CASE REPORT

'Saddle nose' deformity caused by advanced squamous cell carcinoma of the nasal septum

Hamish Edward Graham,¹ Cara Connolly,¹ Gurjinderpal S Pahal,¹ Francis W Stafford²¹St Andrews Department of Plastic, Burns & Reconstructive surgery, Broomfield Hospital, Chelmsford, Essex, UK²Department of ENT Surgery, MEHT, Broomfield Hospital, Chelmsford, Essex, UK

Correspondence to

Hamish Edward Graham,
hamishgraham@doctors.org.uk

Accepted 7 July 2014

SUMMARY

Hidden behind their common garden symptoms, malignant tumours of the nasal cavity are typically advanced when detected. Squamous cell carcinoma (SCC) is the most common histological type of these rare tumours which may simply present with epistaxis or a blocked nose. A 79-year-old woman attended our clinic with a 2-year history of progressive nasal collapse resulting in a 'saddle nose deformity'. Imaging showed a soft tissue mass with absolute destruction of the nasal septum and bones with hard palate and maxillary involvement. Histology confirmed a poorly differentiated SCC. Following discussion at the local head and neck multidisciplinary team meeting, the patient was offered radiotherapy alone due to her advanced presentation. She has had a good response to treatment with no further disease progression. Nasal septal SCC is far more successfully treated in the early stages. A suspicious clinician is the patients best ally.

BACKGROUND

Clinicians may only detect squamous cell carcinoma (SCC) of the nasal septum in its early stages, if they are highly suspicious of nasal symptoms which do not resolve. This affords the patient the best chance of local disease control and survival.¹⁻³

In addition to considering classical or text book aetiologies, the clinician should use the surgical sieve—a classical medical thought process for considering all causes of an usual presentation.⁴

Malignancy, although a rare cause of 'saddle-nose' deformity (SND) should be considered alongside congenital, traumatic, iatrogenic, inflammatory, infective, vasculitic, autoimmune, metabolic, drug-related and degenerative causes; to embrace the surgical sieve.

Congenital: Congenital causes of SND occur classically in congenital syphilis but are also associated with rare syndromes such as Rowat-Thompson, Mowatt-Wilson and Marshall syndrome.⁵⁻⁷

Traumatic: Traumatic causes of SND include fractures to the nasal bones and fracture with collapse of the nasal septum.⁸

Iatrogenic: Most cited examples of iatrogenic causes of SND are the complications of nasal refinement (rhinoplasty).⁹ Although it had been considered as a cause of septal pressure necrosis in this case, there are in fact no documented cases of SND caused by nasal cannulation.

Infective: Most cases of infective SND in the literature relate to leprosy, paediatric septal abscess and rarely cases of septal abscess in HIV.¹⁰⁻¹²

Again, although considered as a possible cause in the community which prompted the initial referral to our unit; there are no specific cited cases of osteomyelitis causing SND in the medical literature.

Vasculitic: Vasculitic causes of SND include Wegner's granulomatosis and the rarely reported PR3 Anca pulmonary vasculitis.^{13 14} Relapsing polychondritis is the only reported autoimmune cause of SND that we have identified.¹⁵

Drug related: Cocaine misuse as a drug-related cause of nasal septal perforation is common knowledge as a result of its prevalence in the popular press. Cases of full-blown SND resulting from cocaine misuse are also reported in the medical literature.¹⁶

Metabolic: We only identified one case of SND caused by a metabolic condition in the literature: a child with fucosidosis, which is a lysosomal storage disorder causing structural abnormalities which resulted in SND.¹⁷

CASE PRESENTATION

A 79-year-old woman presented to outpatients with a 2-year history of progressive nasal collapse and ulceration to the right nostril and columella.

Coincidentally, the ulceration to her nostril had first become apparent around the time of an admission to a local intensive care unit (ICU) for sepsis, following lithotripsy, 2 years previously. Initially, the ulceration was attributed to the nasal cannulation which she had undergone during her ICU admission thus the delay in her referral. There was no other history of trauma or surgery to her nose or mid-face.

