Indian Journal of Basic and Applied Medical Research; March 2020: Vol.-9, Issue- 2. P. 182 - 186
DOI: 10.36848/IJBAMR/2020/12210.51285

Case Report:

A man with progressive wasting of left side of the face: Case report -Hemifacial Atrophy

[1] Anwar N*, [2] Habib A, [3]Islam MR, [4]Shahidullah Md, [5]Anwar NB

1. Nayeem Anwar, Assistant Professor, Dept. of Neurology, BSMMU, Bangladesh.
2. Ahsan Habib, Associate Professor, Dept. of Neurology, BSMMU, Bangladesh.
3. Md. Rafiqul Islam, Professor, Dept. of Neurology, BSMMU, Bangladesh.
4. Md. Shahidullah, Associate Professor, Dept. of Neurology, BSMMU, Bangladesh.
5. Nawreen Binte Anwar, FCPS (Course student-Ophthalmology), Bangladesh.

Corresponding author*; Email :

Abstract:

Hemifacial Atrophy is an uncommon idiopathic medical condition where there is progressive atrophy of one side face. A 37 year old male with progressive atrophy of left side of face from Bangladesh is reported. The literature on this condition is reviewed. Clinically, the skin can be dry, thin and with a dark pigmentation. Some patients present a demarcation line between normal and abnormal skin, resembling a big linear scar, known as coup de sabre (French term which means “cut of the sword”). This finding was not noticed in this case reported. Ocular involvement is common, and the most frequent manifestation is the enophthalmy, due to fat loss around the orbit. The extension of atrophy is frequently limited to on one side of the face, and the ipsilateral involvement of body is rare (23 to 10% of cases were described as being bilateral). In the case presented here, there was involvement of only one side of the face, important features of this disease are enophthalmos, the deviation of mouth and nose to the affected side, and the unilateral exposition of teeth (when lips are involved) , which were not found in our case. Radiographically, teeth of affected side can present some deficiency in root development and, consequently, delayed eruption. However, the affected teeth are normal and vital clinically. The intraoral soft tissue and muscles of mastications are usually normal, and there is no difficulty in jaw movement, speech or deglutition limitations . The dental findings may include delayed ipsilateral tooth eruption, atrophic root development, missing teeth, retarded root formation, oligodontia, microdontia, dilacerations, unilateral crossbite, and pulp stones on the affected side. No such findings were manifested in our patient on clinical observation. Parry-Romberg syndrome is a rare condition manifesting as atrophy of one side of the face. In most cases, the cause of this syndrome appears to be unknown. The pathophysiology of the condition still remains unknown. Treating this condition can be frustrating for both physician and patient as there is no established treatment to limit or improve the condition.

Keywords: Hemifacial Atrophy, Parry-Romberg Syndrome.

Introduction:

Parry in 1825 and Romberg in 1846 was first to describe unilateral progressive atrophy of the face [1]. It was Eulenberg in 1871 introduced the term 'progressive facial hemi atrophy' . This rare condition is characterized by progressive atrophy of the skin as well as subcutaneous tissue, muscle, cartilage, or bone of face and can leave a marked deformity[2,3]. In advanced forms of the

disease, the affected areas can become hyperpigmented, with patches of vitiligo[4]. This process can make the skin thin, dry, and atrophied[5]. Different case reports have also described extension of the disease process to the ipsilateral upper extremity and even to the entire half of the body [3]. This illness brings several functional and psychological problems, of which most concerning is when "symmetric" face loses its identity and creates aesthetic trouble for the patient. The incidence and cause of this condition is yet to be known[6]. Different mechanisms have been proposed describing pathogenesis of the disease, of which one important is cerebral disturbance on fat metabolism [7,8]. Secondary causes such as viral infections, endocrine disturbances, local trauma, autoimmunity and heredity conditions are believed to be associated to the pathogenesis of the disease [9,10,11,12]. This is a disease of early period of life and patients usually presents during the first two decades of life, most of the time it starts in the paramedian part of the face. It is reported that the condition may cease at any stage and leave only a minimal disfigurement on face. This syndrome has been reported to have higher incidence in women [2,7,11]. The extension of the atrophy is frequently limited to on one side of the face, and the ipsilateral involvement of body is rare but has been described [3]. 5% to 10% of cases were described as being bilateral [10].

