Plexiform schwannoma of the floor of the mouth: a case report

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Abstract
Plexiform schwannoma is a type of schwannoma that can be occurred at any age and any site of the oral cavity. In the gross and histopathological features, it shows plexiform or multinodular growth pattern. This variant of schwannoma is sometimes associated with neurofibromatosis type II or schwannomatosis. Histopathologically, plexiform schwannoma is composed of schwann cells that show Anthony A growth pattern. Mitosis are rare or absent. This study reports a case of a healthy 34 year old female with swelling in floor of her mouth; according to the clinical and histopathological findings, the diagnosis was Plexiform schwannoma.

Keywords: Schwannomatosis, Plexiform, Neurofibromatosis type 2, Schwannoma


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شوانومایی پلکسی فرم در کف دهان: گزارش مورد

محمد کوچک زفولی، مریم سیدمجیدی، شبنم سوهانیان، حامد حسینی، کاظمی، خشایار دهشیری

چکیده
شوانومایی پلکسی فرم و ارتباطی از شوانومایی که در هر سنی و هر ناحیهی آن از جلوه دهان امکان مشاهده‌اند وجود دارد و این شوانومایی از نظر نمای gross و استئوپاتولوژی الگویی رنگ‌داده‌ی یک‌پیکسی فرم یا مولتی‌دولار تشکیل‌داده‌ی که گاهی در ارتباط با گروینگ فیبرو ماتورس تیپ II و یا شوانومایی‌تری به مردی می‌شود. از لحاظ استئوپاتولوژی‌کی، شوانومایی پلکسی فرم یا سوله‌سای شوان تشکیل‌شده‌که شان می‌دهد می‌توان نادر و یا اینکه اصل‌اکه‌نی به این شده‌این مطالعه به گزارش مورتری در خانم ۳۲ ساله‌ی سالمی با شکافی از قسم کف دهانی می‌پردازد که در اساسی‌تیه‌های بالینی و استئوپاتولوژی‌برای آن ضعیف، تشخیص شوانومایی پلکسی فرم داده شد.

واژگان کلیدی: شوانومایی‌تری، پلکسی فرم، نورفیبروماتوزی تیپ ۲، شوانومایی

Introduction

Schwannoma or neurilemoma is a benign neoplasm with neural origin derived from schwann cells. Head and neck involvement of this neoplasm is 25-48% and can appear at any age, although the most common site of involvement is the oral cavity, it may be seen in any area of the oral cavity.[1-3] Histopathologically, there are various types of schwannomas including cellular schwannoma, plexiform schwannoma and reticular (microcytic) schwannoma. [4] Cellular schwannoma mainly consists of spindle cells arranged in storiform or nonspecific pattern. In microcystic variant, cystic structure can be seen. Plexiform schwannoma is rare variant of schwannoma, introduced by Harkin and Redd for the first time in 1978 [5] and in the gross and histopathological feature, it shows plexiform or multinodular growth pattern. [1, 4, 5, 6] This variant of schwannoma is sometimes associated with neurofibromatosis type II (NF2) or schwannomatosis [4, 7] and in terms of histopathology, plexiform schwannoma which shows Anthony A growth pattern is composed of schwann cells. Mitosis is rare or absent. Like all types of schwannoma, this tumor is positive for S100 protein. EMA is positive around perineurium briefly. [8] The purpose of this paper was to introduce plexiform schwannoma in oral cavity of a patient with no particular syndrome and this was very significant.

Case report

A 34-year-old female patient with swelling in the floor of the mouth referred to the Oral Medicine Department of Babol University of Medical Sciences. This lesion has been appeared in the floor of the mouth four years ago and its size has gradually increased. In intraoral examination, there was an exophytic mass which was sessile and covered with intact mucosa with the same color. It was 1/5×1×0/5 cm³ lesion in the floor of the mouth on the left side of the midline (figure1). The patient did not have any disorders in submandibular gland. The patient had no marked medical history, but a history of rheumatism.

Figure 1. Clinical feature
The excisional biopsy was done and fibroma, neurofibroma and lipoma were considered as clinical differential diagnosis. The sample in formalin included a half oval-shaped soft tissue which was firm and white-creamy in color. The size was 1/2×1×0/2 cm³ that was solid, uniform, cream-colored and lobular in section. In the microscopic examination, benign neoplastic proliferation of schwann cells that formed multiple neurological fascicles was observed. Fascicles have spherical and elliptical structures with storiform pattern. The lesion was unencapsulated but limited and its surface was covered by stratified squamous epithelium of oral mucosa (figure 2). According to the histopathologic view, the diagnosis was plexiform schwannoma.

