

Squamous cell carcinoma of the nasal vestibule: a 20-year case series and literature review

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Abstract

Squamous cell carcinoma of the nasal vestibule is a rare disease with significant morbidity and mortality, and a five-year recurrence-free survival rate of between 42 and 92 per cent. There are several staging systems: the American Joint Committee on Cancer (AJCC) skin and nasoethmoid complex system, the Union International Centre Cancer (UICC) nasal fossa system, and Wang's system. Treatment options include radiotherapy or surgery for early lesions, but more advanced cases require radical surgery with post-operative radiotherapy. We present a case series spanning the last 20 years in one centre, and we compare this series with cases reported in the literature, paying particular attention to staging, treatment and outcome. We found that patients with tumours staged T₂ or T₃ (Wang system) who received radiotherapy alone did poorly in comparison with those who received surgery or surgery and radiotherapy.

Key words: Squamous Cell Carcinoma; Nasal Cavity; Nasal Septum; Nasal Vestibule

Introduction

Squamous cell carcinoma (SCC) of the nasal vestibule is a rare disease with significant morbidity and mortality and a five-year recurrence-free survival rate of between 42 and 92 per cent.^{1,2} This tumour is frequently misdiagnosed initially, feigning vestibulitis, past local trauma or infection. A delay in diagnosis may necessitate more radical treatment, with reduced survival rates. There are several staging systems: the American Joint Committee on Cancer (AJCC) skin and nasoethmoid complex system, the Union International Centre Cancer (UICC) nasal fossa system and Wang's system.³ Cases reported in the literature are often staged retrospectively. Treatment options include radiotherapy or surgery for early lesions, but more advanced cases require radical surgery with post-operative radiotherapy.

We present a case series spanning the last 20 years in one centre, and we compare this series with cases reported in the literature, paying particular attention to staging, treatment and outcome.

Methods

A retrospective search for patients' records was carried out using available data from both the local head and neck multidisciplinary team database and the personal database of one of the authors (NSJ). The pathology record database (Standardised

Nomenclature of Medicine [SNOMED] coding) was searched, using the key words 'nose' and 'SCC', and the resulting list was reviewed. Only patients with SCC arising from the nasal vestibule and anterior septum were included. We excluded non-squamous cell carcinoma (e.g. basal cell carcinoma) of the vestibule or septum, and SCC from other sites (e.g. skin and paranasal sinuses). Patients were identified and their notes reviewed. We recorded patients' age at diagnosis, original lesion site and any other anatomical areas involved.

The size of the tumour at presentation was recorded from the clinic, theatre and pathology records, as was any clinical or radiological evidence of nodal or distal metastasis. Patients were retrospectively staged using both the Wang and UICC systems. The pathology and degree of differentiation of lesions reported in this series was subsequently checked by one of the authors (RA) by reviewing the original samples. We analysed patients' initial treatments, any subsequent treatments given as a consequence of residual or recurrent disease, and patients' survival rates.

Results

Fifty-two patients were identified in the search. Eighteen were found to have had either a primary SCC of the external skin, an SCC of the sinuses or related to

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a transitional cell carcinoma, or basal cell carcinoma, and were thus excluded. Unfortunately, six sets of notes had been destroyed and one lost, leaving 27 patients. The male to female ratio was 1.5:1. The mean age at diagnosis was 64 years five months (range 32–90 years). The most common sites were the septum (18 cases) and columella (four cases). The lateral alar region was involved in two cases and the nasal floor in two cases, whilst one case was not clearly documented and had not been staged. The staging and treatment results are summarised in Table I.

One patient had a well differentiated tumour and 16 had moderately differentiated tumours (of whom eight had recurrent or persistent disease, and five died of their disease). Six patients had moderately to poorly differentiated tumours, one of whom died of their disease. Three patients had poorly differentiated tumours, of whom one died of their disease. One patient's original histological analysis was performed at a different institution.

