Journal of Pediatric Sciences

Progressive facial hemiatrophy with contralateral maxillary mucocele

Mohd Ashraf, Sheikh Javeed Sultan, Javed Ahmad

Journal of Pediatric Sciences 2010;2:e11

How to cite this article:

Ashraf M., Sultan S.J., Ahmad J. Progressive facial hemiatrophy with contralateral maxillary

mucocele. Journal of Pediatric Sciences. 2010;2:e11

CASE REPORT

Progressive facial hemiatrophy with contralateral maxillary mucocele

Mohd Ashraf¹, Sheikh Javeed Sultan², Javed Ahmad³

Abstract:

We report an 8-year-old boy with a six month history of unilateral, progressive facial atrophy, heralded by the development of a morpheaform plaque on the left cheek. The clinical features and investigations were consistent with a diagnosis of progressive hemifacial atrophy with contralateral maxillary mucocele. This case highlights the substantial overlap of progressive facial hemiatrophy with morphea, masqueraded by contralateral maxillary mucocele.

Keywords: Hemifacial atrophy, Morphea, Mucocele, Parry Romberg syndrome *Received:* 11/03/2010; *Accepted:* 03/04/2010

Introduction

Hemifacial atrophy, originally described by Parry and Henoch Romberg, [1,2] 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, and muscle [3]. Associated abnormalities with their frequencies include: hemiatrophy of contralateral or ipsilateral arm, trunk, or leg (20%), atrophy of tongue (25%), abnormalities (50%). trismus/jaw dental symptoms (including hemi-masticatory spasm) (35%), migraine/facial pain (45%), epilepsy (10%), sometimes associated with ipsilateral brain changes on MRI (5%), vitiligo, hair depigmentation/hyperpigmentation(20%) [4]. Association with right sided maxillary mucocele added to the facial asymmetry in our patient. The condition is rare and can have serious

Mohd Ashraf¹, Sheikh Javeed Sultan², Javed Ahmad³ ¹Department of Pediatrics, Sheri-i-Kashmir Institute of Medical Sciences (SKIMS) Medical College, Srinagar, India. ²Department of Dermatology, Sheri-i-Kashmir Institute of Medical Sciences (SKIMS) Medical College, Srinagar, India. ³Department of PSM, Sheri-i-Kashmir Institute of Medical Sciences (SKIMS) Medical College, Srinagar, India. Corresponding Author: Mohd Ashraf, MD 8-Green Lane Raj Bagh, Near Al-Farooq Masjid, Srinagar, India *e-mail:* aashraf_05@yahoo.co.in

mental and social implications on the patient as well as the parents.

Case Report

8-year-old An boy born out of nonand consanguinous parentage uneventful pregnancy was brought with complaints of facial asymmetry. Six months back, an indurated erythematous plaque appeared on his left upper cheek. The plaque was associated with slight pain. There was no history of trauma, pruritus, respiratory tract infection, dental infection, mouth ulceration or scaling. There was no improvement with oral antibiotics for 10 days. After third week, indurated erythematous plaque resolved leaving behind hyperpigmentation. Over the next four months, the left side of his face developed progressive atrophy that started from the upper part of left cheek and later extended to lips, chin and mandibular area on the same side and with loss lower lid eye lashes (Figure 1).

There was no ocular, respiratory tract, or other neurologic system involvement nor any history suggestive of delay in developmental milestones. Systemic examination was unremarkable. Local examination revealed an asymmetric appearing face with subcutaneous atrophy extending from the left lower eyelid to the ipsilateral temple, cheek, lips, chin, mandibular angle with loss of left lower lid eye lashes. There was no tenderness over the maxillary, ethamoid or frontal sinuses. Brownish hyperpigmented macule was present, extending from ipsilateral mandibular angle to the temple. Complete blood count, erythrocyte sedimentation rate, liver function tests, kidney function tests, chest X-ray, MRI brain and ECG were within normal limits. Antinuclear antibodies were absent. Histopathological examination biopsy of specimen from left cheek revealed normal epidermis with homogenization of collagen bundles in the dermis and decrease in the number of appendages. Computerized tomography (CT) scan of face revealed



Figure 1. showing left facial hemiatrophy with lower eye lash scarcity/ absence, and morpheaform plaque

the atrophic process extending upto the bone with mild left maxillary sinus haziness and a big mucocele producing bulge on right outer side (Figure 2).



