CASE REPORT

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Nasal reconstruction in advanced sinunasal sarcoïdosis*

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SUMMARY

Introduction: *Sarcoidosis can lead to devastating nasal deformities and impairment of nasal respiratory function. There is only very little published experience with nasal reconstruction in this disease entity. In general, a cautious attitude prevails out of fear of complications or failure due to the underlying granulomatous inflammation.*

Case report: *A 57-year-old lady presented to our institution with a 20-year history of sarcoidosis with sinunasal involvement. The findings included a saddle nose deformity, absent nasal septum and conchae, left-sided alar collapse and cutaneous involvement of the nasal tip with discoloration. In a first procedure, we performed a reconstructive nasal surgery with porous polyethylene grafts. In a second procedure, we used a paramedian forehead flap for a partial nasal reconstruction. The functional and asthetic improvement was very satisfactory for the patient during the 2-year follow-up period.*

Conclusion: *This is the first reported use of a forehead flap in nasal sarcoidosis. It demonstrates that major reconstructive nasal surgery and implantation of porous polyethylene grafts can successfully be performed in patients with sinonasal sarcoidosis during remission.*

Key words: sarcoidosis, sinunasal, forehead flap, rhinoplasty, porous polyethylene

INTRODUCTION

Sarcoidosis is a systemic granulomatous disease of unsolved etiology. The first case of sarcoidosis mentioned in the literature [1] presented with cutaneous signs, including the nose "being double its normal volume, of a livid purplish red colour, and shining surface, with dilated sebaceaous gland orifices, and shallow erosions in front of the nostrils". Other typical nasal findings in sarcoidosis include purplish colouring of the mucosa with pale yellowish nodules, typically on the septum and inferior turbinates. Later on, septal perforation and saddle nose deformity occur. Often, as in this report, nasal changes emerge at the beginning of the disease. Sinonasal sarcoidosis (SNS) occurs in less than 10% of all sarcoidosis patients and leads to external disfigurement as well as internal lysis and destruction of large parts of the structural framework of the nose and paranasal sinuses. Systemic and - to a lesser degree - topical steroid treatment is the cornerstone of therapy. There is only very little published experience concerning the surgical treatment of functional and aesthetic impairment in this devastating disease. In analogy to Wegener's granulomatosis, many physicians hesitate and do not recommend surgical therapy, out of fear of a flare-up of the condition [2]. When considering different grafts in rhinoplasty and nasal reconstruction in such patients, it seems that irradiated as well as autologous cartilage grafts are resorbed at a higher rate when compared to the general rhinoplasty population [2]. Therefore, some authors recommend inert graft materials, such as silastic [3].

We present a case of advanced SNS with disfiguring cutaneous involvement of the nasal tip and asymmetric alar collapse, which was treated with a functional rhinoplasty using porous polyethylene and a forehead flap nasal reconstruction.

CASE REPORT

A 57-year-old lady presented to our institution with a history of SNS since 20 years. Her main complaints were left-sided nasal obstruction with collapse of the left nasal vestibule and an unsighty discoloration of the tip of her nose.

ENT examination showed a saddle nose deformity, a marked depression of the anterior nasal spine, absent nasal septum and conchae, left-sided alar collapse and cutaneous involvement of the nasal tip with reddish nodules (Figure 1). Thirteen years before, she had undergone conchal cartilage septorhinoplasty.

The sinus CT showed sinus opacification and marked medial bony erosion, with absence of a bony palate (Figure 2).

Multiple nasal biopsy specimens were available for review, showing non-caseating granulomas with giant cells and occasional central necrosis, with peripheral lymphocytic infiltrates and absence of vasculitis. Fungus and mycobacterial stains were consistently negative. On one single occasion, mycobacterial DNA *(M. chelonei)* was detected by PCR, but an exten-

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Figure 1. A 57-year old patient with 20 years history of sinunasal sarcoidosis. Note the saddlenose deformity, a marked depression of the anterior nasal spine, left-sided alar collapse and cutaneous involvement of the nasal tip with reddish nodules.

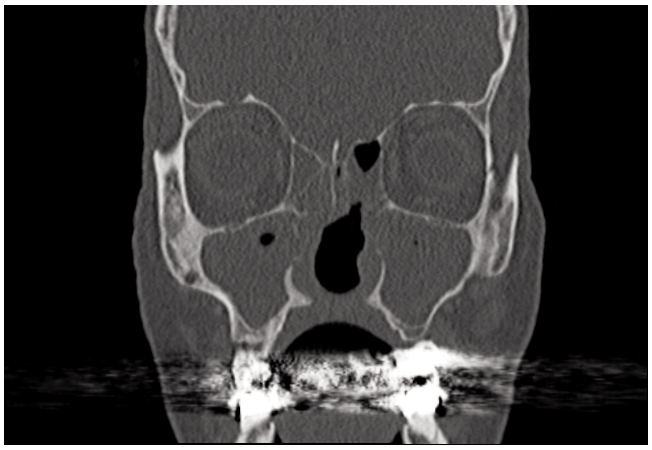

Figure 2. CT appearance: sinus opafication and marked medial bony erosion, with absence of a bony palate.

sive systemic work-up showed this to be of no clinical relevance. The patient's systemic disease activity was low, consisting mainly in an asymptomatic bihilar calcified lymphadenopathy and metacarpal and metatarsal lytic bony lesions causing her only minimal discomfort. Further findings included a moderately elevated serum ACE, a markedly elevated serum IL-2 receptor, and a normal CD4/CD8 T cell ratio in bronchoalveolar lavage fluid.

