CASE REPORT

Recurrent angioleiomyoma of the middle turbinate

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SUMMARY

We present a case of a middle-aged woman with a history of not only progressive nasal obstruction, facial pain, hyposmia and epistaxis, but also excision of the nasal mass diagnosed as a vascular leiomyoma. On examination, a smooth bulge was seen over the middle turbinate. Surgical excision along with histopathology and immunohistochemistry revealed a diagnosis of recurrent vascular leiomyoma of the middle turbinate.

BACKGROUND

Leiomyoma is a benign tumour of smooth muscle origin. Maeska¹ reported a case of leiomyoma of the nasal vestibule in 1966, which is the first reported leiomyoma in the nose in world literature. Approximately 50 cases of leiomyoma in the nose and paranasal sinuses have been reported in the world literature to date. Recurrent vascular leiomyoma of the nose and paranasal sinus has rarely been reported. We present a case of a middle-aged woman who presented with nasal obstruction, facial pain, hyposmia and epistaxis with a history of surgical excision of a nasal leiomyoma.

CASE PRESENTATION

A 69-year-old woman presented to the outpatient department with progressive left-sided nasal obstruction, left-sided frontal headache, hyposmia and intermittent moderate epistaxis of 2 years duration. She did not give a history of nasal discharge, postnasal drip and facial asymmetry. Her history revealed that she underwent an endoscopic excision

of mass which was diagnosed as a vascular leiomyoma 3 years before the onset of presenting symptoms. There was no history of leiomyomas at other sites in the body. Endoscopic examination revealed a reddish mass arising from the middle turbinate extending inferiorly up to the floor of the nasal cavity and anteriorly just up to the internal nasal valve (figure 1). General and systemic examination was normal.

INVESTIGATIONS

Contrast-enhanced CT (CECT) revealed a heterogeneously enhancing expansile soft tissue lesion arising from the middle turbinate measuring 4.3×2.3×3.4 cm obliterating the frontal recess and maxillary sinus mucociliary outflow. There was evidence of focal erosion of the nasal septum and the mass was found to be extending posteriorly into the choana and anteriorly 2.8 cm proximal to the external nares (figure 2). Biopsy from the soft tissue lesion revealed a leiomyoma; however, an immunohistochemical analysis was suggested to confirm the diagnosis.

DIFFERENTIAL DIAGNOSIS

- Haemangioma
- Haemangiopericytoma

Figure 1 Endoscopic image of angioleiomyoma of the middle turbinate. (A) Anterior lobulated part of the tumour. (B) Posterior lobulated part of the tumour. (C) Inferior turbinate. (D) Nasal septum.

Figure 2 Contrast-enhanced CT image showing a heterogeneously enhancing nasal soft tissue lesion with focal septal erosion (depicted by a black arrow).

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- ▶ Angiofibroma
- ▶ Fibromyoma
- ▶ Leiomyoblastoma
- ▶ Angiosarcoma
- ▶ Angiomyolipoma
- ▶ Leiomyosarcoma

TREATMENT

Endoscopic wide excision of the tumour with margin clearance was performed under general anaesthesia. Histopathological analysis of the specimen revealed nasal mucosa lined by a respiratory epithelium overlying a fibrocollagenous stroma with lymphoplasmacytic infiltrate, mucus acini and tumour composed of spindle-shaped cells with cigar-shaped nuclei and a moderate cytoplasm arranged in long intersecting fascicles arranged around a large vascular space lined by endothelial lining with haemorrhage and congested thin-walled blood vessels (figure 3). Mitoses were visualised very rarely (1/20 high-power field) and there was absence of necrosis and other secondary changes. Immunohistochemistry showed positivity for smooth muscle actin (figure 4) and S-100, confirming the diagnosis of a recurrent vascular leiomyoma. Margins sent separately were free of tumour.

OUTCOME AND FOLLOW-UP

The patient was followed up at 2 weeks and 1 month postoperatively and there was no evidence of residual or recurrent disease.

DISCUSSION

Leiomyomas constitute approximately 1% of all benign tumours in the human body.^{2 3} The most common sites of occurrence are the uterus which accounts for 95% of all leiomyomas. Less common sites are the skin (3%) and gastrointestinal tract (1%). Only 0.5% of leiomyomas occur at other sites which include the head and neck region.^{1 3 4} Hachisuga⁵ reported 48 cases of leiomyoma in the head and neck which included five cases in the nose and paranasal sinuses. Wertheimer-Hatch *et al*⁶ in their study reported the occurrence of 139 leiomyomas in the oral cavity and pharynx. Leiomyomas in the nose and paranasal sinuses are extremely rare and their presentation can mimic any other benign lesion in the nasal cavity. The most common sites in the nose are the inferior turbinate, nasal septum and nasal vestibule. Our patient presented with a recurrent angioleiomyoma from the middle turbinate. The female to male ratio is 3.75:1 and it commonly occurs in the age group of 50–60 years.^{7 8}

Figure 3 H & E staining (x5 magnification) showing the spindle cells background (hollow arrow) and vessels (black arrow).

