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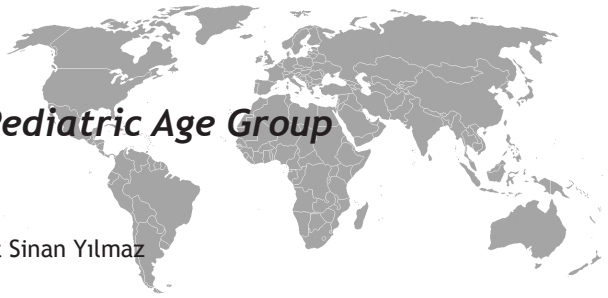
Case Report



Melkersson-Rosenthal Syndrome in Pediatric Age Group

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ABSTRACT

Melkersson-Rosenthal Syndrome (MRS) is a disease characterized by peripheral facial paralysis, facial edema and fissured tongue. Appearance of this triad is rare, generally monosymptomatic or oligosymptomatic involvement is observed and difficulties or delays in diagnosis can occur due to scarcity of the disease. One or two of the findings and presence of cheilitis granulomatosa in biopsy are sufficient for the diagnosis. Melkersson Rosenthal Syndrome should be considered in differential diagnosis of recurrent facial paralysis. Although findings of the syndrome can regress spontaneously or with medical treatment, it can take a progressive course in some patients, requiring surgical treatment (facial nerve decompression). In this article a pediatric case with diagnosis of Melkersson Rosenthal Syndrome is presented.

Key words: *Melkersson-Rosenthal syndrome, orofacial edema, recurrent facial paralysis.*

Pediatrik Yaş Gurubunda Melkersson-Rosenthal Sendromu

ÖZET

Melkersson-Rosenthal sendromu (MRS); periferik fasiyal paralizi, fasiyal ödem ve cografik dil triadı ile karakterize bir hastalıktır. Hastalık yüz, dudaklar, oral kavite ve fasiyal sinirde ödeme yol açan granülomatöz patoloji ile seyreder. Sendromun bulgularının kendiliğinden veya medikal tedaviyle gerileyebilmesine karşın, bazı hastalarda progresif seyredebilmekte ve cerrahi tedavi (fasiyal sinir dekompresyonu) gerektirmektedir. Klasik belirtiler triadının görülmesi nadirdir ve genellikle monosemptomatik veya oligo semtomatik tutulum izlenir ayrıca hastalığın nadir görülmesinden dolayı tanı koymada güçlükler veya gecikmeler söz konusu olmaktadır. Bulgulardan bir veya ikisi ile biyopside granülamotöz keilitin varlığı kesin tanı için yeterlidir. Melkerson Rosenthal sendromu tekrarlayan fasiyal paralizilerin ayırıcı tanısında düşünülmesi gereken bir hastalıklardandır ve bu yazıda Melkersson Rosenthal sendromu tanısı konulan bir pediatrik hasta sunulmuştur.

Anahtar kelimeler: *Melkersson Rosenthal Sendromu, orofasiyal ödem, tekrarlayan fasiyal paralizi*

INTRODUCTION

Melkersson syndrome was first described with findings of peripheral facial paralysis and orofacial edema in 1928. After addition of fissure in tongue finding by Rosenthal in 1930, it was identified as Melkersson Rosenthal Syndrome (MRS) (1). Diagnosis of MRS is made with clinical findings (1-3). Classical triad of the disease is characterized by peripheral facial paralysis, non-pitting orofacial edema and fissured tongue (scrotal tongue, lingua plicata). These findings are seen in 18-70% of the patients whereas oligo (with two symptoms) or monosymptomatic (with single symptom) forms are encountered more frequently (1,4-8). Most common symptom is orofacial edema, occurring in 80-100% of the cases (9). Fissured tongue is seen in 30-40% of the cases (4,9,10). In patients with none of the three findings, diagnosis can be made with mucosal biopsy and these patients are identified as oligo or monosymptomatic form (4, 6-8).Although etiology of MRS is not well known, several factors such as Herpes simplex infections, Epstein-Barr virus, Cytomegalovirus, Campylobacter jejuni, Varicella zoster, odontogenic

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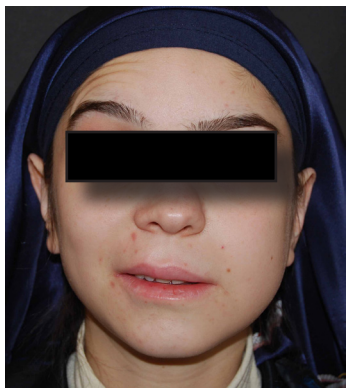

Figure 1.

