Case Report
Progesterone receptor expression in sinonasal leiomyoma: a case report and review of the literature

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Abstract: Leiomyomas are benign myogenic tumors that seldom occur in the sinonasal cavity. They were commonly found in middle-aged adults with a female predominance. Clinical symptoms include nasal obstruction with discharge, nasal bleeding and pain. We describe the case of a 48-year-old woman with a leiomyoma arising from right inferior nasal turbinate. Transnasal endoscopic excision was performed with satisfied result. The tumor was found to be progesterone receptor positive on immunohistochemical analysis. The clinical findings are addressed with a review of the literature. To the best of our knowledge, this is the third reported case that supports the growth of the tumor may be hormone-dependent.

Keywords: Paranasal sinuses, nasal cavity, angioleiomyoma, sex hormone-binding globulin receptor, progesterone receptor, intranasal surgery

Introduction

Sinonasal leiomyomas are extremely rare. Barnes reported that less than 1% of all leiomyomas were found in the head and neck area and within these, only 3% occurred in the nasal cavity [1]. The paucity of smooth muscle in the nasal cavity might explain why the tumors were rare. Since the first report of an intranasal leiomyoma by Maesaka et al in 1966, about sixty subsequent cases had been described in the English-language literatures. We present a new case of sinonasal leiomyoma that immunohistochemical results suggest the tumor may be hormone-dependent with focal positivity to progesterone receptor (PR). The clinical characteristics, histopathological findings, the role of sex hormones in tumor growth and treatment options were discussed.

Case report

Clinical presentation

A 48-year-old woman presented with nasal mucopus and occasional blood-tinged discharge for three months. She was otherwise healthy before and denied any nasal trauma or surgical history. Nasopharyngoscopic survey revealed some yellowish mucopus over bilateral middle meatuses and a 1 cm x 1 cm sized mass located near anterior third of right inferior nasal turbinate. No other abnormality was found in the nasopharynx, oral cavity, larynx and neck. Computed tomography (CT) scan of nose and paranasal sinus showed a homogeneous soft tissue mass originating from right inferior nasal turbinate and mucoperiosteal thickenings with fluid level in bilateral maxillary sinuses (Figure 1). The lesion was completely removed endoscopically under general anesthesia. Postoperative recovery was uneventful. There was no evidence of recurrence four years after surgery.

Gross and histopathology

Grossly, the resected specimen was brownish and elastic. Microscopically, a submucosal tumor consisted of interlacing bundles of bland-looking spindle cells and many thick-walled vessels was seen. The spindled cells had elongated vesicular nuclei with blunt ends, perinuclear vacuoles and eosinophilic cytoplasm (Figure 2). Immunohistochemical analysis showed strong positivity for smooth muscle actin and focal
Progesterone receptor in sinonasal leiomyoma

nuclear positivity for PR (Figure 3A and 3B). Negativity for S-100 protein and estrogen receptor (ER) were also noted (Figure 3C). The feature was consistent with a benign vascular leiomyoma.

Discussion

Sinonasal leiomyoma, a benign neoplasm, is rarely found in the head and neck region. It can be thought as a middle-aged disease with most patients diagnosed in their fifth and sixth decade [2-5]. A female to male ratio of 2 to 1 was noticed [2]. More tumors were found in the right sinonasal cavity, but the reason remained unknown [5]. Inferior nasal turbinate, nasal septum and nasal vestibule were the most common sites of occurrence [2, 3, 5]. The presenting symptoms were nonspecific and usually characterized by nasal obstruction, epistaxis and nasal discharge. These symptoms resulted from the mass effect related airflow change which caused nasal crusting and nasal desiccation. Local pain wasn’t that common in sinonasal leiomyoma.

The etiology of sinonasal leiomyoma remains uncertain. Marioni et al hypothesized the tumor growth might be hormone-dependant due to a female predominance and the increased pain during pregnancy or the menstrual cycle in patients described elsewhere [3]. They published the first case of nasal vascular leiomyoma with immunopositivity for PR and negativity for ER in a 70-year-old woman. He et al reported another case with the same immunoreactivity and suggested the role might also work even in a male patient [6]. However, there were three cases presented totally different results with negativity to both ER and PR [7-9]. The present case had similar result to Marioni et al; the
immunohistochemical study showed positivity for PR and negativity for ER. To our knowledge, this is the third case that supports this hormone hypothesis (Table 1) [3, 6-9]. Furthermore, all three cases showed immunopositivity to PR had symptom of nasal bleeding while the other three didn’t. Maybe the expression of PR could influence the clinical performance. The association between the tumor and the sex steroid receptor should be clarified.

Most authors accepted three hypotheses to explain the origin of the sinonasal leiomyoma. They could stem from aberrant undifferentiated mesenchymal or smooth muscle elements in the blood vessel wall [10]. In the nasal vestibule, they might arise from erector pilae or sweat gland muscles [11]. The observation of many tumors located in nasal turbinates seemed to support the hypotheses; these areas had relatively abundant blood vessels and smooth muscles. But some authors suggested that vascular leiomyoma could be a vascular malformation or a progressive development of smooth muscle proliferation from hemangioma, to an angioma with much muscle, to a leiomyoma with many vessels, and to a solid leiomyoma [12].

World Health Organization (WHO) classified leiomyoma into three groups: nonvascular leiomyoma (solid leiomyoma), vascular leiomyoma (angiomyoma), and epithelioid leiomyoma (atypical or bizarre leiomyoma and leiomyoblastoma). Morimoto divided angiomyoma into three histologic subtypes in 1973: solid or capillary, cavernous, and venous. The vascular subtype is the most common one and angiomyoma of head and neck usually present as venous or cavernous type [12]. Microscopically, interlacing bands of spindle-shaped cells with cigar-shaped nuclei are usually seen. The differential diagnosis includes hemangioma, angiofibroma, hemangiopericytoma, myofibroma and leiomyosarcoma. Immunohistochemical staining such as muscle specific actin, desmin, myoglobin, S-100 protein, vimentin can be of additional value [2, 12].

There was no characteristic image finding for preoperative diagnosis, but CT and magnetic resonance imaging (MRI) could provide the information about the extent of the lesion, bony erosion and sometimes differentiate the neoplasm from inflammatory changes [13]. A tissue biopsy might be performed before surgery, but the accuracy of preoperative diagnosis seemed to be doubtful. In three cases with preoperative biopsy, the results turned out to be the hemangiopericytoma, inflammatory tissue and hemangioma [2, 14, 15]. Therefore, the final diagnosis depended on surgical excision with histopathological study.
The most satisfactory treatment for sinonasal leiomyoma is complete surgical excision. The surgical approach depends on the size, location, the extension of tumor and the experience of the surgeon. In most cases, transnasal endoscopic excision can be performed successfully because many of them were limited in sinonasal cavity. For case in certain site or larger and more extended one, transpalatal, Caldwell-Luc, lateral rhinotomy, external ethmoidotomy with medial maxillectomy or Craniofacial resection is also of the choice.

The prognosis of sinonasal leiomyoma is favorable. No malignant transformation of sinonasal leiomyoma had been published before. The recurrence is rare after complete excision. Yang et al. mentioned one patient with recurrent leiomyoma two years after surgery which was probably due to incomplete excision of tumor margin and adjacent bony shell [13]. The patient then received further surgical excision with no evidence of recurrence for 5 years. Thus, the chance of local recurrence is mainly related to incomplete excision at first surgery.

In conclusion, sinonasal leiomyoma is a middle-aged disease with a female predominance. Patients usually have nonspecific symptoms like nasal obstruction, epistaxis and nasal discharge. Mostly, they are vascular subtype and the tumor growth may be hormone-dependant. For those with immunopositivity to PR, there is a chance that symptom like epistaxis developed. CT and MRI provide useful information about the tumor and the surrounding structure. Complete surgical excision is recommended either by endoscopic or open procedure. The prognosis is favorable with rare local recurrence. Currently, no patient has been reported to have malignant change in previous surgical site after definite surgery.

Table 1. Sex hormone receptor expression in vascular leiomyoma of sinonasal cavity

<table>
<thead>
<tr>
<th>Authors</th>
<th>Sex</th>
<th>Age (yr)</th>
<th>Location</th>
<th>Side</th>
<th>Size (cm)</th>
<th>Symptoms</th>
<th>PR</th>
<th>ER</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marioni et al. (2002)</td>
<td>F</td>
<td>70</td>
<td>NVe</td>
<td>R</td>
<td>1.5</td>
<td>N-O, N-B</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Kim et al. (2004)</td>
<td>F</td>
<td>60</td>
<td>IT</td>
<td>R</td>
<td>2</td>
<td>N-D, Sneezing</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Chen et al. (2007)</td>
<td>M</td>
<td>88</td>
<td>IT</td>
<td>R</td>
<td>0.9</td>
<td>N-D, right hearing impairment</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>He et al. (2009)</td>
<td>M</td>
<td>58</td>
<td>IT</td>
<td>R</td>
<td>1.3</td>
<td>N-O, N-B</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Onesti et al. (2012)</td>
<td>F</td>
<td>46</td>
<td>NT</td>
<td>L</td>
<td>1.45</td>
<td>P-M, skin color change at high temperature</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Present case</td>
<td>F</td>
<td>48</td>
<td>IT</td>
<td>R</td>
<td>1</td>
<td>N-D, N-B</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

Abbreviations: ER: estrogen receptor; F: female; IT: inferior turbinate; L: left; M: male; N-B: nasal bleeding or bloody discharge; N-D: nasal discharge; N-O: nasal obstruction; NT: nasal tip; NVe: nasal vestibule; PR: progesterone receptor; P-M: protruding mass; R: right.

Disclosure of conflict of interest
None.

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