

Journal of Pediatric Sciences

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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

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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%), dental abnormalities (50%), trismus/jaw 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

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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-year-old boy in whom only facial atrophy was present clinically; there was history suggestive of plaque type morphea which was confirmed by histopathological examination. 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 one-half 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.

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