

CASE REPORT



Morpheaform basal cell carcinoma of the nasal ala associated with multiple familial trichoepithelioma reconstructed by anterolateral thigh flap: a case report

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ABSTRACT

The right nasal ala had been destroyed and was diagnosed as morpheaform basal cell carcinoma associated with multiple familial trichoepithelioma (Brooke-Spiegler syndrome). After extended resection, the right buccal and nasal ala defect was reconstructed with a flow-through type anterolateral thigh (ALT) flap, which achieved good functional and cosmetic results.

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Morpheaform basal cell carcinoma; multiple familial trichoepithelioma; nasal ala reconstruction; antero lateral thigh (ALT) flap; Brooke-Spiegler syndrome

Introduction

Multiple familial trichoepithelioma, a disease caused by a genetic mutation, extremely rarely co-occurs with malignant tumor (e.g. basal cell carcinoma [BCC]). We report a case in which morpheaform BCC with extensive defects in the nasal ala was constructed using a free anterolateral thigh (ALT) flap.



Case presentation

The patient was a 50-year-old woman who had a common papular eruption on the nasal area that had been present since 12 years of age. This became blackish and then ulcerated from 48 years of age. When the patient was seen, the nasal wings had already been destroyed, and a biopsy of the 45 × 35 mm ulcer area confirmed the diagnosis of morpheaform BCC. Multiple normal-colored papules on the face were diagnosed as multiple familial trichoepithelioma on a histopathological exam. Multiple familial trichoepithelioma has been implicated in CYLD1 gene abnormalities [1,2]. The diagnosis of Brooke-Spiegler syndrome was based on a clinical diagnosis and family history [1]. Multiple rice-grain-sized nodules on the forehead,

nose, and nasolabial folds were pathologically diagnosed as trichoepithelioma [2]. The father, the father's sister, and the paternal grandfather had the same characteristic symptoms, and the disease was inherited as an autosomal manifestation (dominant inheritance). Based on these two facts, we concluded that the diagnosis in this patient was Brooke-Spiegler syndrome. A genetic analysis including the CYLD gene will be performed in the future.

Surgical intervention

Under general anesthesia, the skin incision line was set with a 10 mm excision margin, and extended resection of the ulcerative lesion was performed. This morpheaform BCC involved the mimic muscles, resulting in extended resection of the right nasal ala, including the greater alar cartilage and a partial anterior wall of the maxilla. This patient was clinically diagnosed with definite BCC at the time of presentation with concomitant ulcerative lesions. Therefore, no immunohistochemical staining was performed to differentiate trichoepithelioma from BCC. The nasal cavity and right maxillary sinus were exposed accordingly (Figure 1). Negative surgical margins were confirmed

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Figure 1. This Aggressive group BCC involved the mimic muscles, resulting in extended resection of the right nasal ala, including the greater alar cartilage and a partial anterior wall of the maxilla. The nasal cavity and right maxillary sinus were exposed at the resection surgery.

by a complete circumferential peripheral and deep margin assessment (CCPDMA).

At two weeks after resection surgery, negative margins were confirmed by examination of the permanent specimen; thus, reconstruction with an ALT flap was planned. A flow-through type ALT flap was harvested from the right thigh, and the flap was filled into the right buccal and nasal ala defects. The lateral femoral circumflex artery and vein (LCFAV) was anastomosed with the facial artery and vein in an end-to-end manner, and the distal end of the descending branch was additionally anastomosed with the superficial temporal artery and vein (Figure 2). The flap was successfully taken and thinning of the flap was performed at 6 months after surgery in order to improve the shape of the nostrils and nasal ala. Further revision of the nasal configuration, including cartilage grafting, will be scheduled in the future (Figure 3).

Pathological findings

Trichoepithelioma lesions

Follicular tissue with hyperplastic sebaceous glands was seen, with deer-horned honeycomb formation of basal cell-like cells in the upper to middle dermis. There was no lacunar formation between the follicle and the surrounding mesenchyme, which was a finding of trichoepithelioma (Figure 4).

Basal cell carcinoma lesions

Atypical hairblast-like cells form spores of various sizes and infiltrative proliferation. The tumor was found to have peripheral palisading and artefactual clefting, invading into the collagen fibers within the dermis and into the adipose tissue, which was a finding of basal cell carcinoma. The mimic muscle layer was also involved, but we secured a negative surgical margin (Figure 5).

Discussion

Multiple familial trichoepitheliomas, also known as Brooke-Spiegler syndrome, are symmetrically distributed on the nasolabial folds, cheeks, forehead, and upper lip, and are characterized by multiple hemispherical, hard, normally colored papules of 3–10 mm in size. The disease is autosomal dominant and has been proven to be caused by mutations in the *CYLD1* gene [1,2]. The mechanism of tumorigenesis has also been elucidated, and it is thought that activation of the NF- κ B signaling pathway results in cell proliferation and—in rare cases—the formation of malignant tumors [3].