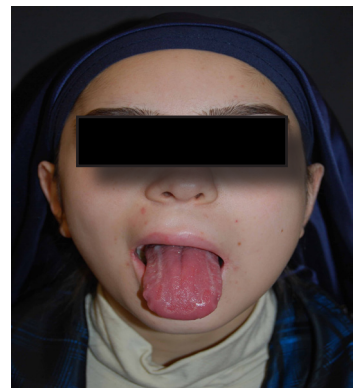

Figure 3.

infections, adenotonsillitis, tuberculosis and T lymphocyte malfunction were held responsible (11). This syndrome, seen rarely in childhood, is more common in 2nd and 3rd decades of life. It is believed to be inherited by autosomal dominant trait (12). In pathologic analysis of samples taken from edematous mucosae of the patients with MRS, non-caseified granulomas, Langerhans-like giant cells, mononuclear infiltrations, nonspecific inflammation and fibrosis are seen. These histopathological findings support the diagnosis. However, when pathology does not support the diagnosis, disease is not necessarily excluded (1). MRS is diagnosed mainly on clinical basis. There is no consensus about treatment protocols in MRS. Treatment options can be either medical or surgical. Among the medical treatment options, corticosteroids, immunosuppressive agents, antibiotics, antihistaminics, danazol, hydrochlorine and clofazimine take place. In situations where no response to medical treatment is obtained, facial nerve decompression can be considered as surgical treatment option (1-3). In this study, clinical findings and treatment of a case with MRS was discussed and it was pointed out that this syndrome, seen rather uncommonly in pediatric age group, should be taken into account in differential diagnosis of facial paralyses.

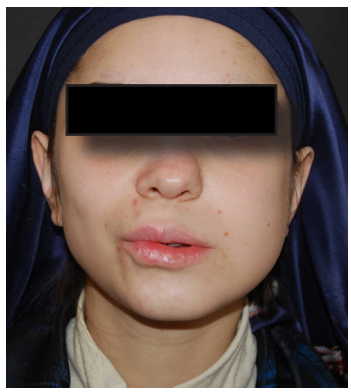

Figure 2.

CASE

Seventeen-year-old female patient admitted with complaints of inability to close the left eyelid, drooping of the mouth corner and swelling in the face. Her history revealed that complaints were present for 5 days, however she had the diagnosis of Bell's palsy in 5 different occasions (3 times right, twice left side). She had her first attack at 10 years of age and right side of her face was involved. There was no one with similar complaints in her family. She had no history of trauma, fever or known systemic disease. She had received 1mg/kg prednisolone for 20 days for the Bell's palsy and she had recovered completely without a sequel. Her last physical examination revealed no pathologic finding except for facial paralysis, non-pitting edema in left side of the face and lips, edema and fissures in the tongue. She had stage 6 facial paralysis according to House-Brackmann classification on left side. (Figure 1,2,3) Other cranial system and neurologic examinations were normal. Laboratory analysis revealed a normal complete blood count, serum biochemistry, sedimentation, CRP, ASO, and immunoglobulin levels. Serologic analysis of Herpes simplex virus, cytomegalovirus, Epstein-Barr virus, coxsackievirus, human immunodeficiency virus, parvovirus infections were done and anti CMV-IGG, herpes simplex

type 1 IGG were found to be positive.

Autoimmune disease panel was negative. Chest X ray, PPD test and audiogram were normal. Temporal and cranial magnetic resonance imaging did not yield any pathologic finding. MRS was suspected and a biopsy was done from edematous area in the upper lip. Non specific inflammatory involvement was found in histopathologic examination. MRS diagnosis was made as a result of clinical and pathologic findings and glucocorticoid (1mg/kg methylprednisolone) treatment was initiated. Following treatment, peripheral facial paralysis of the patient was regressed within 3 weeks. Patient was followed regularly every 3 months and symptoms did not recur in 6th month.