Figure 2. CT face showing right maxillary sinus mucocele, and left sided maxillary haziness

Taking the clinical features and investigations into consideration a diagnosis of progressive hemifacial atrophy following plaque type morphea was made, with right expanded maxillary mucocele.

Discussion

Progressive hemifacial atrophy (Parry-Romberg syndrome) usually occurs in first two decades of life with a female-to-male ratio of 2:1. The relationship between Parry Romberg syndrome where subcutaneous fat and muscle and sometimes bone atrophy is the primary process and the morphea where dermal sclerosis is the primary process has been a subject of debate in the medical literature. Our patient was an 8-yearold boy in whom only facial atrophy was present clinically; there was history suggestive of plaque type morphea which was confirmed histopathological examination. by The atrophic process involved subcutaneous fat, muscle and bone with loss of lower lid eyelashes on the same side and there was marked contralateral maxillary sinus haziness, in the form of big mucocele and mild haziness in ipsilateral maxillary sinus, but without any history of respiratory tract infection. About onehalf of Parry-Romberg syndrome patients can have coexistent en coup de sabre morphea involving the paramedian forehead and scalp; both conditions can involve the muscle, bone, and even meninges and brain underlying affected areas, with seizures and other neurologic manifestations occurring in 10 to 20 percent of patients with either disorder [4-6]. No history suggestive of any ocular symptoms, respiratory tract infections symptoms, seizures or any other neurological manifestation was present in our patient nor was there any abnormality detected on CT/MRI scan of brain. Although dental abnormalities are well recognized fact in parry Romberg syndrome, but their sustained delay in eruption can lead to root resorptions in regions affected by this condition [7,8]. Pharmacologic therapies for Parry-Romberg syndrome have largely been the same as regimens used for the

treatment of morphea [9]. For individuals with active en coup de sabre morphea, methotrexate with or without monthly pulses of systemic glucocorticoids is commonly utilized [7].

Our case highlights coexistent contralateral expanded mucocele, which can masquerade the Parry Romberg syndrome, and will lead to false reassurance to the care givers. 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. To improve the cosmetic appearance, our patient is planned for hyaluronic acid filler injections and autologous fat transfer procedures along with proper otolaryngological management.

References

1. Parry CH. Collections from the unpublished medical writings of the late Caleb Hillier Parry. Vol. I. London: Underwoods (pub.) 1825. Pp. 478 (s)

2. Henoch E, Romberg HM. Klinische Ergebnisse. Berlin: A. Forstner (pub.) 1846. Pp. 75-81. (s)

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. Jon Stone. Parry Romberg Syndrome. Practical Neurology. 2006;6;185-1881.

5. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. J Am Acad Dermatol 2007; 56: 257-263

6. Orozco-Covarrubias L, et al. Scleroderma Ôen coup de sabreÕ and progressive facial hemiatrophy: is it possible to differentiate them? J Eur Acad Dermatol Venereol. 2002; 16:361-366.

7. Foster TD. The effects of hemifacial atrophy on dental growth. Brit.dent J. 1979; 146:148-150

8. O'Flynn S, Kinirons M. Parry-Romberg syndrome: a report of the dental findings in a child followed up for 9 years. Int J Paediatr Dent. 2006; 16: 297-301.

9. Korkmaz C, Adapinar B, Uysal S. Beneficial effect of immunosuppressive drugs on Parry-Romberg syndrome:a case report and review of the literature. South Med J. 2005;98 : 940-942