

Keutel syndrome: Augmentation of the nose with serial fat grafting

Sir,

Keutel syndrome is an extremely rare autosomal recessive hereditary syndrome and is caused by mutations within the matrix Gla protein gene.^[1,2] Here, we report a patient with Keutel syndrome whose soft tissue deficiency and nasal deformity were corrected by soft tissue augmentation accomplished through serial fat grafting.

A 20-year-old female with Keutel syndrome was admitted to our outpatient clinic with cosmetic complaints. Physical examination revealed midface hypoplasia with a depressed nasal bridge, a small nose, and short columella [Figure 1]. Following preoperative assessment and paranasal computed tomography imaging, atraumatic extraction of fat fragments from abdominal area and injection of 8 ml fat graft to the supra perichondrial and supra periosteal plans of the nasal tip, dorsum, side walls, and columella, were repeated seven times with intervals of 6 months under sedoanalgesia. Even though, we planned to perform open rhinoplasty for reconstruction of the cartilaginous nasal frame following fat grafting, autologous cartilage graft was not appropriate due to the original disease [Figure 2]. Upper and lower respiratory cartilage ossification have serious impact on symptoms and prognosis of this syndrome. In addition, respiratory failure and death following general anesthesia were reported in Keutel syndrome cases.^[3]

In our case, the patient needed not only soft tissue augmentation but also nasal framework reconstruction. Even though we planned to perform rhinoplasty for reconstruction of the cartilaginous nasal frame with alloplastic material, patient's refusal of this procedure directed us toward nonsurgical follow-ups [Figure 3].

Although rhinoplasty procedure must be the primary choice for patients seeking aesthetic improvement of the nose, the ability to smooth out deformities and asymmetries using fat grafting holds great appeal due to the apparent simplicity and reliability of this technique. For this reason, it's frequently used both in esthetic and reconstructive surgery. Fat grafting is being performed, especially in the correction of rhinoplasty sequels, such as dorsum irregularities, inverted V deformity, prominent lateral osteotomies, and saddle nose deformity.^[4,5] Furthermore, autologous fat grafting shows itself as an alternative to the aesthetic improvement of nasal shape and profile in patients who refuse open or closed rhinoplasties.

There are not enough reports on preoperative fat grafting in order to expand nasal skin and increase soft tissue volume in congenital nasal hypoplasia cases. Mischkowski and Kübler had reported correction of congenital nasal hypoplasia associated with Kallmann syndrome using self-inflating injectable tissue

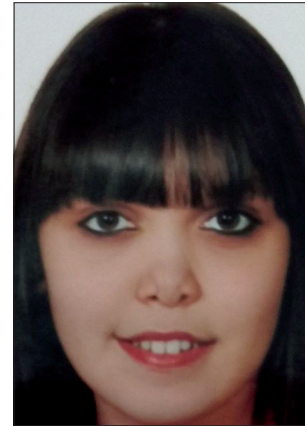


Figure 1: Preoperative view of the case

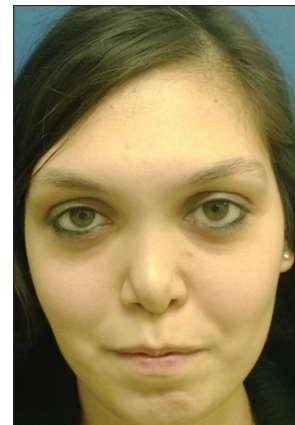


Figure 2: After five sessions of fat grafting

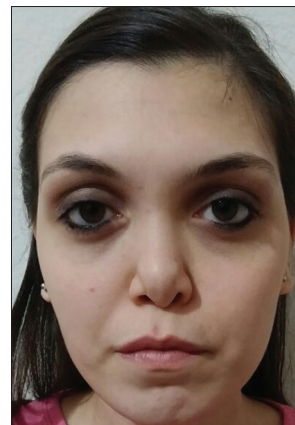


Figure 3: Postoperative view of the case (1 year after seven sessions of fat grafting)

expander pellets. After expansion procedure, in this case, the nasal frame had been reconstructed with an autologous rib graft.^[6]

Even though we did not perform rhinoplasty on our patient, we believe fat grafting to the supra perichondrial and supra periosteal plans can be effectively used in congenital nasal hypoplasia to expand skin and soft tissue volume, and increase skin quality before rhinoplasty procedure.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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