

MELKERSSON-ROSENTAL SYNDROME IN CHILDREN AS AN ORPHAN DISEASE: PREVALENCE, CLINICS AND TREATMENT METHODS

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Abstract.

In this article, the authors, referring to their own practice, cases from practice, present information about a rare syndrome in children - Melkersson-Rosenthal. Based on the results of their own clinical observation, instrumental and laboratory studies, the clinical and diagnostic criteria of the syndrome are described. Historical and modern literary data on the rare Melkersson-Rosenthal syndrome are provided. The authors argue that the disease begins during the transition from childhood to adolescence, and after treatment, the progression of the disease slows down, but does not stop, which requires close monitoring by both pediatricians and general practitioners.

Key words: Melkersson-Rosenthal syndrome, clinical picture, treatment, macrocheilitis, granulomatous cheilitis.

Melkersson – Rozental sindromi (Melkersson – Rosenthal syndrome, MRS) (HKK (МКБ) -10 – G51.2) uchta simptomlarning (lab va/yoki yuzni shishi, yuz nervini paralichi (falaji), skrotal til) yig'indisi sifatida kuzatiladigan kasallik hisoblanadi va bu haqida ko'plab adabiyotlarda ma'lumotlar keltiriladi [1-20].

James, et. al. (2006) MRSni «Misher-Melkerson-Rozental sindromi» deb nomlashgan va kam uchraydigan nevrologik buzilish deb ta'rif berishdi [8]. Kasallik bolalikda yoki erta o'spirinlik yoshida boshlanadi. Periodik qaytalanishlardan keyin (ular orasidagi muddat bir necha kundan bir necha yilgacha bo'lishi mumkin) shish saqlanib qolishi va kattalashishi mumkin va oxir oqibat doimiy bo'lib qoladi. Lablar qizilsimon-jigarrangli qattiq va yorilib ketgan ko'rinishda bo'lishi mumkin. Kasallikni sababini ular genetik moyillik bilan bog'laydilar. Masalan, bir qator tadqiqotchilar bu kasallikni Boliviyaning ba'zi bir etnik guruhlari orasida tarqalgan deb hisoblashadi. U Kron kasalligini yoki sarkaidozni simptomi bo'lishi mumkin deb fikr bildirishadi [10].

Bu simptomlar triadasi birinchi marta rus nevrologi G.I. Rossolimo tomonidan 1901 yili yozilgan. Keyinchalik shved shifokori Ernst Gustav Melkersson 1928 yili 35 yoshlik ayolda makroxeyliya va yuz nervini bir tomonlama falajini diagnostika qiladi [10]. 1931 yili nemis nevrologi Gart Rozental yuz nervini falaji va makroxeyliya bilan birga kuzatilgan buklangan tilni 5 nafar bemorlarda kuzatganini tariflaydi [3, 13]. Shundan beri yuqorida keltirilgan simptomlar triadasi "Melkersson-Rozental sindromi" deb nom oldi va alohida nazologik shaklga ajratildi. Shuni ta'kidlash kerakki, asosan sindromni monosimptom varianti, faqat makroxeyliya ko'rinishidagi klinik namoyon bo'lishi kuzatiladi. Ba'zi mualliflar Misherni granulematoz xeylitini MRSni monosimptom varianti deb hisoblashadi [4].

Melkersson – Rozental sindromini kelib chiqish sabablari to'lig'icha aniqlanmagan. MRSni patogenezi haqida ko'plab tahminlar, farazlar mavjud. Ularning asosiylari quyidagilar hisoblanadi: allergik, angionevrotik, infeksiyon, genetik va immunologik disfunktsiyalar [2].

Kasallikning populyacion chastotasi o'rtacha 0,08% ni tashkil qiladi [19]. MRS har qanday yoshda namoyon bo'lishi mumkin, ammo ko'p holatlarda 25-40 yoshlar oralig'ida ayollarda kuzatiladi. Bolalarda MRS kam aniqlanadi va ko'pincha 7 yoshdan 12 yoshgacha oralig'ida kuzatiladi, eng kichkina bemor bola 22 oylik yoshda bo'lganligi olimlar tomonidan yozib o'tilgan [17].

