CASE REPORT

Clown nose: a case of disfiguring nodular squamous cell carcinoma of the face

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giacomo.colletti@gmail.com**SUMMARY**

'Clown nose' (CN) is the common medical term referring to a reddish-brown bulge involving the tip of the nose, reminding of a clown's fake red nose. Reports about these tumours are scarce. Most reports refer to metastatic skin manifestation of systemic malignancies: this condition has been rarely described as a primary skin neoplasm. We report a case of a 31-year-old patient with a giant cutaneous squamous cell carcinoma of the nose which evolved into a CN. After ruling out genetic or immune risk factors, the patient was treated with surgical excision of the lesion and local reconstruction with good aesthetic outcome and no recurrence over a 2-year follow-up.

BACKGROUND

Clown nose (CN) is a term that refers to the presence of a reddish-brown bulge involving the tip of the nose, resembling the red fake nose of a clown. Although many conditions may produce a brownish mass in the nasal tip, a rapidly growing CN suggests an underlying malignancy, in this case a well-differentiated squamous cell carcinoma (SCC).

The young age of the patient, which showed no malignancy risk factors such as immune deficiencies, smoking, substance abuse or excessive sun exposure, required collecting a complete family history in order to identify potential favouring genetic conditions.

Furthermore, the site of the lesion required a careful excision with impeccable presurgical planning, in order to grant at least an acceptable aesthetic result and good quality of life for the patient.

CASE PRESENTATION

A 31-year-old Caucasian man was referred from the dermatology department to our maxillofacial surgery department for a nodular growing mass on the tip of the nose, appeared 2 months earlier. The lesion had reached a diameter of 3.5 cm after a steady growth (3–5 mm/week) and was otherwise asymptomatic. It appeared reddish-brown in colour, dome-shaped with an incomplete central keratin-filled crater ([figure 1](#)).

The patient was otherwise healthy and had a blank medical history. He was working as an IT technician and he had no previous excessive exposure to sun or radiations. He had no family history of skin malignancies and the only malignancy he could remember was a non otherwise specified hepatic neoplasm in an alcoholic uncle.

INVESTIGATIONS

We performed an incisional biopsy which showed well-differentiated epithelial pearls infiltrating the dermis and did not allow us to differentiate between a keratoacanthoma and an SCC.

A new incisional biopsy confirmed the diagnosis of SCC, due to the aggressive infiltrative behaviour of the tumour cluster that reached the cartilage and to the presence of atypical mitoses and stromal desmoplasia ([figure 2](#)).

Before planning the surgical procedure, we performed routine laboratory investigations and serological tests, chest X-ray, head and neck CT and abdominal ultrasound examination in order to exclude potential distant malignancies or synchronous primary tumours. The investigations ruled out other possible malignancies and lymph node involvement.

After resection, the pathologist reported that the specimen ([figure 3](#)) included a carcinoma deeply extending below the level of sweat glands, infiltrating dermis, subcutaneous fat and surrounding cartilages reaching the nasal mucosa. The neoplasm showed an infiltrative growth pattern without deformation of the cutaneous annexes with variably differentiated areas extending until 1.35 mm from the resection margin. No perineural invasion was detected.

DIFFERENTIAL DIAGNOSIS

Whenever a shouldered lesion with a central keratin-filled crater presents, it is often difficult to differentiate between SCC and keratoacanthoma.

Figure 1 The clown nose is characterised in the patient by a dome-shaped reddish nodular lesion with an incomplete central keratin-filled crater.

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Figure 2 Histological examination. Although the neoplasm showing a tentative crateriform central area, a clear infiltrative growing pattern at the edges was evident ($\times 10$; H&E).

In this case the incomplete crater and, most importantly, the astonishing speed of growth seemed to favour an SCC. The final diagnosis can be made through histological examination, though.

Furthermore, nose tip lesions should always be in differential diagnosis with cutaneous metastasis.

Rapid growth and substantial mass effect help the differential diagnosis with inflammatory diseases and rhinophyma, which are caused by a hyperplasia of the sebaceous glands and easily recognised.

TREATMENT

Owing to the suspicion of a keratoacanthoma, the patient was proposed a tentative treatment with gentamicin ointment twice

Figure 3 Histological examination. This squamous cell carcinoma showing its typical pattern of infiltration below the cutaneous annexes, without deforming them. The carcinoma extends deeply below the level of the cutaneous annexes, infiltrating dermis and the subcutaneous fat ($\times 2$; H&E).

daily. After the final diagnosis of SCC was made, the patient was proposed a radical surgical procedure, with subsequent reconstruction.

The tumour excision was performed under general anaesthesia with a 10 mm macroscopic margin deepening to the lower and upper lateral cartilages and the nasal septum (figure 4). During this same surgical session, a forehead flap was sculpted and raised.

Ten days after the first procedure, when surgical margins were declared clear of the tumour, we started the reconstructive procedure, rotating the forehead flap to cover the defect and grafting the skin into the subsequent forehead defect.

After 4 weeks the flap was elevated with a costochondral graft providing skeletal support for the dorsum and the alae of the new nose, and after four more weeks the pedicle of the flap was detached and the flap inset.