On systems review she had no epistaxis or visual disturbance but latterly she had noticed a clear fluid discharge from her nose which had resolved at the time of presentation.

In her history she had chronic kidney disease, myocardial infarction, hypertension, primary hyperparathyroidism (untreated), recurrent nephrolithiasis and an ICU admission for urinary sepsis following previous lithotripsy.

She was an ex-smoker with a 30-pack-year history and lived independently with her husband in the community.

On examination she had complete collapse of the nasal septum with a SND and nasal deviation to the left. The right nostril sill and nasal bridge were ulcerated (figures 1 and 2). Oral cavity examination was unremarkable and she was clinically cervical node negative. The only other finding on

CrossMark

To cite: Graham HE, Connolly C, Pahal GS, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201765

Figure 1 Photograph at presentation from submental position. Ulceration to the nasal bridge and columella is demonstrated.

clinical examination was malodour from the nose and mouth which were anaerobic in character.

INVESTIGATIONS

Before referral to a specialist outpatient clinic, a plain facial X-ray had been arranged, which showed loss of mid-facial bone density and a suspicion of osteomyelitis was raised. This had prompted her initial referral.

A non-contrast MRI showed a low signal soft tissue mass at the base of the nose and a 10 mm right submandibular lymph node. A non-contrast CT showed a 3.2×2.0×3.0 cm destructive

Figure 2 Photograph from left side at presentation showing loss of nasal projection.

soft tissue mass, with hard palate destruction not detected on clinical examination. The anterior and medial walls of the maxillary sinus along with both nasal bones had been extensively eroded by the lesion. The nasal septum was completely obliterated. The overlying soft tissue involvement at the nasal bridge was also confirmed. The left maxillary and ethmoid sinuses were completely opacified (figures 3 and 4). Inferior to the right lobe of the thyroid gland, a 1 cm parathyroid adenoma was identified, coincidentally.

Following examination under anaesthesia, biopsies were taken of the mass from the nasal bridge, nasal vestibule, nasal floor, septum and roof of the nasal cavity, via the nares. This showed poorly differentiated SCC. The biopsy of the nasal vestibule showed moderate differentiation with overlying dysplasia.

DIFFERENTIAL DIAGNOSIS

Nasal trauma from prolonged cannulation was considered but there are no case reports present in the literature, osteomyelitis was also considered but there are no reports citing osteomyelitis specifically as a cause of SND, congenital syphilis, although unlikely, was also considered as was relapsing polychondritis and Wegener's granulomatosis. Appropriate microbiological, histological and serum samples were collected and sent for investigation.

TREATMENT

The local head and neck MDT reviewed the patient's history, imaging and the results of histological analysis and staged the tumour as an American Joint Committee on Cancer (AJCC) stage IVA (T4a,N0, M0) SCC (tables 1–3). Given the advanced nature of the disease (with extensive destruction of the mid-face), the frailty of the patient and potential significant

Figure 3 Sagittal CT slice showing destruction of the right maxillary wall, nasal septum and nasal bones.

Figure 4 Transverse slice CT showing nasopharyngeal mass, neo-osteogenesis with soft tissue density in the maxillary sinuses and destruction of the right maxillary wall and nasal septum causing nasal collapse to the left.

morbidity associated with surgical resection, a decision was made to offer radiotherapy alone.

OUTCOME AND FOLLOW-UP

At 3 months post radiotherapy, the patient showed a good response to treatment with significant regression of the tumour. She has been left with a defect in the hard palate and further loss of nasal projection. The palatal defect will be managed with an obturator to facilitate speech, eating and drinking. Should problems arise with wearing spectacles, all possible prosthetic solutions will be explored. She remains under close outpatients review and has no other symptoms following radiotherapy.

