm (capsule thickness: about 2.5 mm) and an oval-shaped firm mass covered in a tough yel-

![Fig. 3. Embolization was performed after angiographic confirmation. Lingual artery was the tumor-feeding vessel. (A) Coronal image. (B) Sagittal image.](image)

![Fig. 4. (A) Macroscopic appearance of the tumor shows a fully encapsulated, firm, yellowish-white mass measuring 3.4×2.6×2.4 cm. (B) The cut surface revealed myxomatous changes, accompanied by cystic degeneration and hematomas.](image)
low-white capsule was observed. In the cut surface, the internal side was relatively firm and red or yellow in color, and some myxoid changes and hematomas were observed (Fig. 4).

In the microscopic examination, an Antoni type-A area with highly ordered cellularity and a less-ordered cellular Antoni type-B area were observed (Fig. 5). Hyalinization and focal nuclear atypia with no mitotic figures were observed as degenerative changes (Fig. 6). Immunohistochemical studies revealed that the lesion was S-100 protein positive (Fig. 7). Based on these histopathological features, the lesion was diagnosed as an ancient schwannoma.

The patient recovered well after the surgery. Mild hypoesthesia was present on the left side of the tongue, but the site was observed to be healed well.

**Discussion**

Schwannoma develops in the somatic, cranial, or sympathetic nerves that are covered in the Schwann cell sheath. It does not develop in the olfactory or optic nerves because these nerves do not have the Schwann cells. It is most commonly found in the vestibulocochlear nerve[5]. Only
1% of schwannomas develop in the oral cavity, and the lesion is most often found on the tongue or the floor of the mouth. It can occur at any age, but it is mostly common in people in their 30s and 40s[6].

Clinically, patients do not experience severe discomfort for a long time because the lesion usually has no symptoms and grows slowly, although it does show submucosal swelling. Sometimes, the mass presses down on the neighboring nerve and causes neurological changes or pain[7]. Hwang et al.[8] stated that traumas, such as tongue biting, boring nerve and causes neurological changes or pain[7].

Ancient schwannoma generally has cystic areas and can be radiologically misdiagnosed as malignant fibrous histiocytoma, a malignant peripheral nerve sheath tumor, liposarcoma, synovial sarcoma, or hemangiopericytoma[9]. However, it is not usually misdiagnosed as a hemangioma.

MRI is the most useful tool for a differential diagnosis of the soft tissues of the oral cavity. From an MRI, schwannomas appear as soft, well-bounded, isointense lesions on T1-weighted images. The central area of the lesion shows hypointensity and the peripheral areas shows hyperintensity on T2-weighted images. This inhomogeneous appearance is more pronounced in larger schwannomas because some parts of the mass shows cystic degeneration in the core area, as the lesion does not receive any blood supply as it grows[10]. Both Antoni type-A and Antoni type-B areas are enhanced by an intravenous contrast medium, and the degeneration areas are shown to be non-enhanced[11].

On the T1-weighted image, a hemangioma usually has an iso-to-slightly high signal intensity. On the T2-weighted image, it has heterogeneous intensity with a pattern of lobulations, septations, and central low-intensity dots[12]. In particular, the areas with fast blood flow show multiple low signal intensities and a serpentine appearance[9].

In this case, the preoperative MRI results pointed to a hemangioma. The characteristic features of ancient schwannomas on MRI images, such as a multifocal hyperintense portion on the T2-weighted image, explain this early confusion and the misleading diagnosis of a hemorrhagic lesion.

Rarely, schwannoma has bizarre spindle cells in spite of the lack of mitotic figures and a large cystic myxomatous area. These atypical nuclei are caused by secondary degenerative changes, which are one of the histological characteristics of ancient schwannoma.

Clinically, ancient schwannoma appears encapsulated and well demarcated from the surrounding tissues. Histologically, Antoni type-A and Antoni type-B patterns and characteristic degenerative changes must be present. Secondary degenerative changes include hyalinization and mucoid deposition, cyst formation, increased blood vessels, hemorrhage, and calcification. Long-term schwannoma without symptoms was once observed with the 'ancient' variant[13]. However, ancient schwannomas are clinically difficult to differentiate from typical schwannomas; therefore, a histological evaluation is required[4]. In this case, the long-standing tumor (10 years) showed the secondary characteristics of ancient schwannoma.