Figure 4 Immunohistochemical staining (x5 magnification) showing spindle-shaped smooth muscle cells (hollow arrow) and small vessel walls denoting the smooth muscle component (black arrow) showing positive staining for smooth muscle actin.

The etiopathogenesis of leiomyoma in the nose and paranasal sinus is unknown. The role of Epstein-Barr virus has been implicated in immunocompromised individuals but further studies are required to confirm the role of EBV in the development of leiomyoma in the nose and paranasal sinus. Pre-existing trauma, steroid therapy and hormonal imbalance are other predisposing factors which have a role in the development of leiomyoma, but further clinical studies on a larger cohort need to be carried out to deduce their role in the development of leiomyoma in the nose and paranasal sinus.⁹

Three hypotheses have been suggested for the development of leiomyoma in the nose and paranasal sinuses. They are: (1) Presence of undifferentiated mesenchymal aberrations (2) Presence of smooth muscle in turbinal blood vessels (3) Presence of smooth muscle in vestibular erector pili and sweat glands.^{3 4 7}

Leiomyoma has been classified into two variants—vascular and non-vascular. The vascular variant, which is less common, may progress to develop solid tumours due to a gradual reduction in its vascularity. The WHO has classified myogenic tumours into leiomyoma, epithelioid leiomyoma (leiomyoblastoma) and angiomyoma (vascular leiomyoma).^{2 7} Morimoto utilised this classification and proposed three subtypes—capillary or solid, venous and cavernous based on the compactness of smooth muscle bundles and vascular channels. The presence of a spindle-shaped arrangement of cells in a fascicular pattern and exhibition of cells with blunt cigar-shaped nuclei without atypia and mitotic activity differentiates it histopathologically from other benign and malignant tumours.⁷

Leiomyomas are slow growing and they mimic any benign lesion in the nose and paranasal sinuses. The presentation of the tumour varies. The most common presentations are epistaxis and nasal obstruction. Headache and facial pain are less common symptoms which may occur due to the presence of the blockage of sinus ostia or due to the pressure on the bone and nerves. Our patient presented with facial pain, epistaxis and hyposmia and features of acute maxillary sinusitis due to maxillary ostial obstruction. Crusting and desiccation may occur due to the disturbance of laminar airflow in the nasal cavity, thereby manifesting as epistaxis. Bone erosion in leiomyoma is due to tumour pressure decalcification or due to the local extension of the tumour into the neighbouring tissues.^{2 4 7}

CECT and MRI are imaging modalities which have a minimal role in the diagnosis as leiomyomas do not show classical radiological signs. However, imaging does play a role in the surgical management and planning of the operative approach for

removal of the tumour. Immunohistochemistry on the resected tumour is the gold standard in diagnosis. The presence of α -smooth muscle actin (muscle-specific actin), desmin, S-100, myoglobin and vimentin confirms the diagnosis of leiomyoma.^{2-4 7 9 10} The presence of a positive progesterone receptor has been reported in 20–30% of leiomyoma in the world literature and this hypothesises the role of female predominance in the occurrence of leiomyoma; however, its role was not studied in our patient.^{8 9 11}

Complete surgical excision is the treatment of choice for leiomyoma of the nose and paranasal sinuses. Preoperative embolisation may be indicated in large tumours with locoregional extension; however, it is not required in small tumours limited to the nasal cavity and paranasal sinuses. The approach is largely dependent on the extension of the tumour ranging from an endoscopic transnasal approach for small tumours to craniofacial resection for tumours extending to the skull base. The role of the Potassium Titanyl Phosphate (KTP) laser for bloodless excision of a tumour has been reported in the world literature.^{2-4 7 8 12}

Recurrence is rare in leiomyoma of the nose and paranasal sinuses (less than 1%).¹³ The probable cause for the recurrence in our case was the incomplete primary excision of the tumour

and the involved surrounding bone. Malignant transformation of the leiomyoma has not been reported in the literature; however, Perzin has reported the possibility of a pre-existing benign disease in the development of leiomyosarcoma in the nose and paranasal sinuses.^{4 12}

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Contributors AMB was the principal investigator and prepared the manuscript. BR was a reviewer and coinvestigator. Dr Manna Valiathan reviewed the histopathology. NC collected the images and case data.

Competing interests None declared.

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Learning points

- Leiomyoma constitutes 1% of all benign tumours in the body and less than 0.5% of benign tumours in the head and neck with rare occurrence in the nose and paranasal sinuses.
- Immunohistochemical positivity for α 1 muscle actin is diagnostic of leiomyoma.
- Recurrence of tumour occurs on incomplete surgical excision and malignant transformation of leiomyoma is not documented in the literature.

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