DISCUSSION

The total incidence of all nasal and paranasal cavity cancers is cited as being approximately 1 in 100 000 of the population

Table 1 Primary tumour location within nasal cavity or ethmoid sinus¹⁹

T1	Tumour restricted to any one subsite, with or without bony invasion
T2	Tumour invading two subsites in a single region or extending to involve an adjacent region within the nasoethmoidal complex, with or without bony invasion
T3	Tumour extends to invade the medial wall or floor of the orbit, maxillary sinus, palate or cribriform plate
T4a	Moderately advanced local disease Tumour invades any of the following: anterior orbital contents, skin of nose or cheek, minimal extension to anterior cranial fossa, pterygoid plates or sphenoid or frontal sinuses
T4b	Very advanced local disease Tumour invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than (V ₂), nasopharynx or clivus

Reproduced from AJCC Manual in Cancer staging 7th edn.

Table 2 Classification of regional lymph nodes and metastasis¹⁹

NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in a single ipsilateral lymph node, ≤3 cm in greatest dimension
N2	Metastasis in a single ipsilateral lymph node, >3 cm but ≤6 cm in greatest dimension, or metastases in multiple ipsilateral lymph nodes, ≤6 cm in greatest dimension, or in bilateral or contralateral lymph nodes, ≤6 cm in greatest dimension
N2a	Metastasis in a single ipsilateral lymph node, >3 cm but ≤6 cm in greatest dimension
N2b	Metastases in multiple ipsilateral lymph nodes, ≤6 cm in greatest dimension
N2c	Metastases in bilateral or contralateral lymph nodes, ≤6 cm in greatest dimension
N3	Metastasis in a lymph node, >6 cm in greatest dimension
M0	No distant metastasis
M1	Distant metastasis

with the incidence of nasal septal carcinoma occurring as fraction of this.¹⁸ Up until the past decade, all malignancies of the nasal cavity were classified and reported in conjunction with paranasal sinus cancers because they were anatomical contiguous cavities and possibly because of their low incidence.

Staging

Since 2001, the AJCC has introduced formal staging criteria for nasal cavity malignancies. They are now classified separately from paranasal cancers although nasal vestibule tumours have been added to this group. Nasal cavity tumours are further sub-classified into their site of origin, to help guide treatment and prognosis.¹⁹ The latest version of the AJCC staging is reproduced in tables 1–3 with permission of the AJCC and Springer Publishing.

Treatment: radiotherapy, surgery or both?

Early septal carcinomas, for example, small AJCC T1 tumours can be treated successfully with radiotherapy or surgery. Ho and Coman² reviewed two patients with T1 septal SCC followed up for between 2 and 3 years, all of whom were disease free after surgical resection. Allen *et al*²⁰ demonstrated at least one patient

Table 3 Anatomic stage/prognostic groups¹⁹

Stage	T	N	M
0	Tis	N0	M0
I	T1	N0	M0
II	T2	N0	M0
III	T3	N0	M0
	T1	N1	M0
	T2	N1	M0
	T3	N1	M0
IVA	T4a	N0	M0
	T4a	N1	M0
	T1	N2	M0
	T2	N2	M0
	T3	N2	M0
	T4a	N2	M0
IVB	T4b	Any N	M0
	Any T	N3	M0
IVC	Any T	Any N	M1

Reproduced from AJCC Manual in Cancer staging 7th edn.

treated with radiotherapy alone with T1 septal SCC, who was followed up for 29 months and remained disease free. DiLeo *et al*¹ have also drawn from their metanalysis of nasal septal SCC, that either radiotherapy or surgery are effective for small tumours. For larger AJCC T1 tumours and AJCC T2, T3 and T4, evidence has shown that surgery or surgery combined with radiotherapy offers superior cure rates for SCC of nasal and paranasal carcinoma, reported together.¹⁸ More recent data from large volume units indicate that managing patients with combined surgical resection and radiotherapy for larger nasal cavity carcinoma, is their modus operandi.²⁰

Treatment—elective lymph node treatment

Node positive disease at presentation is usually below 10%³ and regional recurrence in cervical lymph nodes is also low.²⁰ However, there is some emerging evidence that risk of regional lymph node recurrence might be higher in nasal septal SCC and at a level (up to 20%) where cervical lymph node treatment would be justified in this group. As numbers remain small from which to draw conclusions however, only T3 and T4 tumours have elective cervical lymph node treatment.²⁰

Treatment—chemotherapy

Although not part of routine nasal cavity cancer protocols, Allen *et al*²⁰ support the use of platinum-based chemotherapy in specific individual cases, that is, cases of non-resectable cancer.