The patient fulfilled the strict diagnostic criteria for SNS, as proposed by De Shazo et al. [4]. Her SNS was stage III, according to the staging system (stages I to III) developed by Krespi et al. [5].

Initially, the patient mainly wished a functional improvement. In a first reconstructive procedure, the nasal tissue was found to be severely altered by scar formations, and it seemed unlikely to be able to nourish a cartilage graft. Furthermore, we were concerned of rapid resorption due to the underlying granulomatous inflammation, albeit in remission. We therefore used porous polyethylene in a functional nasal reconstructive surgery, due to its stability under such conditions [6]. Two strut-like and alar-Batten-like pieces of porous polyethylene (Medpor®, Porex Surgical, Newnan, USA) were modeled and inserted into the columella and into the left alar rim (Figure 3).

A closed endonasal approach with an alar rim incision was used for placement of the alar implant and a median sublabial incision for placement of the columella strut. In this way, the columella gained some support and the left vestibulum was widened. The columella strut measured 1.5 x 0.4 cm. The alar implant was kidney-shaped and measured 1.8 x 0.5 cm. The implant thickness was 1.5 mm.

Five months later, the patient was more concerned with the aesthetic findings and also not entirely satisfied with the respiratory nasal function, although this was already improved by the procedure described above. After careful consideration of the surgical options, we performed a partial nasal reconstruction with a two-stage right-sided paramedian forehead flap. The area of resection included both nasal alae, half of the columella and the lower third of the nasal dorsum with the nasal tip. Internal lining was provided by a local turn-in flap from the nasal tip skin. The porous PE implants were found to be well integrated and were removed. For structural support we chose rib cartilage in this procedure, due to our experience with the long-term results of porous polyethylene in revision nasal surgery. In a recent review of rhinoplasty procedures with implantation of porous PE [7], we found that this material is not very suitable in multiple revision cases with extensive scar tissue formation, especially if relatively large implant pieces are used. Rib cartilage grafts were modelled to form a columella strut, an anterior septum, bilateral lower lateral cartilages and bilateral lateral wings of alar cartilage. Additionally, a nasal tip graft was modelled and sutured to the rest of the new nasal framework. Four weeks later, the pedicle of the forehead flap was divided and the flap debulked.

The medical management for both surgeries included peri- and postoperative antibiotic prophylaxis with cefuroxime 1.5g i.v. three times daily.

The result was judged very satisfactory by the patient (Figure 4). During the follow-up period of two years, no new manifestations of cutaneous sarcoidosis of the external nose were noted, neither did the surgery provoke a systemic flare-up of the disease. Two years later, the first disease progression occurred, involving the hand and later also the palate, with formation of a palatal fistula.

DISCUSSION

From the literature, it is difficult to estimate the incidence of SNS. Probably it is underestimated in the absence of universal ENT examination in patients that present with thoracic sarcoidosis. The overall incidence of sarcoidosis can be estimated to be between 6 and 10 per 100,000 [8]. Wilson et al. [9] described 27 cases of sarcoidosis (21 biopsy-proven cases) with head and neck involvement among 750 cases of sarcoidosis. In a study from the Mayo Clinic, 220 patients out of 2319 cases of sarcoidosis (9%) had head and neck involvement and only 1% had isolated involvement of the nose and sinuses [10]. It is therefore not surprising that on review of the literature, we did

Figure 3. Status post endonasal approach porous polyethylene functional rhinoplasty.

Figure 4. Status post partial nasal reconstruction with a two-stage right-sided paramedian forehead flap, using a rib cartilage framework.

not find any previously published experience with a forehead flap reconstruction for nasal sarcoidosis.

One main concern in this context is the role that the underlying disease is playing in the postoperative healing period. It is often assumed that surgery, if attempted, should be done in a period of remission. O'Brien used a postauricular full-thickness skin graft in one patient and noted reappearance of cutaneous lesions two years later [(11)]. Reports on reconstructive surgery in granulomatous diseases involving the nose are available for patients with Wegener's granulomatosis. Congdon et al. [(2)] reviewed 13 patients who had mainly undergone dorsal repairs for saddle nose deformity, and found that "disease severity did seem to affect the chance for a successful outcome". But, since all of these patients, like our patient, were operated on during a phase of remission, it remains unclear whether a silent period in the underlying disease activity is necessary for a good result. Another critical point is the choice of graft material. Due to the very limited published experience with patients with SNS, one could reason in analogy to Wegener's granulomatosis, where it has been noted that autogenous materials, such as costal cartilage and calvarial bone had a succes rate of 77% in a series of 13 patients [(2)]. In our patient, porous polyethylene showed a good implantation without complications during five months.

CONCLUSION

In this case of long-standing advanced sinonasal sarcoidosis, with extensive destruction of nasal support structures, functional nasal reconstructive surgery using porous polytehylene as well as reconstruction with a forehead flap yielded a good result and a lasting functional and aesthetic improvement. In patients who are severely affected by this disease, the wish for cosmetic improvement should not be underestimated or overlooked. In the presented case, the patient was not satisfied with the initially desired and accomplished functional gain – and aesthetically corrective surgery was subsequently performed, as per patient wishes.

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