

Hemifacial Atrophy: A Case Report And Review Of Literature

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Citation

T Esan, A Olusile. *Hemifacial Atrophy: A Case Report And Review Of Literature*. The Internet Journal of Dental Science. 2003 Volume 1 Number 1.

Abstract

A case report of hemifacial atrophy and review of literature is presented in this paper.

A variety of treatment techniques have been employed in an attempt to improve the atrophic deformity especially in moderate and severe cases. These have included free fat grafts, dermis fat grafts, fascia, cartilage and bone for augmentation purposes.

However when dealing with mild cases of hemifacial atrophy, prostheses are particularly useful, in that facial disproportion could be easily lifted up to an acceptable aesthetic condition. Their relative cheap cost, ease of fabrication and more importantly psychological relief from the fear of expensive and multi stage surgical technique is a welcome development for the patient. Moreover, in our environment, where facilities for such advanced procedures are lacking, a simple prosthesis may be the treatment of choice.

INTRODUCTION

Hemifacial atrophy, originally described by Parry₁ and Hensch and Romberg₂, consists of slowly progressive atrophy of the soft tissues of essentially half the face which is characterized by progressive wasting of subcutaneous fat, sometimes accompanied by atrophy of skin, cartilage, bone, muscle₃. Although the atrophy is usually confined to one side of the face and cranium, it may occasionally spread to the neck and one side of the body_{4,5} and it is accompanied usually by contralateral Jacksonian epilepsy, trigeminal neuralgia, and changes in the eyes and hair_{6,7}. Evidence of a mendelian basis is lacking. Lewkonja and Lowry₈ reported the case of a 16-year-old boy who developed facial changes at age 7 and had localized scleroderma on one leg and the trunk. The presence of antinuclear antibodies in his serum suggested that the Parry-Romberg syndrome may be a form of localized scleroderma. That Hemifacial atrophy is a form of localized scleroderma is supported by its concurrence with scleroderma.^{7,9,10,11}

Larner and Bennison₁₂ has also reported discordance in a pair of monozygotic twins. The affected twin was a 23-year-old male Caucasian who complained of progressive wasting of the right side of his face, first noted at age 17. The patient was left handed, whereas his twin was right handed. In

addition there was no bony or cerebral asymmetry in the skull radiographs and CT brain scan respectively. Monozygosity was established by DNA fingerprints.

Other synonyms for Hemifacial atrophy include Parry-Romberg syndrome, Romberg-Parry syndrome, Progressive facial Hemiatrophy, Progressive hemifacial atrophy.

A case report of hemifacial atrophy is presented in the study, in which simple prosthetic appliance was used to manage them.

CASE REPORT

A 20- year- old female student presented with a complaint of difficulty with mastication and unpleasing appearance. The left side of her face had been smaller than the right as far as she can remember. She denied any history of trauma or infection to that side of the face.

There was no positive family history of facial atrophy. Neither was there a family history of consanguinity. Her delivery was not assisted per vagina. The medical history was not contributory and was not on any form of medication.

A systemic review was none revealing. Neurological assessment by experts showed no defects with any of the 12 cranial nerves. Neither did a dermatological review reveal

any dermatological pathology.

On physical examination, all her vital signs were within normal limits. There was a furrow in the midline of the forehead. There was a slight shuffling of gait to the left side. Extra-orally, there is facial asymmetry with the left side been slightly smaller than the right side. There was also the collapse of the left cheek (Fig 1&2). She had ptosis of the left eye and refractive errors but no other ophthalmic defects were noted.

Figure 1

Fig 1: Showing facial asymmetry



Figure 2

Fig 2: Left side profile showing facial collapse



Figure 3

Figure 3: Showing atrophic left palate



Figure 4

Figure 4: Showing atrophic left mandible



Figure 5

Figure 5: Showing the upper and lower casts of the jaw s



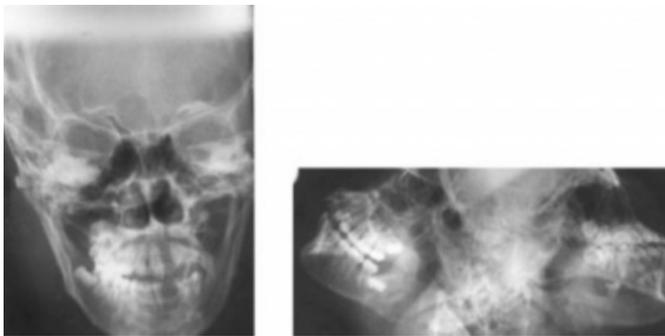
Intra-orally the oral hygiene was fair, the oral mucosa and the tongue looked clinically healthy. The left palatal shelf appeared smaller than the right and so was the left half of the mandible (figure3 &4). Only the right third molars were missing on the upper and lower jaw. The other teeth present

appear to have normal shape and size. On the left however, the maxillary incisors, mandibular canine and the mandibular first molars were present. The maxillary incisors were fused. All the mandibular teeth were small for size and also grossly mobile. The residual ridges were narrow but firm (Figure5).

