CASE REPORT

A rare case of intranasal vascular leiomyoma

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SUMMARY

Vascular leiomyoma (VL) is a solitary and rare form of leiomyoma that usually occurs in the skin or subcutaneous tissue of the lower extremities. Intranasal VL is extremely rare, probably due to the lack of smooth muscle in the nasal cavity. In this study, we report a case of a 70-year-old woman with VL of the inferior nasal turbinate. An endoscopic examination revealed a pinkish globular mass at the inferior turbinate. A preoperative CT scan exhibited a highly enhanced mass originating from the inferior turbinate, and haemangioma was suspected. The patient underwent complete excision of the mass endoscopically, and the histopathological report indicated that the mass was a VL. The tumour was determined to be negative for progesterone and estrogen receptors by immunohistochemical staining. The postoperative period was uneventful. There was no local recurrence during the 12-month follow-up period.

BACKGROUND

Vascular leiomyoma (VL; angioleiomyoma) is an uncommon smooth muscle tumour that is extremely rare in the nasal cavity. Less than 1% of all VLs occur in the nasal cavity.¹ To the best of our knowledge, only 34 cases of VL in the nasal cavity have been reported in the English literature.^{1–30} Recently, a small number of studies regarding the role of the sex hormone receptors and Epstein-Barr virus (EBV) in tumour genesis have been reported.^{2–7} In this study, we report a case of VL in the nasal cavity, provide a review of the English literature, and discuss the role of sex hormone receptors in VL and their relationship with EBV infection.

CASE PRESENTATION

A 70-year-old woman with a history of left nasal obstruction and intermittent spontaneous epistaxis was seen at the ear, nose and throat clinic. She was taking aspirin medication due to a carotid plaque. Beyond that, she had no unusual medical or family history. An endoscopic examination of the nasal cavity revealed a pinkish globular mass at the proximal anterior end of the inferior turbinate (figure 1). The mass had high vascularity over its surface and was attached to the proximal anterior end of the inferior turbinate. No masses were found in the other nasal cavity, and no abnormal nasopharyngeal lesions were observed. A CT scan of the nasal cavity and paranasal sinuses showed a highly enhanced $1.6 \times 1.2 \times 1.4$ cm mass in the anterior nasal cavity, originating from the inferior turbinate. Preoperatively, a nasal haemangioma was suspected based on a contrast-enhanced CT scan (figure 2A, B).

Figure 1 Diagnostic nasal endoscope image. Diagnostic nasal endoscopy photograph of the left nasal cavity showing a pinkish globular mass arising from the inferior turbinate.

DIFFERENTIAL DIAGNOSIS

A woman in her 70s presenting with intranasal mass may have a benign or malignant neoplasm. In the present case, a CT scan showed a highly enhanced mass in the anterior nasal cavity, originating from the inferior turbinate. Preoperatively, a high vascular tumour such as nasal haemangioma was suspected. The differential diagnosis of VL should include myopericytoma, fibromyoma, leiomyosarcoma, angiofibroma and fibroma.

TREATMENT

The patient underwent complete excision of the mass endoscopically under general anaesthesia. Bleeding occurred during the removal of the proximal origin site, but it was stopped by bipolar cauterisation without embolisation. No major intraoperative haemorrhage occurred. The histopathological report of the mass revealed VL. Microscopic examination revealed a tumour composed of many thick-walled vascular channels with inner coats of circumferentially arranged spindle cells (figure 3A). No atypia or mitosis was observed. Immunohistochemical examination revealed strong positivity for smooth muscle actin (SMA), desmin and vimentin in the perivascular cells (figure 3B–D). The Ki-67 proliferation index was up to 1% (figure 3E). The tumour was negative for progesterone receptor (PR) and estrogen receptor (ER). An in situ hybridisation study to detect EBV was negative.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful. There was no local recurrence during the 12-month follow-up period.

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Figure 2 Images of the axial (A) and coronal (B) CT scan showing a highly enhanced mass in the anterior nasal cavity that originated from the inferior turbinate.