There are several reported cases of the development of morpheaform BCC in patients with multiple familial trichoepitheliomas [4,5]; however, there has been no discussion of the reconstructive procedures. In our present case, the destruction of the right nasal

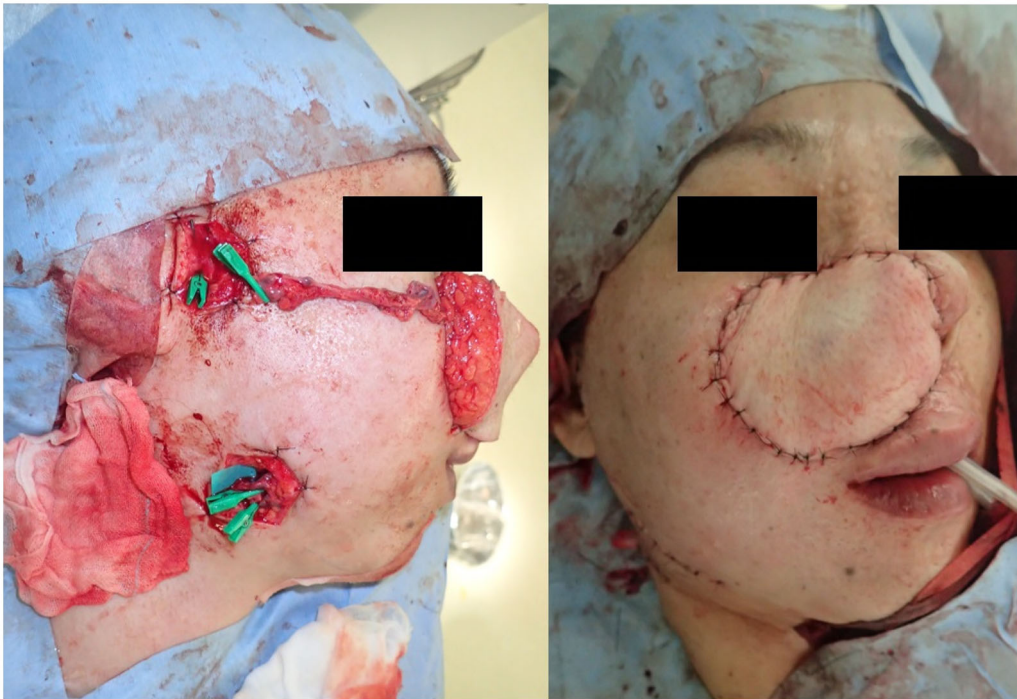


Figure 2. The lateral femoral circumflex artery and vein (LCFAV) was anastomosed with the facial artery and vein in an end-to-end manner, and the distal end of the descending branch was additionally anastomosed with the superficial temporal artery and vein.



Figure 3. The flap was successfully taken and thinning of the flap was performed at 6 months after surgery in order to improve the shape of the nostrils and nasal ala. Further revision of the nasal configuration, including cartilage grafting, will be scheduled in the future.

ala resulted in extensive defects with exposure of the paranasal sinuses and buccal and nasal defects. We decided to use a free flap technique because [1] there were scattered trichoepithelioma lesions in the buccal region as well, and we were concerned about

recurrence due to use a combination of local skin flap, and [2] the defects resected in combination with the nasal ala and maxilla were complex and extensive. A free ALT flap was used to fill the defect, then we tried to reconstruct the nasal shape by folding and

