Summary

Progressive hemifacial atrophy (PHA) is a rare disorder characterized by slow, unilateral atrophy of the soft tissues and bones of the craniofacial region. The defect becomes more pronounced with age leading to esthetic and functional deficits. However, the proper timing and method of surgical reconstruction are still debated. The correction of this defect markedly influencing the quality of life of the patient can be achieved with less to more invasive surgical approaches. A 21-year-old female patient with hemifacial atrophy and extensive alopecia presented to our clinic. Considering the body type and the expectations of the patient, the profunda artery perforator flap was applied for the reconstruction and esthetic improvement of the facial region. The facial asymmetry attenuated following the reconvalescence period. This case shows that the use of microvascular free flaps is still inevitable in the up-to-date surgical treatment of severe PHA in order to achieve an acceptable esthetic result. This is the first time that a profunda artery perforator flap was used to restore facial asymmetry caused by PHA.

Keywords: progressive hemifacial atrophy, profunda artery perforator flap, reconstructive surgery, microsurgery.

Introduction

Progressive hemifacial atrophy (PHA) was first reported by Eulenberg in 1871, but the special craniofacial symptoms accompanied by PHA were first described by Parry in 1825, followed by Romberg in 1846.[1] Despite the early description, the accurate etiological factors are still unknown. Clinical studies emphasize an autoimmune origin supported by the common co-occurrence of other autoimmune disorders (e.g. systemic lupus erythematosus or rheumatoid arthritis), while other research raised the possibility of various other etiologies, namely lymphocytic neuro-vasculitis, trauma or sympathetic hyperactivity/dysregulation.[1-5]

This rare condition usually begins in the first two decades of life, mainly in female patients.[6] The craniofacial manifestation of PHA is characterized by progressive unilateral facial atrophy affecting the skin, subcutaneous tissue, muscles, nerves or the underlying bone.[7] However, PHA may also affect other parts of the body (e.g. neck, shoulders, chest, or lower limb).[8]

As a result of the affected dermatomes of the trigeminal nerve with or without bone atrophy (the basis of classification of PHA) and the constantly deteriorating condition/appearance of the patient, different therapeutic approaches are required.[9] Although immunosuppressive treatment (e.g. MTX) may slow down or even stop/halt the progression of the disease, facial defects already present worsen the quality of life of the patients, and they require surgical management in order to minimize the facial asymmetry.[10] However, the physical condition of the patient may have a significant impact on the surgical management and it may limit the potentially available methods.

Case Report

A 21-year-old Caucasian female patient with PHA causing a significant facial asymmetry (diagnosed when she was 5 years old), presented to our clinic in 2015. Physical examination revealed extensive alopecia, localized hyperpigmentation, severe atrophy of the skin and the underlying subcutaneous tissues on the left side of the face between the zygomatic arch, the body of the mandible (affecting the left submental region), the nasolabial fold and the ear (Fig 1). The mimic muscles were not involved. Radiological investigation revealed bone involvement of the body of mandible; however, a relative stable occlusion was observed (Fig 2). The main complaint of the patient was severe facial asymmetry that influenced her social interactions significantly. We decided on soft tissue reconstruction, first

with free flap and further contingent autologous lipotransfer to replace the facial deformity/defect/tissue loss. At the same time, orthodontic treatment started for the preparation of a further bimaxillary osteotomy. Considering the thin body type and the expectations of the patient and comparing all reconstructive possibilities, we decided on the profunda artery perforator (PAP) flap (Fig 3).

Before the surgery, CT angiography was performed and it revealed more sufficient perforant vessels on the right side. During the surgery, the patient was placed in the lithotomy position and was operated on by two surgical teams at the same time. Following the localization of the right profunda artery perforator with a hand-held Doppler probe, an incision was performed on the lateral border of the planned flap preparing the perforant vessels – which are mostly localized 2 cm behind the gracilis muscle – subfascially. Fibres of the adductor magnus muscle were separated gently, reaching the origin/drainage of perforant vessels, and producing a 9 cm long pedicle with artery and vein, whose diameter was 1 mm (at the origin of the right deep femoral artery) and 2 mm (at the drainage into the right deep femoral vein), respectively (Fig 4A). The forepart of the flap was cut using an epifascial preparation. Finally a flap with a size of 9x5 cm was mobilized.