Kasallik odatda qo'qqisdan boshlanadi va bir necha soat oralig'ida bitta yoki ikkala lablarning qizil hoshiyasi va ularning ostidagi to'qimalar shishib ketadi (birinchi simptomi). Lablarning shakllanmagan shishi hosil bo'ladi, ko'pincha ularning qirg'oqlari xartum singari ag'dariladi yoki odatda tishlardan ortda turadi (tapira tushmug'i, morda tapira). Ba'zida shish shunchalik kattalashadiki, bunda lablar 3-4 marta yiriklashgani kuzatiladi. Lablarning qalinlashishi notekis, labning bir tomoni odatda biroz ko'proq shishli bo'ladi. Ko'pincha bu bilan bir vaqtini o'zida lunj sohasining (pareit) va yuzning boshqa

uchastkalarini shishgani aniqlanadi, ammo shish xududida og'riq va yallig'lanish belgilari kuzatilmaydi. Lablarning shishi yoriqlar hosil bo'lishi bilan kechadi va qoidaga ko'ra labnig barcha qismini egallab oladi. Bunda gapirish, mimika, ovqat qabul qilish buzilishi mumkin. Shishgan lablar oqish-qizil rangda, ba'zida turg'un ko'kimtir-pushti tusli bo'ladi. Palpatsiyada bir maromdagi yumshoqlik yoki zich elastik konsistentsiyali to'qima seziladi. Bir qator bemorlarda shishlar doimiy mavjud bo'lib, asta-sekinlik bilan kuchayib boradi, vaqti-vaqti bilan biroz kamayib turadi. Ba'zi bir bemorlarda shishlar yo'qolib ketadi, ammo keyinchalik turli xil chastotada qaytalanib turadi, so'ng esa doimiy qoladi [1].

Bu kasallikning ikkinchi simptomi bo'lib, yuz nervini parezi/falaji hisoblanadi, undan avval ko'p hollarda turli xil davomiylikdagi prodromal hodisalar bilan namoyon bo'ladi. Bu hodisalar yuz nervini qisman paralich bo'lishiga olib kelishi mumkin, bunda vegetativ, sezuvchanlik hissiy va motor funksiyalarini fragmentar saqlanib qolishi kuzatiladi. Bemorlarning katta qismida vaqt o'tishi bilan yuz nervini parezi o'tib ketadi, ammo kechishida turg'un xarakterlikni kuzatish mumkin bo'ladi. Kam holatlarda kasallik yuz nervini paraze/falaji bilan boshlanishi mumkin, bir necha yillardan (1 yildan 10 yilgacha) keyin esa shishni rivojlanishi kuzatiladi [1].

Bu kasallik uchun xarakterli bo'lgan uchinchi simptom, bu skrotal yoki burmali tilni mavjudligidir. Bu simptom bemorlarni 2/3 qismida kuzatiladi va odatda tilni anomal rivojlanishi sifatida baholanadi. Bunda tilni notekis kattalanishiga olib keluvchi shish aniqlanadi. Uchchala simptomni birga kelishi patologik jarayonni torpid kechishidan, o'tkaziladigan davoga sezuvchanlik pastligidan dalolat beradi [1].

Klinik amaliyotda ko'pincha MRSni monosimptom variantini kuzatishimiz mumkin bo'ladi, bunda kasallikni klinik manzarasida teri rangini (oqish-qizildan to turg'un ko'kimtir-pushti rangda) o'zgarishi bilan kechuvchi yuzni pastki qismini terisining shishi, yuqori va/ yoki pastki labning qizil hoshiyasini shishi bilan xarakterlanadi [1].

Davolashda quyidagi preparatlar guruhi qo'llaniladi: tizimli glyukokortikosteroidlar, antibiotiklar, antigistamin preparatlar, immunosuppressiv vositalar. Ammo ko'plab holatlarda davolashdan samara bo'lmasligi kuzatiladi, ba'zan ba'zi preparatlarning jiddiy nojo'ya ta'sir qilgani aniqlanadi [1, 4].