Outcomes

In the past 20 years the qualification of survival from cancer has evolved to the current terminology of survival, disease free survival, locoregional control and disease failure.²⁰

During this time outcomes have been combined together which unfortunately confound the interpretation of outcomes in the different stages of carcinoma of the nasal cavity, particularly for the specific subgroup of SCC of the nasal septum. Depending on the series cited, 5-year survival for nasal cavity SCC ranges from 86% to 61%.^{3 20} The mean recurrence rate of SCC of the nasal cavity is cited at 18%.²¹

AJCC stage IVa tumour

Our patient with a previously unreported case of SCC-related SND had an AJCC IVa tumour (T4 N0 MO). This is classed as an advanced cancer which is surgically resectable. However, surgical resection of the mid-face may involve rhinectomy, maxillectomy or orbital exenteration to achieve surgical clearance. When facing advanced disease at first presentation the MDT team should consider that such surgery is likely to cause functional, psychological and social morbidity to the patient. Altered routes of nutrition would need to be considered as part of the surgical plan as would potential visual and speech rehabilitation.

It is paramount to balance the opportunity to achieve surgical clearance against the individual patients wishes, general health, reconstructive potential, prosthetic potential and life expectancy. It may be in the patients best interest to offer palliative radiotherapy in advanced disease for symptomatic relief.

The advances in nasal cavity cancer treatment in the past 40 years of last century are heartening and modern protocols in high volume units can achieve 5-year disease-specific survival of over 80%.²⁰ Despite this, the actions of the grass roots clinician can have a greater impact on patient well-being, by detecting early disease.

Learning points

- ▶ Although rare, clinicians should consider malignancy of the nasal cavity as a cause of everyday persistent nasal symptoms. Early detection and specialist referral proffers the patient the best chance of a favourable outcome.
- ▶ Squamous cell carcinoma is the most common histological subtype of malignancy in the nasal cavity. It advances along the architecture of the mid-face to the palate, sinuses, orbit and along the skull base.
- ▶ The complex configuration of intranasal malignancy may require CT and MRI to assess the extent of bony involvement and to differentiate between tumour and inflammatory changes, respectively.²²
- ▶ Malignancy should be considered as a cause of progressive mid-face deformity.
- ▶ The surgical sieve remains a useful tool for qualified doctors and students alike in considering all possible diagnosis for a confounding presentation.⁴
- ▶ Treatment of nasal cavity and therefore nasal septal malignancy has improved in the past 50 years. Some of this improvement in outcome can be attributed to the uniform classification system provided by the AJCC.^{19 20} Today early disease can be treated equally well with surgery or radiotherapy, thereafter (larger T1 tumours and beyond) are best treated with a combination of surgery and radiotherapy.^{2 20} Although theoretically 'resectable' a common sense approach should be adopted to resection of advanced T4 tumours, with special consideration for individual patients, potential morbidity and expected survival time.

Acknowledgements The authors would like to acknowledge the significant contribution of Mr Bhagwat Mathur Consultant Plastic and Reconstructive surgeon who reviewed a number of draft manuscripts and was initially referred the patient and contributed to the initial patient care and assessment.