A preauricular incision was made to prepare the recipient area. The skin of the cheek was undermined to the infraorbital margin proximally, the nasolabial fold medially and the mandibular body distally (Fig 4B); the left facial vessels with a length of 4 cm were prepared from a transverse incision, forming a subcutaneous tunnel between these regions. The flap was transplanted to the left facial region and the pedicle was led to the neck through the tunnel. End-to-end anastomoses were applied with two half continuous stitches in the vein and simple interrupted stitches in the artery using 8.0 and 9.0 monofil, non-absorbable sutures (Prolene®; Ethicon, One Johnson & Johnson Plaza, New Brunswick, New Jersey, USA). The ischemia time was 117 minutes. The flap was de-epithelialized and positioned subcutaneously. A monitor skin with a size of 3x1 cm was left preauricularly to observe the circulation of the flap (Fig 4C and D). The recipient and donor site were closed primarily. Since the patient is young and she has no history of hypercoagulability, anticoagulant therapy was not administered and the patient was mobilized on the day of the surgery.

During the surgery, it is not possible to determine the thickness of the flap at the recipient site because of the interstitial edema and hemorrhage that develop locally. Therefore a thickener flap is more favorable in this situation with its correction at a later time (e.g. cut or liposuction), creating a symmetric facial appearance. It takes approximately 1 year for a scar to reach its final form, and the efficacy of lipofilling can also be determined at that time

together with the indication of further correction. Following the absorption of tissue-fluids, the 3-month follow-up revealed a better esthetic outcome without any complications during the recovery period (Fig 5A). Since the quality and the quantity of the facial skin around the monitor flap were found appropriate, and the preauricular monitor flap might have resulted in a less esthetic appearance, one year after the first surgery the monitor flap was cut out, the flap was thinned, and the wound was primarily closed following lipofilling in the mental region. In view of the improved facial appearance and better symmetry, the main complaint of the patient has been managed successfully (Fig 5B). The alopecia will be resolved with hair transplantation performed by a plastic surgeon.

Discussion

There is professional disagreement concerning the proper date and method of the complex surgical management of patients with PHA. Evidence-based knowledge is not available, but it is generally agreed that the surgery should ideally follow the stabilization of the disease process and the completion of facial growth.[1] However, an early intervention may reduce negative psychosocial effects and improve the patient satisfaction, but it can also necessitate further surgeries.[11]

The main goal of the surgical treatment in PHA is to restore the facial contour and resolve facial asymmetry. The possibilities are numerous and largely dependent on the severity of the defect. Slight deformities can be treated with autologous lipotransfer, while microvascular flap procedures provide long-term contour correction in severe cases.[12] The main drawback of lipotransfer or -injetion is subsequent absorption, resulting in significant shrinkage of the injected fat volume, which may reach a volume of about 65% of the transplanted tissue over the first few months. The results can be improved or maintained with the use of tissue-derived stem cells and proper centrifugation technique, but fat injection and en bloc technique also provide a low absorption rate with a relatively non-visible scar.[13,14] The body type of the patient can also be a problem, as it is very hard to harvest fat tissue from skinny patients.

The selection of the proper flap is based on the amount and shape of soft tissue defect, but sometimes on the expectations of the patient as well. In case of severe defects a combined approach of soft tissue and bone augmentation is commonly required. Microvascular flaps combined with orthognatic surgery and/or bone augmentation are considered the gold standard in the restoration of facial asymmetry.[15] Multiple microvascular free flaps were used