Postero-Anterior X-ray view revealed deviation of the jaws and nasal septum to the left. There is hypoplasia of the frontal and ethmoidal sinuses. The left orbit looked smaller than the right orbit. X rays also revealed teeth with short roots. The ramus of the mandible on the left is shorter than the right with the right ramus appearing more dense than the left.(Fig 6)

Figure 6

Figure 6: Posterior anterior and oblique views of the skull



A diagnosis of hemifacial atrophy was made.

Various options of treatment were given to the patient. Due to financial constraints, we decided to conservatively manage her with the aim of improving her facial profile and mastication.

She was then referred to the oral surgery unit where extraction of all upper and lower left teeth were done.

Upper and lower alginate impressions were taken to fabricate upper and lower acrylic based dentures. Three days later secondary impressions were taken after border moulding the special tray with green stick impression compound and the wash impression was taken with polysulphide impression material.

The impression was boxed-in and then poured in dental stone. The upper and lower record blocks were constructed on the master cast for registration of occlusion (figure 7).

Figure 7

Figure 7: Showing the bite blocks



Bite registration was taken about a week later, where the labial fullness of the upper and lower left record rim were adjusted to satisfactory position. Labially, the record rim was about 2mm below the level of the upper lip at rest.

With the occlusal surfaces of the teeth on the right making even contact, the record blocks were sealed. The bite blocks with the master cast were then mounted on an articulator. Selection of teeth was done together with the patient using shade guide under natural light. Semi anatomical teeth were selected for the posterior teeth.

The anterior teeth were set in front of the alveolar ridge, while the lower and upper posterior teeth were set over the center of the residual alveolar ridge and lateral to the upper ridge respectively. Subsequently trying-in of the upper and lower dentures was done, and at this phase the fit, stability, base extension, tooth position, support and patient's facial profile were evaluated. The patient was pleased with the appearance in wax and did not make any request for adjustment. The trial dentures were properly sealed down and then processed in acrylic and later fitted. This design was to uplift the left side of her face and also to improve her masticatory efficiency (Fig 8). After delivery, the patient was satisfied and discharged after a few uneventful recall visits.

Figure 8

Fig 8: Showing the facial profile after fitting of dentures



DISCUSSION

Hemifacial atrophy is a rare condition that occurs sporadically although some familial distribution has been found¹³. The majority of cases are sporadic with no definite inheritance being proven in the literature.

The disease occurs more frequently in women with female to male ratio been 3:2³. It has a slight predilection for the left side and appears in the first or second decades of life³. It progresses over a time course of between two and 10 years, and atrophy appears to follow the distribution of one or more divisions of the trigeminal nerve. The resulting facial flattening may be mistaken for Bell's palsy¹⁴.

The aetiology has been the subject of considerable debate^{7,15,16}. Wartenburg⁷ considered the primary factor to be a cerebral disturbance leading to increased and unregulated activity of sympathetic nervous system, which in turn produced the localized atrophy through its trophic functions conducted by way of sensory trunks of trigeminal nerve. Other workers^{15,16} suggested extraction of teeth, local trauma, infection and genetic factors could also be a cause. In a paper published in 1973, Poswillo¹⁷ attributed the

development of facial deformities to the disruption of stapedia artery. The stapedia artery functions as a stopgap vascular channel during days 33-45 of embryologic development. Poswillo fed pregnant rats with triazene and pregnant monkeys' thalidomide and showed the consistent mal development of first and second branchial arch structures. Robinson¹⁸ in 1987 supported Poswillo's theory by demonstrating carotid flow abnormalities in two and defects related to vascular disruption in a third child with craniofacial microsomia.

Hemifacial atrophy is a syndrome with diverse presentation. The most common early sign is a painless cleft, the "coup de sabre", near the midline of the face or forehead. This marks the boundary between normal and atrophic tissue. A bluish hue may appear in the skin overlying atrophic fat¹⁹.

The affected area extends progressively with the atrophy of the skin, subcutaneous tissue, the muscles, bones, cartilages, alveolar bone and soft palate on that side of the face. In addition to facial wasting that may include the ipsilateral salivary glands and hemiatrophy of the tongue, unilateral involvement of the ear, larynx, oesophagus, diaphragm, kidney and brain have been reported^{11,14}.

It starts in the first decade and lasts for about 3 years before it becomes quiescent. The final deformity varies widely, burning itself out in some patients with minimal atrophy, while in others progressing to marked atrophy.