Parry-Romberg Syndrome/progressive hemifacial atrophy is an auto-limitable condition and has some differencials such as localised scleroderma, traumatic fat necrosis, hemifacial microsomia, atrophy secondary to facial paralysis and unilateral patrial lipodystrophy[13].

Case Report:

In 2018 a 37 year old man presented to department of Neurology, BSMMU (Bangabandhu Sheikh Mujib Medical University), Shahbag, Dhaka, Bangladesh with history of progressive wasting of left side of his face since childhood. Patient stated that he noticed mild wasting of his left side of face which was slowly progressive in nature but there was no history of trauma, itching, discoloration, pain, fever, joint stiffness or any systemic illness. On enquiry he denies any weakness of face, there was no history of difficulty of chewing.

After his history taking meticulous physical examinations was done. His general vital signs were normal (Blood pressure 110/70 mm Hg with no postural drop, Pulse: 82/min, regular). On examination of the face there was wasting of left side of the face which extended from cranium to lower jaw. Abnormal pigmentation was seen on involved side of face but tenderness, loss of sensation was not found. There was no proptosis of left eye and power of facial muscles was normal on both sides. There was no malocclusion of teeth or change in voice. Thorough physical examination revealed no wasting or fasciculation in any part of body and other neurological, musculoskeletal, cardiological findings were within normal limit.

On laboratory investigations he had normal complete blood count with normal ESR. A MRI of brain with contrast was done which revealed nothing significant. Findings of X-ray chest P/A view, serum electrolytes including Calcium was within normal limit.

Patient was later properly counseled about the condition and was referred to Department of Maxilofacial surgery and Plastic surgery. He was prescribed with multivitamins.

Patient was reviewed 3 months later and his symptoms were static.

Indian Journal of Basic and Applied Medical Research; March 2020: Vol.-9, Issue- 2. P. 182 - 186
DOI: 10.36848/IJBAMR/2020/12210.51285

A

B

C

Discussion

Progressive hemifacial atrophy, as described in the case above, is a rare pathology, of unknown cause. It is characterized by a slow and progressive unilateral atrophy of the facial tissues, including muscles, bones and skin [2,3]. The condition is more often found in female population and has predilection for the left side of the face[14] which was seen in case of this patient.

Clinically, the skin can be dry, thin and with a dark pigmentation. Some patients present a demarcation line between normal and abnormal skin, resembling a big linear scar, known as coup de sabre (French term which means "cut of the sword")[3,6]. This finding was not noticed in this case reported. Ocular involvement is common, and the most frequent manifestation is the enophthalmy, due to fat loss around the orbit[15]. The extension of atrophy is frequently limited to on one side of the face, and the

ipsilateral involvement of body is rare (23 to 10% of cases were described as being bilateral)[10]. In the case presented here, there was involvement of only one side of the face, important features of this disease are enophthalmos, the deviation of mouth and nose to the affected side, and the unilateral exposition of teeth (when lips are involved) [11], which were not found in our case. Radiographically, teeth of affected side can present some deficiency in root development and, consequently, delayed eruption. However, the affected teeth are normal and vital clinically. The intraoral soft tissue and muscles of mastications are usually normal, and there is no difficulty in jaw movement, speech or deglutition limitations [16]. The dental findings may include delayed ipsilateral tooth eruption, atrophic root development, missing teeth, retarded root formation, oligodontia, microdontia, dilacerations, unilateral crossbite, and pulp stones on the affected side[17]. No such findings were manifested in our patient on clinical observation.