successfully in the treatment of PHA, including omentum, rectus, latissimus, serratus, deep inferior epigastric perforators (DIEP), anterolateral thigh or parascapular flaps.[16-20] The largest disadvantage of musculocutaneous flaps is the inactivity-related tissue atrophy that is hard to predict, and which results in failure to fill the atrophied soft tissues. Inframammary extended circumflex scapular (IMECS) flaps are commonly used at our Department to restore hemifacial atrophy of various causes (e.g. facial palsy). This flap has a robust blood supply with low anatomical variability, it can provide the appropriate size, and during the surgery the structure/extension (e.g. full-thickness flap with deepithelialized skin; flap containing fat and fascia, where fat can be thinned; flap containing only fascia folded according to the needs of the recipient site) can be modified, as necessary.[21,22] The primary arguments against an IMECS flap in this case were (1) the asthenic body type of the patient, which would have resulted in a 4-5 mm thinner flap than a PAP flap; (2) the visible scar on the back; and (3) the position of the patient during the operation, which would not allow the simultaneous work of two surgical teams. DIEP flaps are usually too thick for facial reconstruction and very vulnerable for flap thinning techniques.[23,24] Thoracodorsal artery perforator (TDAP) flaps can be also used in head-and neck reconstructive surgery and it is modifiable as a IMECS flap.[25] Although flap harvesting can be performed with the patient in the supine position, it does not provide appropriate accessibility for simultaneous surgery. As our patient refused the use of surgical methods with visible scars at the donor sites (e.g. IMECS or TDAP flap) and the DIEP flap was not feasible because of the body type of the patient, another solution had to be found.

Because of the unmodifiable explicit absorption of musculocutaneous flaps, fasciocutaneous flaps are more frequently adopted and used for the reconstruction of hemifacial microsomia or facial asymmetry.[17,26] The posteromedial thigh region, first described by Ahmadzadeh R in 2007, is a relatively novel donor site in reconstructive surgery.[27] Allen RJ first used it for breast reconstruction, and this surgical team also provided a more detailed anatomical description of the special characteristics of this particular flap.[28,29] Recently, PAP-flaps have been used for breast reconstruction where the abdominal donor sites were not desirable, but some articles demonstrated their use as a pedicle flap for vulvar reconstruction.[30,31] Just a few publications can be found regarding the use of PAP flaps in the maxillofacial region. However, these show a survival rate similar to conventional flap types (radial forearm; anterolateral thigh flaps, ALT) for head and neck reconstruction, as well as a relative ease of harvest, as observed.[32-34] The main advantages are reliable blood supply, sufficient pedicle length (approximately 10 cm) to reach the neck, thick donor tissue and

relatively favorable donor site.[29,34] Publications suggest that PAP flaps may be a good alternative to ALT flap in head and neck reconstruction if the perforant vessel of the ALT flap is not suitable (in approximately 5% of cases), or if the esthetic result at the donor site is more important (e.g. young patients).[33-35]

Since the asthenic body type and the expectations of our patient did not allow the use of other fasciocutaneous flaps, we hypothesized that the PAP flap would offer a good alternative. This flap permits simultaneous surgery by two teams, provides a relatively large size (up to 8x28 cm) and long pedicle length with proper vessel diameters, and if the surgical incision does not extend beyond the midline of the gluteal crease, the scar is barely visible.[29,34] This flap harvested from the upper medial and posterior thigh contains skin and fat tissue, providing a sufficient flap size even in a thin patient for reconstructing soft tissue defects; however, the narrow width can limit the harvested tissue volume.[4] Unlike IMECS flaps, where almost always the same anatomical situation can be found, in case of PAP flaps CT angiography is necessary to determine the suitability of the perforant vessel.[21,36] Although PAP flaps are not as modifiable as IMECS flaps, its thickness matched the recipient site more in this case, and it provided a barely visible scar in the gluteal fold. As in our case, PAP flaps are suitable to minimize facial asymmetry and to replace soft tissue defects. This case report may draw attention to the importance of interdisciplinary cooperation and individualized surgical management of patients. However, further clinical investigations/experience or prospective multicenter studies are required to evaluate the clinical significance and more common use of this flap and in head and neck reconstructive surgery.

Conclusion

This is the first description of the use of a PAP-flap for the reconstruction of facial deformity in a patient with PHA. To choose the best therapeutic option for patients, all potential treatment modalities have to be considered, which may be influenced by the physical status or request of the patient, sometimes necessitating the use of uncommon reconstructive surgical solutions. We have to emphasize the importance of interdisciplinary cooperation, the development of surgical skills and knowledge, which are essential for the proper management of complex cases. PAP flaps may be useful in certain cases where the individual surgical plan has a particular intended impact, and the surgeon has the possibility to choose from a wider armamentarium instead of being forced to apply the most frequently used flaps (radial forearm and ALT flaps) when solving a surgical situation in the head and neck region.

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Conflict of Interest Disclosures

None of the authors reported any disclosures.