Neurological disorders are found in 15 % of patients with this disorder, while ocular findings occur in 10-40%, the most common one being enophthalmos^{20,21}

Rarely, half of the body may be affected. This condition may be accompanied by pigmentation disorders, vitiligo, pigmented facial naevi, contralateral jacksonian epilepsy, contralateral trigeminal neuralgia and ocular complications. Dental abnormalities include incomplete root formation, delayed eruption, severe facial asymmetry resulting in facial deformation and difficulty with mastication²².

The evaluation of hemifacial atrophy includes a thorough history and physical examination, photographic and cephalometric analysis, and three dimensional computed tomographic study. Family history of consanguinity, intrauterine exposure to infection and toxins, and problems with delivery should be explored. Physical examination should focus on facial asymmetry as well as on isolated findings consistent with this syndrome. Investigations such as three dimensional computed tomography provides

accurate reconstruction of patients' craniofacial skeletons, alleviates the need for constructing physical models and allows for faster and more accurate surgical planning. Photographs and cephalometry will allow for monitoring of facial symmetry over time and aid in planning surgical approaches to individual patients.

Its differential diagnosis includes post traumatic fat atrophy, hemifacial microsomia (first and second branchial arch syndrome), Goldenhar's syndrome, and partial lipodystrophy which is, however, always bilateral^{7, 11, 23, 24}.

Hemifacial atrophy remains almost as much an enigma today as it was when first reported by Romberg in 1846.

Presently there is no known definitive treatment but all available treatment schemes are adapted to the specific dysmorphology of individual patients which is geared to improving the facial profile and also the masticatory efficiency of the patient.

The basic principle of surgical treatment is to correct first the bony tissue deficits by the use of onlays, inlays, osteotomies and ostectomies with or without bone graft followed by soft tissue augmentation. The mandible is addressed initially since correction of mandibular malformations often stimulates maxillary growth. Maxillary growth is further enhanced with the use of maxillary activators. Costochondral grafts must be used in TMJ reconstructions. Soft tissue deficits are corrected with local and microvascular free flaps. Facial nerve defects usually are permanent and hearing must be assessed early to allow for hearing augmentation.

A variety of treatment techniques have been employed in an attempt to improve the atrophic deformity. These have included free fat grafts, dermis fat grafts, fascia, cartilage and bone for augmentation purposes^{3, 21}. As the grafts, in many instances, are partially or completely reabsorbed, pedicle flaps and more recently omentum and de-epithelialized skin flaps transferred utilizing microvascular techniques have been employed^{3, 4}. While pedicle and microvascular free flaps allow for the permanent correction of large deformities, they have the disadvantages of being more lengthy procedures and result in additional scarring to both the donor and the affected sites. Deformities have also been corrected with alloplastic materials including liquid silicone which was previously used with great enthusiasm^{21, 25, 26, 27, 28}. The large volumes of silicone often required, however, have led to late complications which include chronic relapsing inflammation, skin ulceration and sinus tracts²⁹.

As subcutaneous fat is the tissue most commonly affected in hemifacial atrophy, it should ideally be replaced. In recent years, the injection of aspirated fat has been used for the correction of acne scars as well as for furrows, deep wrinkles and depressions resulting from liposuction surgery^{30, 31, 32}. The correction of hemifacial atrophy utilizing fat injection has also been described³³. Unlike prior fat grafting, which involved the harvesting of large pieces of fat, extraction of adipose tissue through a cannula or needle creates a culture of adipocytes which is believed to lead to more rapid revascularization and less reabsorption. The long-term survival of injected fat is as yet unknown; however, good results at three years appear encouraging^{30, 31, 32, 33}.

However when dealing with mild cases of hemifacial atrophy, Prostheses are particularly useful, in that facial disproportion could be easily lifted up to an acceptable aesthetic condition. Their relative cheap cost, ease of fabrication and more importantly psychological relief from the fear of expensive and multi stage surgical technique is a welcome development for the patient.

Moreover, in our environment, where facilities for such advanced procedures are lacking, a simple prosthesis may be the treatment of choice.