Figure 3 Images of the resected mass and microscopic examination. (A) The tumour consisted of thick-walled vessels, with spindle cells surrounding it (H&E stain;×200). (B) The tumour cells were highly positive for smooth muscle actin (×200). (C) The tumour cells were highly positive for desmin (×100). (D) The tumour cells were highly positive for vimentin (×100). (E) The Ki-67 proliferation index was up to 1% (×200).

DISCUSSION

VL is a solitary and rare form of leiomyoma that usually occurs in the skin or subcutaneous tissue of the lower extremities.¹ VL in the nasal cavity is extremely rare, probably due to the lack of smooth muscle in the nasal cavity.¹ Hachisuga *et al*¹ described only 5 cases of 562 VLs located in the nasal cavity and did not describe the origin site of those VLs. No more than 34 cases of VL in the nasal cavity and paranasal sinuses have been reported in the English literature (table 1).^{1–30}

In the nasal cavity, VLs have been reported to arise most frequently at the inferior turbinate (13 of 30; 43.3%), probably due to the relatively abundant blood vessels and smooth muscle tissues in the inferior turbinate. Other sites of VLs in the nasal cavity that have been reported include the nasal septum (5 of 30; 16.7%), nasal vestibule (5 of 30; 16.7%), paranasal sinus (3 of 30; 10%), other turbinate (2 of 30; 6.7%) and nasal fossa floor (1 of 30; 3.3%). Only 1 of 30 cases was reported to have a VL in the entire nasal cavity, nasopharynx and anterior cranial fossa (table 1).⁸

Table 1 Vascular leiomyoma of the nasal cavity and paranasal sinuses reported in the English literature

Authors	Year	Country/race	Age/ sex	Symptoms	Site	PR	ER	EBV	Treatment	Outcome (follow-up period)
Maesaka <i>et al</i> ¹¹	1966	Japan	49/F	Facial pain	Vestibule	ND	ND	ND	Local excision	NED
Wolfowitz and Schmamman ¹²	1973	Africa	42/F	Epistaxis	IT	ND	ND	ND	Local excision	NED (2.5 years)
Schwartzman and Schwartzman ¹³	1973	Caucasian	57/M	NO, headache	SS,ES,MS	ND	ND	ND	Transantral ethmoid sphenoidectomy	NED (3 years)
McCaffrey <i>et al</i> ¹⁴	1978	USA	76/F	Epistaxis, NO	IT	ND	ND	ND	Excision	ND
Daisley ¹⁵	1987	Trinidadian	32/F	NO, headache	MT	ND	ND	ND	Excision (Caldwell-Luc approach)	ND
Hanna <i>et al</i> ¹⁶	1988	Caucasian	64/F	Epistaxis, NO, facial pain	IT	ND	ND	ND	Excision	NED (1 year)
Zijlker and Visser ¹⁷	1989	Caucasian	33/M	Periorbital swelling, NO, rhinorrhoea	ES	ND	ND	ND	Transnasal antrostomy, ethmoidectomy	NED (2 years)
Sawada ¹⁸	1990	Japan	41/F	Nasal mass	Vestibule	ND	ND	ND	Excision	NED (1 year)
Ragbeer and Stone ¹⁹	1990	MD	49/M	Epistaxis, facial pain	Nasal floor	ND	ND	ND	Excision	NED (1 year)
Harcourt and Gallimore ²⁰	1993	Caucasian	55/F	Epiphora	ES	ND	ND	ND	Patterson's external ethmoidectomy	NED (1 year)
Trott <i>et al</i> ²¹	1994	USA	43/F	Epistaxis, NO, PND	IT	ND	ND	ND	Excision	NED (1 year)
Khan <i>et al</i> ²²	1994	UK	71/F	NO	IT	ND	ND	ND	Excision	NED (1 year)
Ardekian <i>et al</i> ²³	1996	Israel	54/F	NO, nasal pain, rhinorrhoea	Septum	ND	ND	ND	Excision	ND
Nall <i>et al</i> ²⁴	1997	USA, African-American	43/F	Epistaxis, NO, facial pain headache	ST	ND	ND	ND	Embolisation and excision	NED (21 months)
Nicolai <i>et al</i> ⁸	1996	Italy	45/F	NO	Whole nasal cavity, anterior cranial fossa	ND	ND	ND	Excision via a combined craniofacial resection	NED (30 months)
Murono <i>et al</i> ²⁵	1998	Japan	69/F	Epistaxis	IT	ND	ND	ND	Endoscopic excision	NED (1 year)
Bloom <i>et al</i> ¹⁰	2001	USA	50/F	NO, headache	Septum	ND	ND	ND	Endoscopic excision (re-excision at 8 months)	NED(3 years)
Bloom <i>et al</i> ¹⁰	2001	USA	70/F	Asymptomatic	Septum	ND	ND	ND	Endoscopic excision	NED (6 months)
Marioni <i>et al</i> ²	2002	Italy	70/F	NO, epistaxis	Vestibule	+	–	ND	Excision	NED (3 months)
Kim <i>et al</i> ⁵	2004	Korea	60/F	Rhinorrhoea, sneezing	IT	–	–	ND	Endoscopic excision	NED (6 months)
Chen <i>et al</i> ⁶	2007	China	88/M	Rhinorrhoea	IT	–	–	–	Excision	NED (1 year)
Campelo <i>et al</i> ²⁶	2008	Brazil	44/F	Epistaxis, NO	IT	ND	ND	ND	Endoscopic excision	NED (10 months)
Singh <i>et al</i> ²⁷	2008	India	31/M	Epistaxis, NO	Septum	ND	ND	ND	Endoscopic excision	NED (6 months)
Vafiadis <i>et al</i> ²⁸	2008	Greece	68/M	NO	Vestibule	ND	ND	ND	Excision (intraoral incision)	NED (2 years)
He <i>et al</i> ⁴	2009	China	58/M	Epistaxis, NO	IT	+	–	–	Endoscopic excision	NED (1 year)
Michael and Shah ²⁹	2009	India	34/M	Epistaxis, NO	IT	ND	ND	ND	Endoscopic excision	ND
Navarro Junior <i>et al</i> ³⁰	2010	Africa	62/F	Epistaxis, NO, facial pain	Septum	ND	ND	ND	Endoscopic excision	ND
Kim <i>et al</i> ⁷	2013	Korea	58/F	NO, rhinorrhoea	Vestibule	–	–	ND	Endoscopic excision	NED (6 months)
Tseng <i>et al</i> ³	2014	Taiwan	48/F	Rhinorrhoea, epistaxis	IT	+	–	ND	Endoscopic excision	NED (4 years)
Present case	2014	Korea	70/F	Epistaxis, NO	IT	–	–	–	Endoscopic excision	NED (1 year)

