Endoscopic Treatment of Sinonasal Leiomyosarcoma: A Case Report in Light of the Literature

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ABSTRACT
A 49-year-old Japanese man presented with a rare case of sinonasal leiomyosarcoma with left nasal bleeding for 12 months. He had no history of irradiation or malignancies, including retinoblastoma. Preoperative histological examination suggested vascular leiomyoma. Complete resection with endoscopic surgery was performed. Histological examination during the operation suggested that the tumor was a leiomyoma. However, immunohistochemical staining for α smooth muscle actin and desmin were helpful in establishing a definitive diagnosis of sinonasal leiomyosarcoma. The resection margins were positive for tumor cells. Staging with CT of the neck and thorax, ultrasound of the abdomen, and MRI of the head ruled out metastases. Second endoscopic tumor resection surgery was performed for positive resection margins. The patient’s condition was successfully managed with additional excision, and he remains well with no evidence of recurrence and metastasis 36 months after treatment. Endoscopic management should be considered in suitable cases.

Key words sinonasal leiomyosarcoma; endoscopic surgery; nasal bleeding

Sinonasal leiomyosarcoma is a rare type of tumor.1 Leiomyosarcoma is an aggressive mesenchymal tumor that shows smooth muscle differentiation by histology and immunophenotype.2 Common affected sites are uterus, skin and retroperitoneum, with only around 3% in the head and neck region.¹, ² Clinically, sinonasal leiomyosarcoma is associated with nasal obstruction and nasal bleeding.³ Histologically, leiomyosarcoma is made up of spindle cells forming fascicular, storiform, or whorled patterns showing immunoreactivity for α smooth muscle actin and desmin.⁴ The mainstay of treatment is complete surgical extirpation to avoid recurrence.¹–⁴ We report the case of a 49-year-old Japanese man who presented with left nasal bleeding. Complete resection with endoscopic surgery was performed. Diagnostic and therapeutic options are discussed in the light of the literature.

PATIENT REPORT
A 49-year-old Japanese man was referred to our otolaryngology department for evaluation of left nasal bleeding for 12 months. He had bilateral polisinecctomy and deviation at 22 years and endoscopic sinus surgery at 27 years, respectively, and no history of irradiation or malignancies, including retinoblastoma. Endoscopic revealed a soft, gray, elastic mass in the left nasal cavity (Fig. 1). Preoperative coronal and axial computed tomography (CT) of the paranasal sinuses showed a low-density, homogeneous lesion occupying the left nasal cavity (Fig. 2). Preoperative pathological examination was performed without massive bleeding and suggested vascular leiomyoma. The patient was admitted to our department for definitive diagnosis and treatment. First endoscopic resection of the tumor was performed under general anaesthesia. The tumor was resected with 0-degree-of-view, 4-mm-diameter rigid endoscopes. Histological examination during the operation suggested that the tumor was a leiomyoma. Since the tumor adhered to the nasal septum, it was resected from...
the bony portion of the septum. The volume of bleeding during surgery was 5 mL. Histologically, it consisted of a proliferation of atypical spindle-shaped cells showing spindled and oval nuclei with partially surrounding hyalinization and slightly branching vascular structures (Fig. 3). The tumor cells were positive for α smooth muscle actin and desmin (Fig. 3). In addition, tumor cells were immunohistochemically positive for vimentin, CAM5.2, EMA and bel-2 and negative for S-100, CD34, CD31 and p53. MIB-1 immunohistochemistry showed positive nuclear staining in approximately 40% of the tumor cells. According to the grading standardized by the Fédération nationale des Centres de lutte contre le cancer (France), tumor differentiation was Score 2, mitotic count was Score 3, and tumor necrosis was Score 0. The definitive diagnosis was sinonasal leiomyosarcoma, Grade 2 (intermediate grade). Following these findings, radical second endoscopic surgery was performed for positive resection margins. The right and left middle turbinate were first partially resected to obtain a clearer and well defined visualization of the safety margin. Second, the anterior upper portion of the nasal septum was resected, and multiple biopsies of surrounding

Fig. 1. Preoperative endoscopic photograph of the left nasal cavity occupied by a mass of about 25 mm × 20 mm × 30 mm. (* tumor, # middle turbinate, + nasal septum)

Fig. 2. (A, B, C) Preoperative coronal, axial and sagittal CT scan of the paranasal sinuses showed a low-density, homogeneous lesion occupying the left nasal cavity (A, B, C). The tumor had low signal intensity in the T1-weighted image and low to high signal intensity in the T2-weighted image (D, E). Axial MRI showed that the tumor had high signal intensity in the contrast-enhanced T1-weighted image indicating strong enhancement with poor enhancement in the center lesion of the tumor (F). Axial diffusion image indicated inhomogeneous low to high signal intensity lesion. (G) CT, computed tomography; MRI, magnetic resonance imaging.
neighbouring structures were taken to confirm the safety margin. The patient’s condition was successfully managed with excision of the tumor without further radiotherapy, and he remains well with no evidence of recurrence 36 months after treatment (Fig. 4).