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References

1. Parry CH: Collections from the unpublished medical writings of the late Caleb Hillier Parry. Vol. I. London: Underwoods (pub.) 1825. Pp. 478 only.
2. Hensch E, Romberg HM: Klinische Ergebnisse. Berlin: A. Forstner (pub.) 1846. Pp. 75-81.
3. Rogers BO: Progressive facial hemiatrophy: Romberg's disease: a review of 772 cases. Proc 3d Int Cong Plast Surg. Excerpta Medica ICS 1964; 66: 681-689.
4. Rogers BO: Embryology of the face and introduction to craniofacial anomalies. In: Converse JM (ed). Reconstructive Plastic Surgery, 2nd ed. WB Saunders, Philadelphia, 1977: 2296-2358.
5. Hickman JW, Sheils WS: Progressive facial hemiatrophy: report of a case with marked homolateral involvement. Arch Intern Med 1964; 113: 716-720.
6. Walsh FB: Facial hemiatrophy: report of 2 cases. Am. J. Ophthalm. 1939;22: 1-10.
7. Wartenberg R: Progressive facial hemiatrophy. Arch. Neurol. Psychiat. 1945; 54: 75-96.
8. Lewkonia RM, Lowry RB: Progressive hemifacial atrophy (Parry-Romberg syndrome) report with review of genetics and nosology. Am. J. Med. Genet. 1983;14: 385-390.
9. Lakhani PK, David TJ: Progressive hemifacial atrophy with scleroderma and ipsilateral limb wasting (Parry-

- Romberg syndrome). *J. Royal Soc. Med.* 1984; 77: 138-139.
10. Adebajo AO, Crisp AJ, Nicholls A, Hazleman BL: Localized scleroderma and hemiatrophy in association with antibodies to double-stranded DNA. *Postgrad. Med. J.* 1992; 68: 216-218,.
11. Rees TD: Facial atrophy. *Clin Plast Surg* 1976; 3: 637-647.
12. Lerner AJ, Bennison DP : Some observations on the aetiology of progressive hemifacial atrophy ('Parry-Romberg syndrome'). (Letter) *J. Neurol. Neurosurg. Psychiat.* 1993;56: 1035-1039.
13. Witkop CJ, jr : Genetic disease of the oral cavity: In Teiecke, R.W (ed): New York, McGraw Hill 1965.
14. Lederman RJ: Progressive facial and cerebral hemiatrophy. *Cleve Clin Q* 1984; 51: 545-548.
15. Davis WB: Reconstruction of hemiatrophy of the face: case report. *Plastic Reconstr. Surg* 1968; 42: 489-491.
16. Vickey IM: Hemifacial atrophy. *Brit.J.oral surg* 1972; 9: 102-109.
17. Poswillo D: The pathogenesis of the first and second branchial arch syndrome. *Oral Surg Oral Med Oral Pathol* 1973;35:302-328.
18. Robinson LK, Hoyme HE, Edwards DK, Jones KL: Vascular pathogenesis of unilateral craniofacial defects. *J Pediatr* 1987; 111:236-239.
19. Ho KH. Hemifacial atrophy (Romberg's disease). *Brit. Dent. J* 1987;162(5):182-84.
20. Poskanzer DC: Hemiatrophies and hemihypertrophies. In: Vinken PJ, Bruyn GW (eds). *Handbook Clin Neurol*, Amsterdam, North-Holland, 1975; 22: 545-554.
21. Muchnick RS, Aston SJ, Rees TD: Ocular manifestations and treatment of hemifacial atrophy. *Am J Ophthalmol* 1979; 88: 889-897.
22. Foster TD. The effects of hemifacial atrophy on dental growth. *Brit.dent.J* 1979; 146:148-150
23. Gorlin RJ, Jue KL, Jacobsen U, Goldschmidt E: Oculoauriculovertebral dysplasia. *J Pediatr* 1963; 63: 991-996.
24. Gorlin RJ, Pindborg JJ, Cohen MM: *Syndromes of the Head and Neck*. McGraw-Hill, New York, 1976: 546-552.
25. Rees TD, Coburn RJ: Silicone treatment of partial lipodystrophy. *JAMA* 1974; 230: 868-873.
26. Rees TD, Ashley FL, Delgado JP: Silicone fluid injections for facial atrophy. *Plast Reconstr Surg* 1973; 52: 118-127.
27. Ashley FL, Thompson DP, Henderson T: Augmentation of surface contour by subcutaneous injections of silicone fluid. *Plast Reconstr Surg* 1973; 57: 8-13.
28. Ashley FL, Braley S, McNall EG: The current status of silicone injection therapy. *Surg Clin Alorth Am* 1971; 51: 501-509.
29. Jurkiewicz MJ, Nahai F: The use of free revascularized grafts in the amelioration of hemifacial atrophy. *Plast Reconstr Surg* 1985; 76: 44-54.
30. Bart RS, Kopf AW: Hemifacial atrophy. *J Dermatol Surg Oncol* 1978; 4: 12-14.
31. Ellenbogen R: Free autogenous pearl fat grafts in face a preliminary report of a rediscovered technique. *Ann Plast Surg* 1986; 16: 179-185.
32. Asken S: A manual of liposuction surgery and autologous fat transplantation under local anesthesia, 2nd ed. Keith C. Terry and Associates, Irvine, 1986, pp 119-151.
33. Agris J: Autologous fat transplantation: a 3-year study. *Am J Cosm Surg* 1987; 4: 95-102.

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