**Figure 2.** Histopathologic feature, A-multiple neurological fascicle in storiform pattern were covered by stratified squamous epithelium (×10), B-Fasicles had spherical and elliptical structures (×400), C and D-Antoni A pattern (×400)

**Discussion**

Schwannoma or neurilemoma is a benign neoplasm with neural origin derived from schwann cells. This is an encapsulated slow-growing tumor that is usually associated with nerve trunk and appeared as an asymptomatic mass at any age. The most common site for oral schwannoma is the tongue, but it may involve any area of the oral cavity. There are also more intraosseous cases seen in the posterior part of the mandible and in radiography, it is observed as unilocular or multilocular radiolucency. Intraosseous tumors have pain and paresthesia. [1-3] Schwannoma may be a part of an inherited disorder, called neurofibromatosis type II (NF₂). NF₂ is an autosomal dominant inherited disorder related to the mutations in the NF₂, which is tumor suppressor gene located on the chromosome 22 and produces Merlin protein. It is necessary to mention that the bilateral vestibular nerve schwannoma, progressive sensoneural deafness, dizziness, tinnitus as symptoms of this genetic disorder. Neurofibroma and café-au-lait pigmentation are sometimes seen. Schwannomatosis is another disorder which is associated with the mutation of SMARCB1 on chromosome 22. One of the characteristics of this disorder is multiple painful schwannomas in different areas, but there is not auditory nerve vestibular involvement. [1] Microscopic feature of schwannoma is
**Plexiform schwannoma**

An encapsulated tumor with two types of growth patterns. Antoni A pattern includes the areas of spindle cells organized in palisaded whors and waves. These cells often surround an acellular eosinophilic zone which is named Vero cay body, representing reduplicated basement membrane and cytoplasmic cellular processes. The other pattern is the so-called Antoni B tissue, consisting of spindle cells haphazardly distributed in a delicate fibrillar microcystic matrix.  

Microscopically, schwannoma is divided into cellular schwannoma, plexiform and reticular (microcytic) types.  

Plexiform schwannoma is a variant of schwannoma, which shows Anthony A pattern composed of schwann cells. Few mitosis is usually seen or may not exist at all. Like the other types of schwannoma, this tumor is positive for S100 protein. EMA is positive around Perineurium briefly. Rare cases of this type are observed in the literature review.

Table 1. Intraoral plexiform schwannoma reported in PubMed since 1994

<table>
<thead>
<tr>
<th>No.</th>
<th>Author</th>
<th>Year</th>
<th>Location</th>
<th>Sex</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Krolls et al</td>
<td>1994</td>
<td>Hard palate</td>
<td>Female</td>
<td>21</td>
</tr>
<tr>
<td>2</td>
<td>Lobo et al</td>
<td>2009</td>
<td>Lip mucosa</td>
<td>------</td>
<td>----</td>
</tr>
<tr>
<td>3</td>
<td>Vera-Sempere et al</td>
<td>2010</td>
<td>Mandible</td>
<td>------</td>
<td>----</td>
</tr>
<tr>
<td>4</td>
<td>Nisa et al</td>
<td>2011</td>
<td>Tongue</td>
<td>Female</td>
<td>38</td>
</tr>
<tr>
<td>5</td>
<td>Kapetanakis et al</td>
<td>2012</td>
<td>Soft palate</td>
<td>Female</td>
<td>21</td>
</tr>
<tr>
<td>6</td>
<td>Al-Mahdi et al</td>
<td>2012</td>
<td>Tongue and mandibular region</td>
<td>------</td>
<td>27</td>
</tr>
<tr>
<td>7</td>
<td>Lambado et al</td>
<td>2013</td>
<td>Intramaxillary</td>
<td>------</td>
<td>----</td>
</tr>
<tr>
<td>8</td>
<td>Couto et al</td>
<td>2014</td>
<td>Hard palate</td>
<td>Female</td>
<td>14</td>
</tr>
<tr>
<td>11</td>
<td>Present case</td>
<td>2017</td>
<td>Floor of the mouth</td>
<td>Female</td>
<td>34</td>
</tr>
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</table>
Malignant peripheral nerve sheath tumor, palisaded encapsulated neuroma, plexiform neurofibroma, lipoma, hemangioma, eosinophilic granuloma, epidermoid and dermoid cysts, epithelial hyperplasia, granular cell tumor, benign tumors of the salivary glands, rhabdomyoma, leiomyoma, lingual thyroid, mucocele and lymphangioma were considered as differential diagnosis of plexiform schwannoma in several studies. MPNSTs have mitotic activity and metastasis. PEN contains axons that can be seen by silver staining. Plexiform neurofibromas are less cellular, less S100 immunoreactive and have a mucin-rich matrix. Plexiform schwannoma can be easily differentiated from the other lesions mentioned above by the microscopic and immunohistochemical studies.

In some cases, the malignant processes of cancer such as squamous cell carcinoma and sarcoma show similar clinical features and in these cases the accurate diagnosis is crucial, because plexiform schwannoma has a benign process and is treated by an excisional surgery.

References