DISCUSSION

Sinonasal leiomyosarcoma is a rare aggressive sarcoma that represents only 2% to 3% of non-epithelial neoplasia of the nasal cavity, paranasal sinuses and nasopharynx.\(^1\) Its most frequent localization is the nasal cavity.\(^2\) An association with previous therapeutic irradiation and hereditary retinoblastoma has been described.\(^1\)–\(^3\)

The diagnosis of sinonasal leiomyosarcoma is often difficult, as patients often present late, owing to the non-specific symptoms and non-specific clinical and radiological findings.\(^4\)–\(^5\) The presenting symptoms depend on the localization of the leiomyosarcoma within the sinonasal tracts.\(^6\) While involvement of
the nasal cavity, as in our case, presents usually with non-specific symptoms such as nasal obstruction and bleeding, involvement of the paranasal sinuses and skull base may present, with a range of symptoms from non-specific to severe functional impairment of the neighbouring cranial nerves. Symptoms include unilateral or bilateral nasal obstruction, rhinorrhoea, epistaxis, facial pressure, pain, postnasal drip and a reduction or loss in smell sensation. CT and MRI of the paranasal sinuses can help in determining the localization and the involvement of neighbouring structures before surgical intervention. Sinonasal leiomyosarcoma shows only moderate hyperintensity on both T1 and T2 weighted MRI, which does not provide additional diagnostic information. Imaging therefore may be more helpful in surgical planning than in diagnosis. A definitive diagnosis is made by histopathology and immunohistochemistry. Sinonasal leiomyosarcoma stains positive for smooth muscle actin and desmin and negative for myogenin, cytokeratin MNF116 and AE1AE3, and S100/Melan-A. Leiomyosarcoma tends to have aggressive local behavior characterized by local displacement of neighbouring structures rather than invasion. Metastasis of sinonasal leiomyosarcoma is not common, but vascular metastases are often more common than lymph node metastases. The workup must include imaging studies involving the lung, brain, and liver, to where sinonasal leiomyosarcoma metastasizes preferably. Because of its invasive nature, the treatment of choice is complete surgical resection, with adjuvant chemotherapy or radiotherapy reserved for patients with locally advanced, recurrent or metastatic disease. If the primary tumor is not resectable, neoadjuvant chemotherapy to shrink the tumor followed by palliative surgery or primary radiotherapy with or without chemotherapy has been tried. In our case, the tumor was located in the nasal septum and radical second endoscopic surgical resection could be performed, and following negative margin results, further radiotherapy was not selected. As endoscopic tumor excision is expected to result in clear margin excision, an open approach, especially with its better controlled removal of the nasolacrimal duct, should be considered. However, endoscopic surgery for tumors in the nasal cavity is still challenging because of the narrow working space, the angled, anatomically variable paranasal sinus and the proximity to the orbit structure, anterior ethmoid artery and skull base. Advances in imaging, surgical instrumentation, intraoperative navigation systems and multi-angle visualization might enable radical resection to be performed by endoscopic surgery.

However, several external surgical procedures, including the Denker, medial maxillectomy, lateral rhinotomy, and midfacial degloving approaches, have been used in advanced cases.

Prognosis is often poor owing to the late presentation because of the lack of specific symptoms. Moreover, achieving wide surgical margins for locally advanced disease is often difficult in the sinonasal region. Several factors associated with poor prognosis have been reported, including the size of the tumour, higher histological grading and positive surgical margins. The reported 5-year survival ranges from 88% in patients with well differentiated tumours to 53% for poorly differentiated tumours. It was reported that the disease specific mean survival rate was 66% and the disease specific mean survival period was 38.24 months in a review of the sinonasal leiomyosarcoma. Late recurrence has been reported despite clear surgical clearance with postoperative adjuvant radiotherapy. Long-term careful follow-up might be important in its management.

In conclusion, although sinonasal leiomyosarcoma is extremely rare, it should be considered in the differential diagnosis of masses in the nasal cavity even for perioperative benign pathological examination. Immunohistochemical staining for smooth muscle actin and desmin are helpful in establishing a definitive diagnosis. Endoscopic management should be considered in suitable cases.

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The authors declare no conflict of interest.

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