EBV, Epstein-Barr virus; ER, estrogen receptor; ES, ethmoid sinus; F, female; IT, inferior turbinate; M, male; MD, Middle Eastern; MS, maxillary sinus; MT, middle turbinate; ND, no data; NED, no evidence of disease; NO, nasal obstruction; PND, postnasal drip; PR, progesterone receptor; SS, sphenoid sinus; ST, superior turbinate; UK, United Kingdom; USA, United States of America.

Table excludes five cases mentioned in a series by Hachisuga *et al*.¹

This tumour predominantly occurs in females (22 of 30; 73.3%) and is relatively common at middle age (mean age; 54.5 years). The most common symptoms are nasal obstruction, epistaxis and facial pain. The incidence of intranasal pain is higher for VLs of the nasal septum and vestibule (table 1). Preoperatively, it is very difficult to diagnose VL through clinical history and endoscopic findings. A correct diagnosis can be made by histological confirmation postoperatively. A preoperative CT scan can be helpful for determining extension, bony erosion and vascularity. Remarkable enhancement of the preoperative CT scan can reveal hypervascularity of the mass lesion and prevents dangerous events from occurring during the biopsy and aspiration.

The WHO histological classification of tumours divides leiomyomas into three groups: leiomyoma, angioleiomyoma (VL or angiomoma) and epithelioid leiomyoma (bizarre leiomyoma and leiomyoblastoma).^{9–10} Microscopically, VL consists of spindle-shaped smooth muscle cells and many blood vessels where the smooth muscle cells are concentrated. Immunohistochemistry for vimentin and SMA are useful for diagnosing this tumour.