Treatment in progressive hemifacial atrophy is primarily directed towards augmentation of the affected areas for aesthetic reasons. Secondary procedures on uninvolved areas of the face may improve the result. The introduction of free tissue transfer by microvascular anastomosis opened a new era in reconstructive surgery[18].

Conclusions

Parry-Romberg syndrome is a rare condition manifesting as atrophy of one side of the face. In most cases, the cause of this syndrome appears to be unknown. The pathophysiology of the condition still remains unknown. Treating this condition can be frustrating for both physician and patient as there is no established treatment to limit or improve the condition.

ACKNOWLEDGMENTS AND DISCLOSURE STATEMENTS

The authors report no conflicts of interest related to this study.

Reference

1. Parry CH. Collection from unpublished papers, London: Underwood, 1825; 1: 478.
2. Jurkiewicz MJ, Nahai F. The use of free revascularized grafts in the amelioration of hemifacial atrophy. Plast Reconstr Surg 1985;76:44-55.
3. Lakhani PK, David TJ. Progressive hemifacial atrophy with scleroderma and ipsilateral limb wasting (Parry Romberg Syndrome). Journal of the Royal Society of Medicine. 1984;(2):138-9.
4. O'Flynn S, Kinirons M. Parry-Romberg syndrome: A report of the dental fi ndings in a child followed up for 9 years. Int J Paediatr Dent 2006;16:297-301.
5. Pinheiro TP, Silva CC, Silveira CS, Botelho PC, Pinheiro Md, Pinheiro Jde J. Progressive hemifacial atrophy — Case report. Med Oral Patol Oral Cir Bucal 2006;11:E112-4.
6. Witkop CJ. Genetic disease of the oral cavity: In Teiecke RW editor. New York, Mcgrant Hill 1965.
7. Finesilver B, Rosow HN. Total hemiatrophy. JAMA 1938;5:366-8.
8. Foster TD. The effects of hemifacial atrophy of dental growth. Br Dent J 1979;146:148-50.
9. Mazzeo N, Fisher JG, Mayer MH, Mathieu GP, Mcade FGG. Progressive hemifacial atrophy (Parry Romberg Syndrome). Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1995;79:30-5.
10. Miller MT, Sloane H, Goldberg MF, Grisolano J, Frenkel M, Mafee MF. Progressive hemifacial atrophy (Parry Romberg Disease). J Pediatr Oftamol Strabismus 1987;24:27-36

11. Pensler JM, Murphy GF, Muliken JB. Clinical and ultra-structural studies of Romberg´s hemifacial atrophy. Plast Reconstr Surg 1990;85:66976.
12. Zafarulla MY. Progressive hemifacial atrophy: a case report. Br J Ophthalmol 1985;69:545-7.
13. Rees TD. Facial atrophy. Clin PlastSurg 1976; 3: 637-46.
14. 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.
15. Lazaridou E, Giannopoulou C, Apalla Z, Fotiadou C, Trigoni A, Ioannides D. Parry-Romberg syndrome. J Dermatol Case Rep 2010;4:30-2.
16. Goldhammer KJ, Tadmor T, Braham J, Leventon G. Progressive hemifacial atrophy (Parry-Romberg's disease) principally involving bone. J Laryngol Otol 1981; 95: 643-7.
17. Sujatha SR, Devaraju D, Kunal A. Parry Romberg syndrome: Report of two cases with rare dental and skeletal fi ndings. J Dent 2012;2:212-5.
18. Harashina T, Fujino T. Reconstruction of Romberg's disease with free groin flap. Ann Plast Surg 1981; 7: 289-94.

Date of Submission: 19 January 2020
Date of Peer Review: 24 February 2020
Date of Acceptance: 18 March 2020
Date of Publishing: 30 March 2020
Author Declaration: Source of support: Nil , Conflict of interest: Nil
Ethics Committee Approval obtained for this study? NA
Was informed consent obtained from the subjects involved in the study? Yes
For any images presented appropriate consent has been obtained from the subjects: NA
Plagiarism Checked: Urkund Software

DOI: 10.36848/IJBAMR/2020/12210.51285