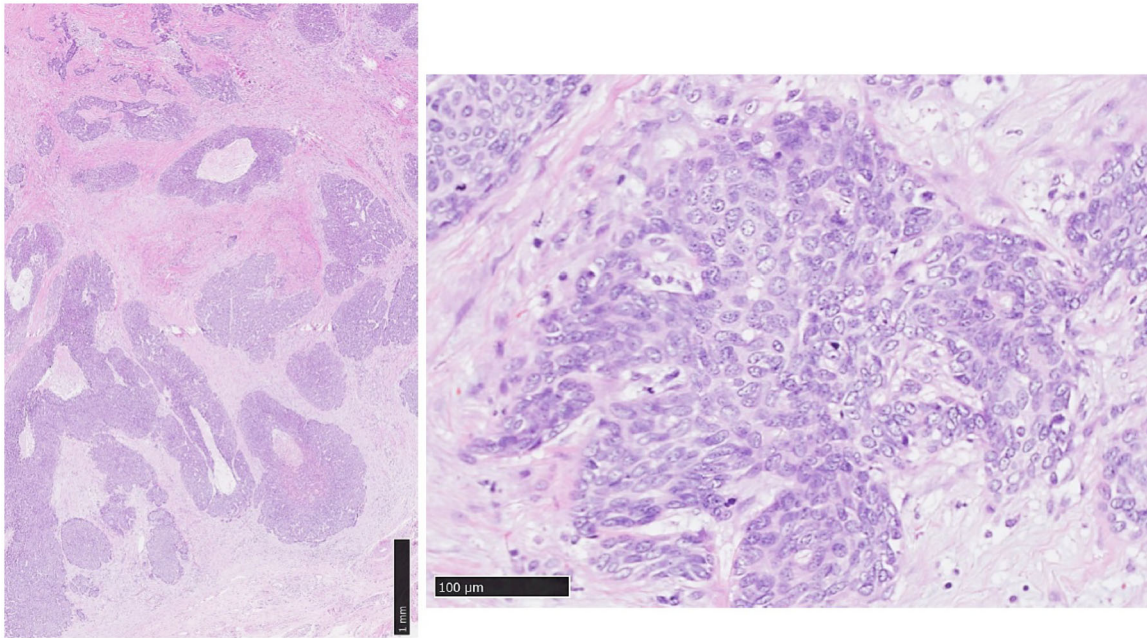


Figure 4. Pathological images of the trichoepithelioma lesions are shown.

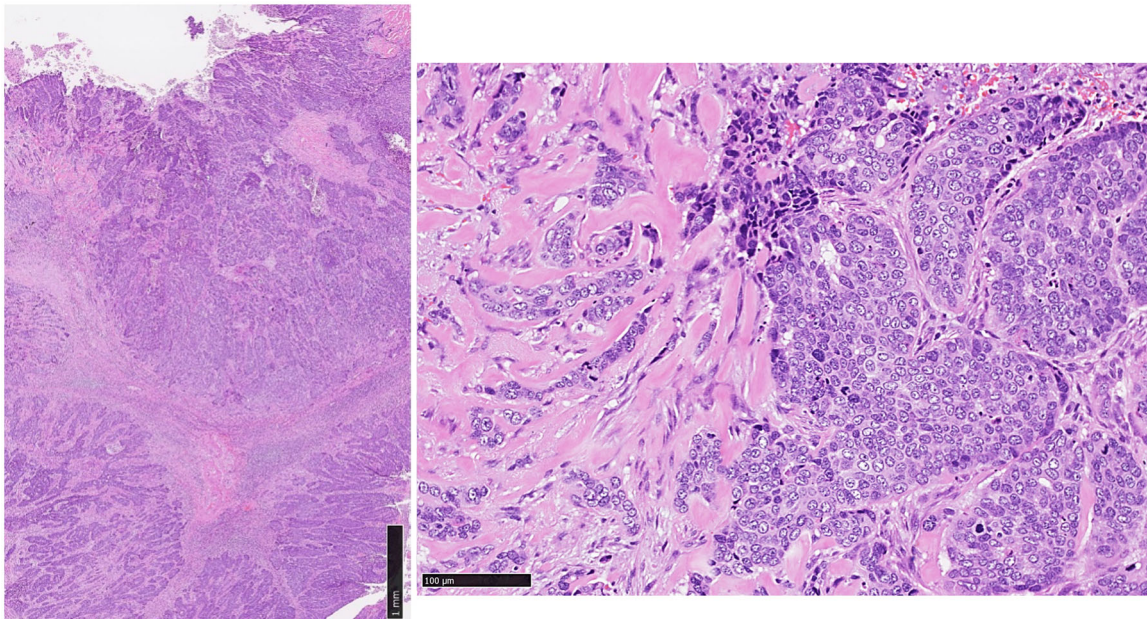


Figure 5. Pathological images of the basal cell carcinoma lesions are shown.

anchoring the flap. The outer margin of the reconstructed nasal ala was thus refixed with inverted sutures to form an acute angle between the skin flaps. Multiple-stage flap reconstruction may be performed for similar defects in combination with forehead flap or rotational cheek flap [6].

However, good functional and cosmetic results could be achieved with reconstruction using a single free flap. In cases involving extensive facial defects caused by this disease, one-stage reconstruction with a free ALT flap can be an alternative reconstructive option.

Conclusion

We experienced the case of a patient diagnosed with morpheiform BCC associated with multiple familial trichoepithelioma (Brooke-Spiegler syndrome). In the past, there have been several reported cases of malignancy associated with multiple familial trichoepithelioma, but there has been no discussion of reconstructive procedures. In particular, there have been no reports of reconstruction using free flaps. Because multiple familial trichoepithelioma involves

scattered lesions in the buccal and forehead areas, reconstruction of the defect using a combination of local skin flaps (e.g. forehead and cheek flaps) carries a risk of recurrence. The first choice for reconstruction in these cases should therefore be free flaps. Even in cases with complex, extensive defects with combined resection of the nasal ala and maxilla, good functional and cosmetic reconstruction is possible if careful corrective surgery is performed.

Informed consent and patient details

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

Disclosure statement

The authors declare no conflicts of interest in association with the present study.

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