Thirteen patients were treated primarily with radiotherapy alone. Five of these patients went on to undergo salvage surgical removal of recurrent disease at the primary site, and, of these patients, three also underwent a neck dissection for cervical nodal disease. One further patient who was treated primarily with radiotherapy had a neck dissection for cervical lymphadenopathy alone. Two of these thirteen patients had no sign of recurrence (after six months' and 13 years 10 months' follow up). One patient had palliative care alone for an orbital recurrence. Another, who had no recurrence, had reconstructive surgery for radionecrosis.

Ten patients were treated with surgery as the primary modality. Of these, two went on to undergo further resection – one for recurrence at

the primary site (this patient remained disease-free for six years) and the other for cervical metastasis (this patient died five years 10 months after treatment from metastatic disease).

In this series, five patients had tumours staged as T₂ or T₃ (Wang system), treated by radiotherapy alone, and three died of their disease.

In comparison, the five patients who received surgery or surgery plus radiotherapy all remained disease-free.

The overall survival rate for vestibular carcinoma was 68 per cent with a mean follow up of eight years one month (range, six months to 13 years 10 months). Six patients died of their disease. Two patients died of an unrelated cause.

Discussion

Nasal vestibular SCC is rare. In the present series, in a catchment area of approximately 810 000 there were 25 cases, giving an estimated incidence of 1.99 per million population (the other cases were tertiary referrals from outside our area). Because of the nature of the treating author's (NSJ) practice, there were extra-regional referrals, many of which had received initial treatment elsewhere which had failed. Thus, it may be that this series describes a population with a skew to the worse end of the spectrum, affecting our recurrence and survival rates.

It is unfortunate that seven sets of notes had been destroyed or lost, as uncommon cancers such as nasal vestibular SCC need lengthy historical review in order to generate reasonable numbers to enable comment on management. All of these notes were more than 15 years old, and as such the earliest patient in this series was actually diagnosed in 1991. Current UK Department of Health policy states

TABLE I
COMPARISON OF TNM AND WANG SYSTEMS OF NASAL VESTIBULAR SCC STAGING

Stage	Treatment	n	Recurrence			Result		
			L	R	LR	DoD	Salvaged	NSR
<i>Wang</i>								
T ₁	DXT	6			2	2	0	4
	Surg	6	1	1		1	1	4
	DXT + Chem	1				0		1
T ₂	DXT	3		1	1	1	1 (early)	1
	Surg	2				0		2
T ₃	DXT	3	2		1	2	1	0
	Surg	2 (1 N+)				0		2
	Surg + DXT	2				0		2
	DXT + Chem	1 (N+)				1	0	0
<i>TNM</i>								
T ₁	DXT	8	1	1	2	3	1 (early)	4
	Surg	7	1	1		1	1	5
T ₂	DXT	2			1	0	1	1
	Surg	2 (1 N+)				0		2
	DXT + Chem	1				0		1
T ₄	DXT	2	2			2	0	0
	Surg	1				0		1
	Surg + DXT	2				0		2
	DXT + Chem	1 (N+)				1	0	0

N refers to nodal status. TNM = tumour–node–metastasis; SCC = squamous cell carcinoma; L = local; R = regional; LR = locoregional; early = patient with only 3 months' follow up after salvage treatment; DoD = died of disease; NSR = no sign of recurrence; DXT = radiotherapy; surg = surgery; chem = chemotherapy