A skin expander placed in the adjacent forehead granted defect closure with a local rotation flap with a good aesthetic outcome (figure 5).

OUTCOME AND FOLLOW-UP

We monitored the patient with clinical examinations every 2 months for the first year and then every 4 months. An ultrasound tomography of the neck and abdomen was added every 6 months. We decided to further investigate the lungs with a chest X-ray every year.

After 2 years of follow-up the patient is healthy and no signs of recurrences or other localisations were detected.

Even if the whole surgical treatment took almost 4 months to complete, the patient was deeply satisfied not only for the oncological results but also for the aesthetic appearance of the new nose, which granted him a good quality of life.

DISCUSSION

Disfiguring reddish-brown masses involving the tip of the nose are fortunately uncommon at best. If we take into account all rapidly growing nasal tip reddish-brown masses, only eight papers describe what may be defined as a CN.¹⁻⁸ In six of these papers the condition was due to a skin metastasis of a lung or breast cancer and most of these patients already had an existing diagnosis of breast or lung neoplasm. In only one paper the cause was a basal cell carcinoma and this is the only reported

Figure 4 The nose region after the tumour resection. We can observe the full thickness defect involving the upper and lower lateral cartilages and the anterior septum.

Figure 5 The nose region in its final shape after the reconstructive procedures.

literature case of a primary skin malignancy producing such disfigurement.¹ Our patient is the first case in which an SCC is involved. Chest X-rays and neck and abdominal ultrasound are therefore necessary, since most of the reports regard metastases from lung or breast cancer.

The surgical treatment consists in the removal of the lesion with a good safety margin. Some authors suggest a 1 cm safety margin while dealing with SCC of the skin.⁹ This wide 10 mm margin, which may sound questionable, was chosen since the lesion involved more than one aesthetic subunit of the nose: in these patients a complete demolition of the whole nasal aesthetic unit with subsequent reconstruction yields significantly better results. Luckily enough the large safety margin granted a tumour-free resection despite the neoplasm reaching up to 1.35 mm from the surgical resection in the final examination.

Mohs micrographic surgery could have been indicated in this case. Unfortunately the procedure was not yet available in our department at time of the lesion excision.

After removing the tumour, we decided to wait for the histopathological examination of the specimen to assess the resection margins. This cautious approach follows the rule of halting the reconstruction until confirmation of clear margins on histological examination.¹⁰ Moreover, total and subtotal nasal reconstructions are multistaged procedures that can jeopardise a potentially good outcome if performed in a non-radical field.

The reconstruction was carried on with a three-staged forehead flap as suggested by Menick¹¹ with minor variations (such as prefabricating the inner surface of the forehead flap with a split thickness skin graft and using a costochondral graft to reconstruct the dorsum and two modelled costal cartilage grafts to reconstruct the alae, similarly to what was previously described by Brusati and Colletti).¹²

The young age of the patient, who had no immune deficiencies and had no risk factors whatsoever, posed the suspect of a genetic condition favouring the malignancy, such as Muir-Torre (MTS) and Lynch (LS) syndromes. These syndromes are characterised by the loss of mismatch repair genes, thus increasing the risk of malignancies.^{13 14} An accurate oncological family history is therefore required to identify the patient at high malignancy risk and to provide adequate counselling. The Amsterdam

criteria—which were not met for this patient—could help identifying LT and MTS candidates on whom genetic laboratory tests could be performed.

Last but not least a multidisciplinary approach involving dermatologists, maxillofacial/plastic surgeons, oncologists and radiotherapists is advisable also for ‘minor’ skin malignancies: a destructive surgical approach can be justified only if all specialists agree with the planned treatment and less radical approaches have been ruled out.

Learning points

- ▶ Many diseases can cause reddish swelling on the tip of the nose but they do not produce any mass. A clown nose (CN) is a condition characterised by a rapidly growing brownish mass which invariably suggests the presence of a malignancy.
- ▶ CNs are generally due to metastatic lung or breast diseases. The histological examination is the only instrument that allows to distinguish these secondary malignancies from the less common primary cutaneous squamous or basal cell carcinoma.
- ▶ The differential diagnosis between squamous cell carcinoma (SCC) and keratoacanthoma is not always easy. Even if the final diagnosis requires histological examination, the lack of a complete central crater and, most importantly, a rapid growth usually characterises SCC.
- ▶ An SCC requires wide excision margins of the skin and the underlying structures. The reconstruction requires verified clear margins, lest jeopardising oncological safety.
- ▶ SCC usually affects elderly patients, often with personal history of excessive sun exposure and light skin types. The appearance of an SCC in a young patient should always elicit the suspect of a genetic condition such as Muir-Torre or Lynch syndrome. Accurate family history is therefore mandatory and should be followed in selected patients by genetic screening.

Contributors GC performed all of the surgical treatments. LM performed the histological examination of the specimens. MP was the first dermatologist who examined the patient; after surgery she monitored the patient with clinical examination every 4–6 months. FA performed the biopsies and wrote the article.

Competing interests None.

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