Table 1. Reported cases of ancient schwannoma in the oral cavity, including the presented case

<table>
<thead>
<tr>
<th>No.</th>
<th>Author</th>
<th>Age/Sex</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Eversole and Howell[15]</td>
<td>58/F</td>
<td>Floor mouth and ventral tongue</td>
<td>2.5</td>
<td>Unknown</td>
</tr>
<tr>
<td>2</td>
<td>Marks et al. (1976)</td>
<td>65/F</td>
<td>Right floor of mouth</td>
<td>3.5</td>
<td>Unknown</td>
</tr>
<tr>
<td>3</td>
<td>McCoy et al. (1983)</td>
<td>36/F</td>
<td>Maxillary posterior vestibule</td>
<td>2.0</td>
<td>Unknown</td>
</tr>
<tr>
<td>4</td>
<td>Dayan et al. (1989)</td>
<td>52/F</td>
<td>Left Maxillary vestibule</td>
<td>0.9</td>
<td>Unknown</td>
</tr>
<tr>
<td>5</td>
<td>Nakayama et al. (1996)</td>
<td>40/F</td>
<td>Floor of mouth and ventral tongue</td>
<td>5.5</td>
<td>2 months</td>
</tr>
<tr>
<td>6</td>
<td>Ledesma et al. (1999)</td>
<td>21/F</td>
<td>Floor of mouth and ventral tongue</td>
<td>3.0</td>
<td>5 months</td>
</tr>
<tr>
<td>7</td>
<td>Chen et al. (2006)</td>
<td>34/M</td>
<td>Floor of mouth</td>
<td>3.0</td>
<td>18 years</td>
</tr>
<tr>
<td>8</td>
<td>Tobita et al.[17]</td>
<td>60/M</td>
<td>Buccal mucosa</td>
<td>2.0</td>
<td>23 years</td>
</tr>
<tr>
<td>9</td>
<td>Subhashraj et al.[17]</td>
<td>18/M</td>
<td>Mandibular posterior vestibule</td>
<td>3.1</td>
<td>8 months</td>
</tr>
<tr>
<td>10</td>
<td>Salehinejad et al. (2009)</td>
<td>40/M</td>
<td>Mandibular gingiva</td>
<td>1.9</td>
<td>3 years</td>
</tr>
<tr>
<td>11</td>
<td>Amirchaghmaghi et al. [18]</td>
<td>14/M</td>
<td>Mandibular gingiva</td>
<td>1.5</td>
<td>1 year</td>
</tr>
<tr>
<td>12</td>
<td>Biliçi et al. (2011)</td>
<td>45/M</td>
<td>Tongue tip</td>
<td>3.0</td>
<td>Unknown</td>
</tr>
<tr>
<td>13</td>
<td>Humber et al.[13]</td>
<td>82/F</td>
<td>Upper right lip</td>
<td>2.0</td>
<td>2 years</td>
</tr>
<tr>
<td>14</td>
<td>Kim et al.[16]</td>
<td>66/F</td>
<td>Mandibular posterior vestibule</td>
<td>2.0</td>
<td>13 years</td>
</tr>
<tr>
<td>15</td>
<td>Kim et al.[16]</td>
<td>35/F</td>
<td>Left mandible body</td>
<td>3.0</td>
<td>Unknown</td>
</tr>
<tr>
<td>16</td>
<td>Gainza-Cirauqui et al.[14]</td>
<td>35/F</td>
<td>Hard palate</td>
<td>2.0</td>
<td>5 years</td>
</tr>
<tr>
<td>17</td>
<td>Lee et al. (2013) (the present case)</td>
<td>29/F</td>
<td>Ventral tongue</td>
<td>3.3</td>
<td>10 years</td>
</tr>
</tbody>
</table>

F, female; M, male.
degenerative changes that are observed in previous reports[14]. The histological findings showed hyalinization and focal nuclear atypia with no mitotic figures, and the case was finally diagnosed as ancient schwannoma.

Ancient schwannoma that develops in the oral cavity was first reported by Eversole and Howell[15] in 1971. Sixteen cases of the disease were later reported in the English-language literature; however, only five cases were reported in the floor of the mouth and the ventral tongue[16]. The average age of the patients was 44 years (ranging from 14 to 82 years). The longest duration of the disease was 23 years[17]. In the current case study, the duration was 10 years (Table 1)[7,13-18].

Ackerman and Taylor[4] argued that ancient schwannoma has a diffuse cellular overgrowth due to the initially increased vascularization; it then causes hyalinization due to the subsequent decrease in vascularity. Histologically, the areas that show hypercellularity and nuclear atypia can be misdiagnosed as malignant. Specifically, it can be diagnosed as a malignant lesion by pathologists who are examining the frozen section of the mass[19].

To date, no ancient schwannomas have been reported to have malignant transformation in the oral cavity. However, in some cases, ancient schwannomas were reported to show malignant transformation in other anatomical locations; thus requiring attention[20].

Intraoral ancient schwannomas can be removed by conservative surgical excision. If complete excision is possible, the prognosis is good, with almost no recurrence. If the tumor is well encapsulated, surgical removal can be conducted easily. However, if the tumor is adhered to the nerves, careful dissection is required. In the present case, the originating nerves were not found, and the encapsulated mass was easily removed. After surgery, the patient had mild hypoesthesia but is showing good sensory recovery.

In conclusion, we have presented a rare case of an intraoral ancient schwannoma on the ventral surface of the tongue, which was initially misdiagnosed as a hemangioma. Hemangioma like lesion with cystic area showing heterogenous intensity on MRI should be considered in the differential diagnosis of ancient schwannoma.

References

4. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax: A clinicopathological evaluation of forty-eight cases, Cancer 1951;14:669-91.