References

1. Tolkachjov SN, Patel NG, Tollefson MM: Progressive hemifacial atrophy: a review. Orphanet J Rare Dis 10:39, 2015
2. Pensler JM, Murphy GF, Mulliken JB: Clinical and ultrastructural studies of Romberg's hemifacial atrophy. Plast Reconstr Surg 85:669; discussion 675, 1990
3. Fry JA, Alvarellos A, Fink CW, Blaw ME, Roach ES: Intracranial findings in progressive facial hemiatrophy. J Rheumatol. 19:956, 1992
4. Garcia-de la Torre I, Castello-Sendra J, Esgleyes-Ribot T, Martinez-Bonilla G, Guerrerosantos J, Fritzler MJ: Autoantibodies in Parry-Romberg syndrome: a serologic study of 14 patients. J Rheumatol 22:73, 1995
5. Lonchampt P, Emile J, Pélier-Cady MC, Cadou B, Barthelaix A: Central sympathetic dysregulation and immunological abnormalities in a case of progressive facial hemiatrophy (Parry-Romberg disease). Clin Auton Res 5:199, 1995
6. Raposo-Amaral CE, Denadai R, Camargo DN, Artioli TO, Gelmini Y, Buzzo CL, Raposo-Amaral CA: Parry-Romberg syndrome: severity of the deformity does not correlate with quality of life. Aesthetic Plast Surg 37:792, 2013
7. Cory RC, Clayman DA, Faillace WJ, McKee SW, Gama CH: Clinical and radiologic findings in progressive facial hemiatrophy (Parry-Romberg syndrome). AJNR Am J Neuroradiol 18:751, 1997
8. Duymaz A, Karabekmez FE, Keskin M, Tosun Z: Parry-Romberg syndrome: facial atrophy and its relationship with other regions of the body. Ann Plast Surg 63:457, 2009
9. Iñigo F, Jimenez-Murat Y, Arroyo O, Fernandez M, Ysunza A: Restoriation of facial contour in Romberg’s disease and hemifacial microsomia: experience with 118 cases. Microsurgery 20:167, 2000

10. Tollefson MM, Witman PM: En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. J Am Acad Dermatol 56:257, 2007
11. Slack GC, Tabit CJ, Allam KA, Kawamoto HK, Bradley JP: Parry-Romberg reconstruction: optimal timing for hard and soft tissue procedures. J Craniofac Surg 23:1969, 2012
12. Agostini T, Spinelli G, Marino G, Perello R: Esthetic restoration in progressive hemifacial atrophy (Romberg disease): structural fat grafting versus local/free flaps. J Craniofac Surg 25:783, 2014
13. Livaoğlu M, Yavuz E: Soft tissue augmentation with autologous fat graft: the dissected pouch technique. J Cutan Aesthet Surg 2:21, 2009
14. Koh KS, Oh TS, Kim H, Chung IW, Lee KW, Lee HB, Park EJ, Jung JS, Shin IS, Ra JC, Choi JW: Clinical application of human adipose tissue-derived mesenchymal stem cells in progressive hemifacial atrophy (Parry-Romberg disease) with microfat grafting techniques using 3-dimensional computed tomography and 3-dimensional camera. Ann Plast Surg 69:331, 2012
15. Hu J, Yin L, Tang X, Gui L, Zhang Z: Combined skeletal and soft tissue reconstruction for severe Parry-Romberg syndrome. J Craniofac Surg 22:937, 2011
16. Jurkiewicz MJ, Nahai F: The use of free revascularized grafts in the amelioration of hemifacial atrophy. Plast Reconstr Surg 76:44, 1985
17. Siebert JW, Anson G, Longaker MT: Microsurgical correction of facial asymmetry in 60 consecutive cases. Plast Reconstr Surg 97:354, 1996
18. Cheng J, Shen G, Tang Y, Zhang Z, Qiu W, Lu X: Facial reconstruction with vascularised serratus anterior muscle flap in patients with Parry-Romberg syndrome. Br J Oral Maxillofac Surg 48:261, 2010