The aetiology of VL in the nasal cavity remains unclear. Most authors have hypothesised the origin of VLs in the nasal cavity. It has been proposed that this tumour could originate from aberrant undifferentiated mesenchymal or smooth muscle elements in blood vessel walls. However, some authors have suggested that it could be a vascular malformation or a progressive development of smooth muscle proliferation from haemangioma to angiomoma, which is composed mostly of muscle cells, to a leiomyoma with many blood vessels and, finally, to a solid leiomyoma.³

Sex hormone receptors may influence the tumour genesis of smooth muscle tumours, such as uterine VL. Marioni *et al*² hypothesised that this tumour growth might be progesterone dependent due to its predominance in females and the fact that pregnancy and menstruation result in increased pain in these patients. Marioni *et al*² reported the first case of nasal VL that was immunopositive for PR. In the case reported here, the immunohistochemical studies for PR and ER were negative. To the best of our knowledge, six cases of VL were analysed for sex hormone receptors.^{2–7} Including our case, three of seven cases were immunopositive for PR (table 1). Whether or not sex hormones have a role in tumour genesis in nasal VL is still controversial, so more studies are necessary.

EBV infection is associated with smooth muscle tumours in immunocompromised patients such as patients with AIDS and patients with organ transplantation.⁶ Although the incidence and prevalence of sinonasal VLs are not known, they have been more commonly reported in East-Asian countries (China, Japan and Korea) and Central and South America (Brazil, Africa), which are areas where EBV infection is endemic (table 1). However, studies of the relationship between sinonasal VL and EBV infection are lacking (3 cases; table 1). Previously, studies of EBV in nasal VL cases were reported to be negative. Therefore, more studies regarding EBV infection in sinonasal VL cases are needed.

The differential diagnosis of VL should include myopericytoma, fibromyoma and leiomyosarcoma, angiofibroma and fibroma.

Complete surgical excision is the treatment of choice. The surgical approach depends on the tumour location and its extension into the surrounding tissue. Complete endoscopic resection is a good alternative treatment modality with a satisfactory recovery rate, except in cases of extensive invasion of VL. In larger and more extensive cases of VL, lateral rhinotomy, Caldwell-Luc, external ethmoidectomy with medial maxillectomy or craniofacial

resection are also other good treatment modalities. In one case of skull base invasion, extensive resection with craniofacial resection was performed.⁸ Attention should be paid to avoiding intraoperative and postoperative bleeding due to the hypervascularity of VL. Preoperative embolisation may be considered in cases of extensive invasion or hypervascular lesions. For our case, the endoscopic resection was successful without massive perioperative bleeding.

VLs are benign tumorous lesions that are capable of malignant transformation. Although malignant transformation of VL in the nasal cavity has never been reported, malignant transformation of VL at other sites, such as the hands, has, rarely, been reported.³¹ The malignant counterpart of VL, angioleiomyosarcoma, exists and exhibits dysplastic features histopathologically.

Recurrence of VL is extremely rare if the tumour is completely resected. Bloom *et al*¹⁰ reported a case of VL recurrence after incomplete resection; the mass was biopsied in the clinic, and the patient sneezed out the remaining tumour. Additionally, Hachisuga *et al*¹ reported two cases of VL recurrence. Therefore, complete resection is necessary to prevent recurrence.

In conclusion, although VL occurring in the nasal cavity is extremely rarely, it should be considered in the differential diagnosis of all vascular neoplasms of the nasal cavity.

Learning points

- ▶ Although vascular leiomyoma very rarely occurs in the nasal cavity, it should be considered in the differential diagnosis of all vascular neoplasms of the nasal cavity.
- ▶ The treatment of intranasal vascular leiomyoma is based on local complete excision. Complete resection endoscopically is a good alternative treatment modality with a satisfactory recovery rate.
- ▶ Attention should be paid to avoiding intraoperative and postoperative bleeding due to the hypervascularity of vascular leiomyoma.
- ▶ Sex hormone receptors, especially progesterone receptors, may influence the tumour genesis of the intranasal vascular leiomyoma.

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