DISCUSSION

MRS is a syndrome described with classical triad of recurrent orofacial edema, recurrent peripheral facial paralysis and fissured tongue (lingua plicata). Since MRS can manifest as various clinical pictures, the diagnosis can be challenging in some cases. Many cases with MRS can be missed when all symptoms do not coexist. Facial paralysis and orofacial edema are more common findings in MRS. Facial paralysis usually coexists with edema, however it may develop before or after the edema. Fissured tongue is more uncommon finding (it is seen in only 40% of the cases) and 2/3 anterior region of the tongue is affected. Fissured tongue is not always specific for diagnosis; it can be seen in healthy population as well (4,12,16). MRS is usually presented in oligosymptomatic form and many patients can be overlooked and misdiagnosed as Bell's palsy. MRS diagnosis is made clinically. Imaging methods such as computerized tomography (CT) and magnetic resonance (MR) are used for excluding the other diseases in differential diagnosis (17). All three symptoms corresponding classical triad was present in our case. Although diagnosis was certain, imaging techniques were performed in order to exclude the other diseases. Facial paralysis can recur in 3-11% of the cases (13). Causes of recurrent peripheral facial paralysis include Bell's palsy, MRS, infectious mononucleosis, syphilis, herpes zoster, otitis media, multiple sclerosis, diabetes mellitus, leukemia, Myasthenia Gravis, Guillan-Barre syndrome, polyarteritis nodosa and tumors (4). Positive findings regarding these disease in our patient were as follows: anti CMV-IGG, herpes simplex type 1 IGG positive.

This syndrome is rare in children, it usually occurs in young adults, in 2nd and 3rd decades. Only 30 patients in childhood age group were defined in the literature. It is more common in girls than boys; male/female ratio is 1/3 (4,14). Facial paralysis is generally unilateral in MRS, however bilateral involvement has been also reported. It can resolve spontaneously or can be permanent (18). In MRS, vagus nerve, glossopharyngeal nerve and hypoglossal nerve paralysis can accompany as well (19,20). Etiology of MRS is not fully known. Bacterial and viral infections, hypersensitivity against some food additives such as monosodium glutamate and heavy metals, stress, autoimmunity and genetic factors were blamed (9,10). In some studies, it was reported that MRS was transmitted in autosomal dominant inheritance and responsible gene was localized in short arm of the 9th chromosome (21). In histopathologic examination of the edematous region, observation of non-caseating granulomas and high level of angiotensin converting enzyme in these cases suggested that MRS could be a variant of sarcoidosis and Crohn disease. MRS has similar clinical and histopathologic characteristics with Miescher's cheilitis (oligosymptomatic form of MRS), Crohn's disease, sarcoidosis and orofacial granulomatosis. Some researchers consider orofacial granulomatosis as an initial finding in Crohn's disease, while others consider orally localized Chron's disease as oligosymptomatic form of MRS. Therefore patients with MRS diagnosis should be followed in terms of development of Crohn's disease and sarcoidosis (22,23). Besides, among the diseases that can accompany MRS are inflammatory bowel disease, syphilis, craniopharyngioma, otosclerosis, trigeminal neuralgia, migraine-type headache and psychosis. In differential diagnosis, angioedema, sarcoidosis, hypothyroidism, superior vena cava syndrome, recurrent lymphangioma, lymphoma, chronic herpes simplex labialis, Crohn's disease should be considered (15,16,20). As a result of clinical and laboratory examinations, these disorders were not encountered in our case.

Although systemic (1mg/kg/day) corticosteroid treatment is frequently applied in medical treatment of MRS, there is no standard treatment protocol. It was reported that corticosteroids prevent edema and tissue damage. Glucocorticoid administration into lesion was found to be as efficient as systemic glucocorticoid treatment. Other medical treatment options include antihistaminics, immunosuppressive agents, antibiotics, hydrocholrine and danazol (1). We administered 1mg/kg methyl-

prednisolone for two weeks in our case and almost total recovery was achieved in patient's complaints following the treatment.

MRS, a rare disease, should be considered in differential diagnosis of recurrent facial paralysis. Its classical triad is peripheral facial paralysis, non-pitting orofacial edema and fissured tongue. Mono/oligosymptomatic forms which are difficult to diagnose can be encountered. In uncertain situations, diagnosis should be supported by histopathologic findings. Although it is more uncommon in children, diagnosis and treatment is same with that of adults.

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