19. Wójcicki P, Zachara M: Surgical treatment of patients with Parry-Romberg syndrome. Ann Plast Surg 66:267, 2011
20. Si L, Zeng A, Qiao Q, Liu Z, Zhao R, Wang Y, Zhu L, Wang X: Microsurgical correction of progressive facial hemiatrophy using free anterolateral thigh adipofascial flap. J Craniofac Surg 23:2051, 2012
21. Siebert JW, Longaker MT, Angrigiani C: The inframammary extended circumflex scapular flap: an aesthetic improvement of the parascapular flap. Plast Reconstr Surg 99:70, 1997
22. Albarah A, Kishk T, Megahed M, Elsakka D, Ghareeb F: Pre-expanded Extended Island Parascapular Flap for Reconstruction of Post-burn Neck Contracture. Ann Burns Fire Disasters 23:28, 2010
23. Chana JS, Odili J: Perforator flaps in head and neck reconstruction. Semin Plast Surg 24:237, 2010
24. Prasetyono TO, Bangun K, Buchari FB, Rezkini P: Practical considerations for perforator flap thinning procedures revisited. Arch Plast Surg 41:693, 2014
25. Guerra AB, Lyons GD, Dupin CL, Metzinger SE: Advantages of perforator flaps in reconstruction of complex defects of the head and neck. Ear Nose Throat J 84:441, 2005
26. Tanna N, Wan DC, Kawamoto HK, Bradley JP: Craniofacial microsomia soft-tissue reconstruction comparison: inframammary extended circumflex scapular flap versus serial fat grafting. Plast Reconstr Surg 127:802, 2011
27. Ahmadzadeh R, Bergeron L, Tang M, Geddes CR, Morris SF: The posterior thigh perforator flap or profunda femoris artery perforator flap. Plast Reconstr Surg 119:194, 2007
28. Allen RJ, Haddock NT, Ahn CY, Sadeghi A: Breast reconstruction with the profunda artery perforator flap. Plast Reconstr Surg 129:16e, 2012

29. Saad A, Sadeghi A, Allen RJ: The anatomic basis of the profunda femoris artery perforator flap: a new option for autologous breast reconstruction--a cadaveric and computer tomography angiogram study. J Reconstr Microsurg 28:381, 2012
30. Healy C, Allen RJ Sr: The evolution of perforator flap breast reconstruction: twenty years after the first DIEP flap. J Reconstr Microsurg 30:121, 2014
31. Huang JJ, Chang NJ, Chou HH, Wu CW, Abdelrahman M, Chen HY, Cheng MH: Pedicle perforator flaps for vulvar reconstruction--new generation of less invasive vulvar reconstruction with favorable results. Gynecol Oncol 137:66, 2015
32. Scaglioni MF, Kuo YR, Yang JC, Chen YC: The Posteromedial Thigh Flap for Head and Neck Reconstruction: Anatomical Basis, Surgical Technique, and Clinical Applications. Plast Reconstr Surg 136:363, 2015
33. Mayo JL, Canizares O, Torabi R, Allen RJ Sr, St Hilaire H: Expanding the Applications of the Profunda Artery Perforator Flap. Plast Reconstr Surg 137:663, 2016
34. Wu JC, Huang JJ, Tsao CK, Abdelrahman M, Kolios G, Cheng MH: Comparison of Posteromedial Thigh Profunda Artery Perforator Flap and Anterolateral Thigh Perforator Flap for Head and Neck Reconstruction. Plast Reconstr Surg 137:257, 2016
35. Lu JC, Zelken J, Hsu CC, Chang NJ, Lin CH, Wei FC, Lin CH: Algorithmic approach to anterolateral thigh flaps lacking suitable perforators in lower extremity reconstruction. Plast Reconstr Surg 135:1476, 2015
36. Haddock NT, Greaney P, Otterburn D, Levine S, Allen RJ: Predicting perforator location on preoperative imaging for the profunda artery perforator flap. Microsurgery 32:507, 2012

Figure legends

FIGURE 1. Preoperative appearance of the patient.

FIGURE 2. Orthopantomogram image of the patient.

FIGURE 3. Surgical management of the patient.

FIGURE 4. Preparation of the profunda artery perforator flap (A) and the subcutaneous tunnel on the left face (B). Positioning (C) and fixation of the de-epithelized flap (D).

FIGURE 5. The appearance of the patient 3 months (A) and approximately 1 year after the first surgery (B).