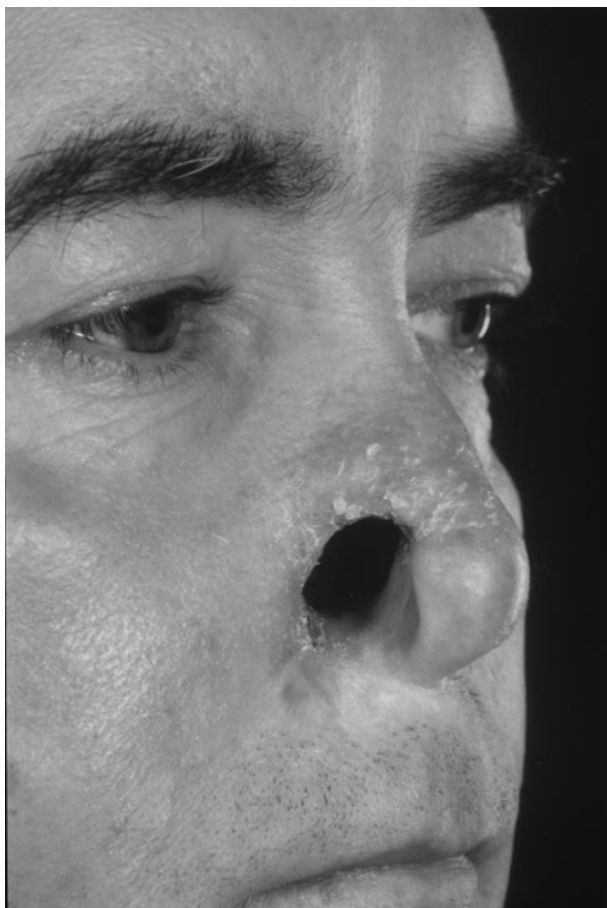


FIG. 1

Necrosis following radiotherapy for squamous cell carcinoma of the nasal vestibule.

that patient records should be retained for a minimum period of 30 years for oncology (furthermore, permanent preservation should be considered for research purposes), and for a period of 50 years or to age 75 years, whichever is the longer, for radiation dose records.⁴

As with all cancers, accurate note-taking at presentation and biopsy provides vital data for initial staging, thus aiding the choice of treatment options and providing valuable audit data for future review. In cases of vestibular carcinoma, it is essential to record both tumour size and a detailed description of the affected anatomical sites. The availability and use of clinical photographs or diagrams helps this process.

As there is no universally recognised staging system for nasal vestibular SCC, a number of previous authors have attempted to assess and compare both of the staging systems used, with varying results. Horsmans *et al.*⁵ and Langendijk *et al.*⁶ concluded that Wang's system did not show any prognostic value, whereas Vendelbo Johansen *et al.*⁷ found both the Wang and the tumour-node-metastasis (TNM) systems to be strong and useful predictors of overall and disease-free survival. Jeanon *et al.*⁸ found Wang's system to be better. Kummer *et al.*⁹ found both the Wang and the AJCC systems had significant prognostic value, but

they commented that the latter system allowed 5 cm lesions to be categorised as T₃, and that it was extremely unlikely that this stage would occur without deeper involvement, therefore making the lesion a T₄. We could only find one paper that reported any T₃ tumours according to the UICC classification.⁸ Poulsen and Turner¹⁰ and Mendenhall *et al.*¹¹ used the UICC TNM system and found that it did not predict recurrence-free survival.

The accepted treatment for early or stage one (Wang or TNM system) disease is radiotherapy, although local surgical excision also achieves good results. We found that those patients with involvement of the deep tissues of the septum and columella, particularly around the anterior nasal spine (i.e. T₂ or T₃ by Wang's system), had a worse prognosis. In all but one of our T₂ and T₃ (Wang system) patients treated solely with radiotherapy, the tumour recurred; in comparison, those who received surgery or surgery plus radiotherapy remained disease-free. This indicates more aggressive or advanced disease, and these findings have also been reported by others.^{3,11-15} Treatment of these patients by radical surgery with post-operative radiotherapy is more successful than radiotherapy alone.^{11,14,16} However, these individual series are not large enough to reach statistical significance. Jeanon *et al.*⁸ were able to show a significant difference between the five-year overall survival for superficial (72 per cent) and deep (40 per cent) tumours. Many series report their local control rate but do not clearly report the number of cases of post-radiotherapy recurrence requiring surgery. For T₂ lesions, this is approximately 35–60 per cent.^{9,16} Complete surgical resection and high quality reconstruction, followed by radiotherapy, would give patients optimum quality of life and a better survival rate.