Contributors HEG and CC contributed to initial patient diagnosis and care, conducted literature reviews and prepared the manuscript. GSP contributed to initial patient care and provided imaging FWS is the senior clinician providing ongoing care and who has reviewed and revised previous drafts of this manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 DiLeo MD, Miller RH, Rice JC, *et al*. Nasal septal squamous cell carcinoma: a chart review and meta-analysis. *Laryngoscope* 1996;106:1218–22.
- 2 Ho YM, Coman WB. Nasal septum malignancy. *J Surg* 2011;81:533–6.
- 3 Bhattacharyya N. Cancer of the nasal cavity: survival and factors influencing prognosis. *Arch Otolaryngol Head Neck Surg* 2002;128:1079–83.
- 4 Hartley E, Trafford G, Bhangu S, *et al*. How to answer questions. *Student BMJ* 2009;17:b2173 (accessed 13 Mar 2014).
- 5 Larizza L, Roversi G, Volpi L. Rothmund-Thomson syndrome. *Orphanet J Rare Dis* 2010;5:2.
- 6 Garavelli L, Mainardi PC. Mowat-Wilson syndrome. *Orphanet J Rare Dis* 2007;2:42.
- 7 Onile BA, Rotowa A, Osoba AO, *et al*. Marshall syndrome: a condition resembling congenital syphilis. *Br J Vener Dis* 1981;57:100–2.
- 8 Won TB, Kang JG, Jin HR. Management of post-traumatic combined deviated and saddle nose deformity. *Acta Otolaryngol* 2012;132(Suppl 1):S44–51.
- 9 Thomson C, Mendelsohn M. Reducing the incidence of revision rhinoplasty. *J Otolaryngol* 2007;36:130–4.
- 10 Menger DJ, Tabink IC, Trenité GJ. Nasal septal abscess in children: reconstruction with autologous cartilage grafts on polydioxanone plate. *Arch Otolaryngol Head Neck Surg* 2008;134:842–7.

- 11 Sandel HD IV, Davison SP. Three spontaneous occurrences of nasal septal abscess in patients with chronic asymptomatic HIV—the need for early intervention and reconstruction. *Ear Nose Throat J* 2009;88:1058–66.
- 12 Malaviya GN, Husain S. Surgical correction of saddle nose deformity in leprosy—one stage procedure. *Acta Leprol* 1994;9:76–82.
- 13 Sepehr A, Alexander AJ, Chauhan N, *et al*. Detailed analysis of graft techniques for nasal reconstruction following Wegener granulomatosis. *J Otolaryngol Head Neck Surg* 2011;40:473–80.
- 14 Zycinska K, Wardyn KA, Piotrowska E, *et al*. Rhinologic and sinonasal changes in PR3 ANCA pulmonary vasculitis. *Eur J Med Res* 2010;15(Suppl 2):241–3.
- 15 Cantarini L, Vitale A, Brizi MG, *et al*. Diagnosis and classification of relapsing polychondritis. *J Autoimmun* 2014;48–49:53–9.
- 16 Shipchandler TZ, Chung BJ, Alam DS. Saddle nose deformity reconstruction with a split calvarial bone L-shaped strut. *Arch Facial Plast Surg* 2008;10:305–11.
- 17 Soltani AE, Moharari RS, Ghaffari R, *et al*. Fucosidosis and anesthesia. *Saudi Med J* 2007;28:1446–8.
- 18 Dulguerov P, Jacobsen MS, Allal AS, *et al*. Nasal and paranasal sinus carcinoma: are we making progress? A series of 220 patients and a systematic review. *Cancer* 2001;92:3012–29.
- 19 Edge SB, Byrd DR, Compton CC, *et al*, eds. *AJCC cancer staging manual*. 7th edn. New York: Springer, 2010:69–78.
- 20 Allen WM, Schwartz DL, Rana V, *et al*. Long term radiotherapy outcomes for nasal cavity and septal cancers. *Int J Radiat Oncol Biol Phys* 2008;71:401–6.
- 21 Cooper Scurry W, Goldenberg D, Chee MY, *et al*. Regional recurrence of squamous cell carcinoma of the nasal cavity a systematic review and meta-analysis. *Arch Otolaryngol Head Neck Surg* 2007;133:796–800.
- 22 Zinreich SJ. Imaging of the nasal cavity and paranasal sinuses. *Curr Opin Radiol* 1992;4:112–16.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow