CASE REPORT

Smooth muscle tumour of uncertain malignant potential (SMTUMP) in the nasal cavity: an incidental finding

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SUMMARY

Sino-nasal smooth muscle tumours of uncertain malignant potential (SMTUMP) are very rare neoplasms of mesenchymal origin with features in between a benign leiomyoma and a leiomyosarcoma. We report a rare case of SMTUMP in a 44-year-old woman, who presented with vague symptoms of pharyngitis. Nasal endoscopy revealed a smooth mass in left nasal cavity. Contrast-enhanced CT and MRI scans showed features likely to be inverted papilloma or olfactory neuroblastoma or meningioma. Excision was planned and intraoperatively, frozen section revealed a probable spindle cell lesion. Final histopathological report following immunohistochemistry (IHC) & immunofluorescence (IF) confirmed it to be a SMTUMP. This patient underwent complete resection via endoscopic KTP laser assisted, anterior skull base surgery with no recurrence on follow-up.

BACKGROUND

Smooth muscle tumours are usually seen in adult life, and the majority are genitourinary, gastrointestinal or cutaneous in origin. Even among the non-epithelial neoplasms of nasopharynx and sino-nasal tract, very few cases of leiomyoma and leiomyosarcoma have been reported.¹ These tumours are commonly seen in fourth and fifth decades with slight predilection for woman.² The clinical behaviour of smooth muscle cell tumours is dependent on site and degree of differentiation.^{2,3} In view of lack of clear knowledge on clinical course, challenging histopathological diagnosis and rarity of SMTUMP, this case is being reported.

CASE PRESENTATION

A 44-year-old woman with no comorbidities, presented to the Department of Medicine for a routine health check. On analysing the history, she gave symptoms of occasional throat irritation and postnasal discharge for the past 2 years. She was referred to the department of ENT—head and neck surgery, suspecting a nasal and paranasal sinus focus of infection. A detailed history was taken, and she reported of occasional episodes of throat irritation, lasting for about a week, usually during winter and gets relieved with or without medication. She also gives history of post-nasal discharge and hawking sensation in the throat since 2 years; occasionally associated with cough with yellowish expectoration that was relieved on antitussive medication. She did not give any history of nose block.

No history of sneezing, rhinorrhoea, epistaxis, altered smell perception, headache and fever was noted. There was no history of gastric reflux, change in voice or speech. The patient has never approached a physician for her throat symptoms nor had taken any treatment for the same. She took mixed diet and had normal sleep and appetite. She did not have any significant family history. She was evaluated to rule out infection in nose and paranasal sinuses. External nasal framework looked normal. Anterior rhinoscopy showed a pinkish globular mass in the superior part of left nasal cavity. Probing was not performed. The findings of oral cavity examination and posterior pharyngeal wall were normal.

INVESTIGATIONS

Diagnostic nasal endoscopy revealed a 2×2 cm pink smooth-surfaced globular mass with prominent blood vessels, medial to the left middle turbinate appearing to arise from skull base obscuring the anterior end of left middle turbinate (figure 1). Minimal mucoid discharge was present in the middle meatus, which was suctioned out. In view of possible bleed, no attempt was made to probe or to take a biopsy. A contrast-enhanced CT scan revealed a well-defined round-to-oval isohyperdense lesion with heterogeneous enhancement (2.7×1.7×2.6 cm) along the roof of left nasal cavity, remodelling the adjacent bony structures (figure 2). Superiorly, the lesion was eroding left cribriform plate, base of anterior cranial fossa with minimal intracranial extension, and also the left frontal recess with hypoplastic left frontal

Figure 1 Diagnostic nasal endoscopy showing mass in superior part of left nasal cavity.

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Figure 2 Coronal section of contrast-enhanced CT—paranasal sinus (PNS) showing the lesion.

sinus. Inferiorly, the lesion was eroding the left osteomeatal complex and anterior parts of left superior and middle turbinates. Laterally, it is infiltrating the left anterior ethmoid cells with thinning, bowing and focal erosion of left lamina papyracea, with minimal extension into extraconal compartment of the left orbit. Fat plane with medial rectus was maintained. Medially, mild bowing of nasal septum towards right was noted with focal erosion and abutting the middle turbinate. Minimal mucosal thickening was noted in left maxillary sinus. MRI revealed a well-defined round-to-oval isointense on T1WI, iso to hyperintense on T2WI and FLAIR imaging, showing homogenous postcontrast involving left anterior ethmoidal cells and left nasal cavity, suggesting a neoplastic lesion ([figure 3](#)).

DIFFERENTIAL DIAGNOSIS

- ▶ Haemangioma/angioma
- ▶ Olfactory neuroblastoma
- ▶ Meningioma
- ▶ Inverted papilloma
- ▶ Schwannoma and malignant lymphoma.⁴

TREATMENT

The patient underwent KTP 532 laser-assisted endoscopic excision. Intraoperatively, frozen section revealed a possible spindle cell lesion. Debulking of tumour was performed using a microdebrider to facilitate space for endoscopic dissection ([figure 4](#)). Mucosal incision was made with a 5 mm margin using KTP 532 laser, and mucosa was elevated from the bone up to the attachment of tumour ([figure 5](#)). Periosteal blood vessels were coagulated using laser. Middle turbinate was resected, and the tumour was removed from anterior ethmoid, along with normal mucosa. After separation of mass, bony area was drilled with a diamond burr over the anterior end of cribriform plate, and normal dura was exposed. The adjacent mucosa and remnant tumour mass was finally separated from dura. There was no involvement of dura. There was a breach of lamina papyracea,

Figure 3 Iso to hyperintense on T2 weighted MRI and fluid-attenuated inversion recovery (FLAIR) imaging.

Figure 4 Debulking with microdebrider.

Figure 5 Mucosal incision using KTP 532 laser.

but orbital periosteum appeared normal. All projecting bony spicules from anterior ethmoid, fovea ethmoidalis, up to the anterior wall of sphenoid sinus were drilled out. Left maxillary sinusotomy was performed, and sphenoid ostium was widened. The resected specimen along with left middle turbinate, superior turbinate attachment was marked and sent for histopathological examination. Margins were sent separately, including from left frontal recess, to look for adequate clearance. A posterior septal flap was elevated, rotated and covered over the exposed olfactory nerve endings and dura (figure 6).

OUTCOME AND FOLLOW-UP

Histopathological examination revealed nasal epithelium overlying a non-encapsulated tumour (figure 7), comprising fascicles of spindle-shaped cells with slender vesicular nuclei and scanty to moderate cytoplasm noted (figure 8). Tumour had <4 mitotic figures/10 high power field (HPF). All margins were clear except that the margin near to frontal recess was found to have a close margin. Therefore, a second-look endoscopy was performed, and the mucosa near frontal recess area was resected with KTP 532 laser. Repeat biopsy was free of tumour. Immunohistochemistry showed positivity of tumour cells for smooth muscle antigen (SMA) (figure 9) and was negative for cytokeratin (CK), Desmin, S-100 and CD34 (figure 10). Ki-67 proliferative index was <5% (figure 11). The patient was on

Figure 6 Septal flap to cover dura and olfactory nerve endings.

Figure 7 Nasal epithelium overlying a non-encapsulated tumour haematoxylin & eosin (H&E) (×20).

Figure 8 Fascicles of spindle-shaped cells with slender vesicular nuclei and scanty to moderate cytoplasm H&E (×100).

Figure 9 Immunohistochemistry showing that the tissue was positive for SMA (×100). SMA, smooth muscle antigen.

Figure 10 Immunohistochemistry showing the tissue was negative for CK (×100). CK, cytokeratin.

regular follow-up and has no evidence of local recurrence or any metastasis 18 months following the surgery.