Wang T₃ tumours are not suitable for radiotherapy alone,⁹ and our results confirm this. Patients without neck node involvement have a much better prognosis than those with such involvement.¹⁶

Table II summarises the staging and treatment modalities used in published series for which recurrence rates have been reported. This Table includes data from studies which reported staging and outcomes without specifying treatment;^{2,3,5-11,13,14,16-18,20} thus, the total numbers shown are not the sum of the figures in the preceding rows. Data from the present series are included in this Table. Unfortunately, the numbers collated are too small to generate statistical significance.

We were unable to correlate patient prognosis with degree of tumour differentiation. Pantelakos *et al.*¹⁵ found that the degree of tumour differentiation correlated directly with patient survival. Jeanon *et al.*⁸ found that tumour grade was associated with a worse disease-free survival period but not with overall survival. However, Poulsen *et al.*¹⁰ found that the degree of tumour differentiation was not a prognostic indicator, and Langendijk *et al.*⁶ found that patients with better differentiated tumours did worse than those with poor differentiation.

Despite the existence of evidence that more advanced tumours (T₂ and T₃, Wang system) are best treated with primary surgery and radiotherapy,¹⁶

TABLE II

STAGING AND TREATMENT MODALITIES USED IN NASAL VESTIBULAR SCC CASES FROM STUDIES REPORTING RECURRENCE RATES

Treatment	Wang (<i>n</i> (%))			TNM T ₄ (<i>n</i> (%))	N+ or recurrent disease (<i>n</i> (%))
	T ₁	T ₂	T ₃		
DXT	169 (12)	109 (30)	10 (70)	34 (35)	28 (36)
Surgery	6 (33)	7 (13)	6 (33)	1 (0)	5 (40)
Surgery + DXT			3 (0)	5 (20)	4 (25)
Other			1 S + DXT + Ch (0)		4 (2 DXT + Ch, 2 S + Ch) (75)
Total	300 (16)	176 (35)	44 (55)	48 (40)	44 (37)

Percentages indicate patients who had a recurrence or died of their disease. SCC = squamous cell carcinoma; TNM = tumour–node–metastasis; N+ = nodal metastasis; DXT = radiotherapy; S = surgery; Ch = chemotherapy

surgery is often omitted. Radiotherapy alone may be given because of the surgeon's concerns about being unable to reconstruct the surgical defect with a good cosmetic result.¹⁶ There is a perception amongst both patients and physicians that radical surgery with reconstruction (surgical or prosthetic) is disfiguring, and that a good cosmetic result is better achieved with radiotherapy. Indeed, this is often the case where the local surgical and prosthetic service does not have an interest or expertise in this area. This situation will lead to more patients being treated with radiotherapy, limiting the options available to the patient and, as discussed above, worsening the prognosis. Levendag *et al.*² assessed cosmesis after radiotherapy for T₁ and T₂ N₀ tumours, and found that 65 per cent of patients had a good or excellent cosmetic result. The morbidity associated with radiotherapy should also be taken into account in the decision-making process. Patients suffer from dryness, crusting and telangiectasia after radiotherapy. Significant radionecrosis, requiring surgical reconstruction or hyperbaric oxygen, has been reported in 14 patients.^{6,9,10,11,13,17} There has been one case reported of sarcoma developing *de novo*, seven years after radiotherapy for SCC of the nasal vestibule.⁶

With the introduction of Moh's micrographic surgery in our unit in 1997, in conjunction with the aesthetic results achieved from surgical reconstruction, we feel that T₂ and T₃ (Wang system) lesions are best managed by Moh's surgery, reconstruction and radiotherapy.

Conclusions

Squamous cell carcinoma of the nasal vestibule should be staged carefully, and those tumours involving the deeper tissues should be treated by primary surgical excision followed by radiotherapy.

Because of the rarity of this carcinoma, patient survival and morbidity will be improved by centralised, multidisciplinary team led treatment, incorporating Moh's micrographic surgery and a full range of surgical reconstructive options.

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