Angioleiomyoma of the Nasal Cavity
- A Case Report -

Key Words: Angioleiomyoma–Nasal Cavity–Steroid Receptor

Angioleiomyoma of the sinonasal area is an extremely rare benign neoplasm. To the best of our knowledge, only 26 cases have been described. Here, we report a case of angioleiomyoma arising in the nasal cavity of a 60-year-old woman. Microscopically, the tumor consisted of proliferating smooth muscle cells punctuated with thick-walled vessels with slit-like lumina. The tumor was negative for estrogen and progesterone receptor by immunohistochemical study. Further studies are needed to clarify whether the growth of this tumor is sex steroid-dependent.

CASE REPORT

A 60-year-old woman presented with a two-month history of intermittent rhinorrhea and sneezing. Endoscopic findings showed a polypoid tumor originating from the right inferior turbinate. The tumor was in close contact with the mucosa without evidence of ulceration. No other abnormalities in the nasopharynx, oral cavity, larynx or ears were described. Her medical and family histories were unremarkable. The lesion was completely removed under endoscopic control and local anesthesia. Grossly, the specimen was an ovoid, firm, pinkish gray mass, measuring 2 × 1.2 × 0.9 cm; its cut surface was solid and pale gray. Microscopically, the tumor consisted of a well-demarcated nodule of smooth muscle tissue punctuated with thick-walled vessels with partially patent lumina (Fig. 1). The smooth muscle cells had rod-like nuclei and showed no cellular atypia or mitoses. Many vessels were nested in the major portion of the lesion. The inner layers of the vessel wall smooth muscle were arranged in an orderly circumferential fashion whereas the outer layers merged with a less orderly background muscle fibers. The vessel walls lacked internal and external elastic laminae by elastic staining. Immunohistochemical studies produced a positive result for smooth muscle actin (1:300, Dako, Glostrup, Denmark) in the perivascular proliferating smooth muscle cells and in the muscular wall of thick blood vessels (Fig. 2A), and for factor VIII-related antigen (1:3, Dako, Carpinteria, CA, U.S.A.) in the endothelial cells of thick-walled vessels (Fig. 2B). The perivascular proliferating smooth muscle cells, the muscular wall of the thick blood vessels and endothelial cells showed negative immunoreactivity for the
estrogen receptor (ER) (1:50, Dako, Glostrup, Denmark) and for the progesterone receptor (PR) (1:50, Dako, Carpinteria, CA, U.S.A.). There was no evidence of recurrence during the 6-month postsurgical period.

DISCUSSION

Since Maesaka et al.\textsuperscript{4} first reported an intranasal angioleiomyoma in 1966, 26 cases have been reported,\textsuperscript{2,3} which demonstrates the rarity of this condition even taking underreporting into account. A review of literature indicated a predominance in middle-aged women and a predilection for the right side of the nasal cavity, as in the present case; however, the reasons are unknown. Though subcutaneous angioleiomyomas are commonly painful,\textsuperscript{1} tumors occurring in the nasal cavity are not known to be painful. Patients with nasal angioleiomyomas usually complain of epistaxis and nasal obstruction.\textsuperscript{2} These tumors are histologically classified into three types: capillary or solid, cavernous, and venous. In the extremities, the tumors are mainly of the solid type whereas in the head and neck region, they are mainly venous.\textsuperscript{1} Our case, which arose in the nasal cavity, was also of the venous type. Surgical excision is recommended as treatment. The prognosis of nasal angioleiomyoma is usually favorable. A case of nasal angioleiomyoma extending into the nasopharynx and the anterior cranial fossa showed no evidence of recurrences until 30 months after the operation.\textsuperscript{5}

The origin of nasal angioleiomyoma is uncertain. Lesions may arise from smooth muscle elements in the nasal cavity or from embryonic tissue remnants. In the nasal cavity, smooth muscles are present as either piloerector muscle of the vestibule or in the

Fig. 1. The nasal tumor consists of a well-demarcated nodule of smooth muscle tissue punctuated with thick-walled vessels with slit-like lumina.

Fig. 2. (A) Immunohistochemical staining for smooth muscle actin shows a positive reaction in perivascular proliferating spindle cells and thick blood vessel walls. Thick-walled blood vessel shows a circumferential arrangement of the inner smooth muscle layer and less well-ordered outer layer blended with the background smooth muscle tissue of the tumor. (B) Immunohistochemical staining for factor VIII-related antigen shows a positive reaction in the endothelial cells of the thick-walled vessels.
walls of blood vessels elsewhere. Most of the cases reported in the sinonasal area have developed from the inferior or middle turbinate. Therefore, it seems more likely that any leiomyoma developing in the nasal cavity is of vascular origin, though a hamartomatous basis cannot be excluded. The vessels in these tumors are difficult to classify because they are not altogether typical of veins or arteries. Their thick walls and small lumina are reminiscent of arteries, but they consistently lack internal and external elastic laminae, as in our case.

Whether differential incidence is associated with the presence of ER and/or PR on smooth muscle cells is speculative, although some evidence exist to show a relationship between the presence of sex steroid receptors and the proliferation of smooth muscle cells. The prevalence of female cases of nasal angioleiomyoma and the increased pain during pregnancy and menstrual cycle described in cases of angioleiomyoma at several sites suggest that sex steroid receptors influence nasal angioleiomyoma development. Di Tommaso et al. reported that a focal positivity for PR was detected in 6 of 10 cases of subcutaneous angioleiomyoma and that the same cases showed no positivity for ER. Marioni et al. described a case of nasal angioleiomyoma with PR-positivity but ER-negativity by immunohistochemical study, and suggested that PR though expressed had no functional role or, alternatively, that PR was activated by a different pathway in nasal angioleiomyomas. However, in the present case, the tumor showed negative immunohistochemical findings for ER and PR. These discrepancies are probably due to the sex steroid receptors being expressed in only a small number of copies in mesenchymal tumors. The reason why nasal angioleiomyomas are not painful during pregnancy or the menstrual cycle might be related to the total number of cells expressing sex steroid receptors. Further studies using more cases are needed to clarify whether the growth of this tumor is sex steroid-dependent.

REFERENCES