DISCUSSION

The tumours of the upper aerodigestive tract are mainly epithelial tumours. Smooth muscle tumours arising in the upper

Figure 11 Ki-67 proliferative index was <5% (×100).

aerodigestive tract are rare and are extremely rare in nose and paranasal sinuses.^{1 3 5 6} They arise from the smooth muscle in the walls of blood vessels.^{7–9} Lesions, when occur here, are more commonly seen in the nose when compared to paranasal sinuses.¹ Fu *et al* in their study on 256 cases of sino-nasal and nasopharyngeal non-epithelial tumours found that only eight cases were smooth muscle tumours.⁷ Gross appearance of sino-nasal smooth muscle tumours is usually pale, polypoidal or nodular, pale pink to grey and soft to rubbery mass in the nasal cavity.² They present as asymptomatic, painless, slow-growing lesions. Patients usually complain of epistaxis, nasal obstruction and facial pain.^{5 6} Unlike this case, epistaxis or nasal obstruction was the most common presenting feature in the previously published case reports. There is a 1:2 male-to-female ratio in incidence, and the mean age for presentation was 40 years.^{2 10} They are derived due to aberration in the differentiation of mesenchymal tissue, vascular smooth muscle and myoepithelial cells in submucosal glands.^{11–13} Although they can develop anywhere in nasal cavity and paranasal sinuses, their development was found to be more common posteriorly than in anterior nasal cavity.³ Microscopically, smooth muscle tumours of the nasal cavity are usually subepithelial and may have mucosa ulcerations.¹ The tumour cells are found embedded in the musculature of tunica media of blood vessels. Histologically, smooth muscle tumours may range from a leiomyosarcoma to SMTUMP to leiomyoma. Leiomyomas have low cellularity and absence of nuclear pleomorphism with no mitotic figures.² SMTUMP show low cellularity, mild-to-moderate pleomorphism and 1–4 mitotic figures per 10 HPF. Leiomyosarcoma shows high-grade pleomorphism and more than four mitotic figures per 10 HPF.² Leiomyomas are mainly seen in uterine myometrium of uterus (95%). It may be seen involving skin (3%) and gastrointestinal tract (1.5%), and fewer than 1% of cases are seen in the head and neck region.^{11 13} The behaviour of the smooth muscle tumour depends on its site and also on the degree of cellular differentiation.³ A pathological examination SMTUMPs show mitotic figures ranging from 1 to 4/10 HPF, low MIB-1 index. The factors affecting the prognosis of a sino-nasal smooth muscle tumours are yet to be determined due to its rare incidence. Kuruvilla³ have indicated that the size and extent of involvement at the time of presentation has prognostic significance, but Huang and Antonescu² found that few cases with extensive involvement at presentation have had a favourable outcome without recurrence. Smooth muscle tumours exhibit no characteristic CT or MRI findings, but these

radiologic investigations are helpful in analysing extent of tumour, to look for any invasion or erosion, and in planning the treatment. Complete surgical resection is found to be the favourable treatment for these tumours.² In the era of minimally invasive surgery, endoscopic resection of the tumour is preferred over open rhinotomy approaches whenever feasible. KTP 532 laser delivered by a handheld device using fibreoptic cable was useful in our case as it precisely excised and coagulated the pedicle of the mass attached to the nasal septum. KTP laser is a very efficient tool for coagulation as it is absorbed by haemoglobin. Hence the intraoperative bleeding was negligible, and no nasal pack was required in our patient. The patient is on regular follow-up and has no local recurrence or distant metastasis 18 months postsurgery.

Learning points

- ▶ Smooth muscle tumour of uncertain malignant potential (SMTUMP) is a rare tumour in the sino-nasal cavity.
- ▶ These tumours although present mostly with epistaxis can be relatively asymptomatic.
- ▶ Owing to unknown malignant potential, the patients need to be followed-up closely.
- ▶ SMTUMP can be considered as a differential diagnosis while looking at a smooth vascular mass in the nasal cavity in women above the age of 40 years.

Contributors DRN involved in diagnosis; is the operating surgeon; contemplated to report this case; contributed to intraoperative findings, steps of procedure performed and recording and editing of the surgery; obtained postoperative pictures and involved in drafting and proof reading of the paper. ADG involved in review of literature and contributed to drafting the paper, editing of pictures, documentation, formatting and proof reading of the paper. ANR involved in procuring the consent, assisted in the surgery and helped in drafting the operative findings and procuring the histopathology slides. SS obtained details of histopathology and immunohistochemistry. All authors were not only involved in the patient care but also have actively participated in drafting of the paper. All authors have personally procured the data regarding the case, have reviewed the literature and have given inputs for the case report.

Competing interests None declared.

Patient consent Obtained.

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