Yuqorida keltirilgan preparatlarning bemor tomonidan qabul qilinishi to'xtatilganida kasallikning qaytalanishi kuzatiladi.

MRS bilan og'riqan bemorni klinik kuzatuv.

Bemor SH.L. 2018 y.t., "Melkersson-Rozental sindromi" diagnozi 2024 yili qo'yilgan. Peditrarga bemor qizni ota-onasi birinchi marta yuqori labini shishib ketishiga shikoyat qilib olib borishgan.

Bemorning otasini so'ziga ko'ra, 2024 yilda qizini yuzini o'ng tomonida og'riq paydo bo'lgan va uni fonida o'ng yuzi tomonidan ptoz, labini o'ng burchagida shish aniqlangan. Peditr va nevropatolog tavsiyasiga ko'ra kortikosteroidlar, qon tomirlari preparatlarini, vitaminlarni qisqa kurslarda olib davolangan. Shuningdek, fizioterapevtik usullar qo'llanilgan va shu mutaxassislarning nazoratida turgan.

O'tkazilgan davolash natijasida yuz nervi funksiyasi qisman tiklangan. Bemor qizni otasini so'zlariga ko'ra, bir necha oylardan keyin, kutilmaganda qizini yuqori labini shishi paydo bo'lgan va bu shish asta-sekinlik bilan yuqori labni to'liq egallagan. Bemorni otasi qizini turli xil vrach-mutaxassislarga olib borib ko'rsatgan. Allergolog, otolaringologlar konsultatsiyasini olishgan. Toshkent shahridagi ilmiy markazlarda ham vrachlar konsultatsiyasida bo'lishgan. Standart tekshiruvlardan so'ng MRS diagnozi qo'yilgan. Antigistamin preparatlar, sensibilizatsiyani oluvchi vositalar, topik kortikosteroidlar qo'llanilganiga qaramay, davodan samara past bo'lgan.

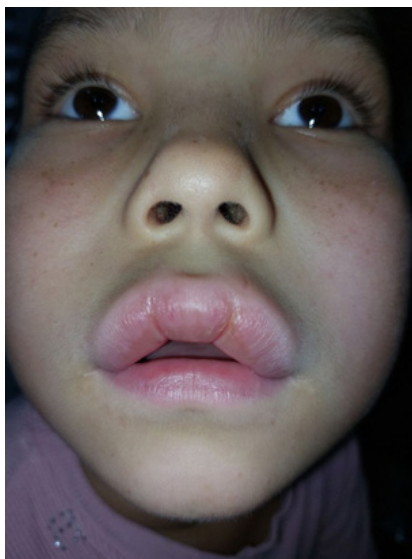
Bemor qiz vizual ko'rilganida, yuqori labini qizil hoshiyasida yaqqol shish namoyon bo'lgan. Palpatsiyada zich elastik, hamirsimon konsistentsiya aniqlanadi. Og'iz shilliq qavati ko'zdan kechirilganida: shilliq qavatlar oqish-pushti rangda, o'rtacha namlikda, patologik toshmalarsiz. Til normal konfiguratsiyaga ega, burmali (skrotal) tuzilish yaqqol namoyon bo'lmagan, biroz shishgan. Laborator tekshirishlarning (qonni klinik va biokimyoviy tahlili) natijalari referens ko'rsatkichlari darajasida (расим 1, 2, 3).

Klinik manzaraga asoslanib "MRS" diagnozi qo'yildi va kompleks terapiya tavsiya etildi: azitromicin 250 mg sutkasiga 10 kun davomida ichish, Kenalog 1 ml muskul orasiga 1 kun yuborish, Levosetil 20 ml 10 tomchidan 1 mahaldan suv bilan, natriy tiosulfat 5%-200 ml 1 osh qoshiqdan 3 mahaldan ichga qabul qilish, Xelzibon 1 tabletkadan 2 mahaldan ovqatdan oldin ichishga 1 oy. Keyinchalik Dotal 5 mg 1 tabletkadan 1 mahaldan 1 oy ichish maslahat berildi. Tashqi davoga labni qizil hoshiyasiga Momeyd kremi 1-2 mahaldan bir oy davomida surtishi tavsiya etildi.

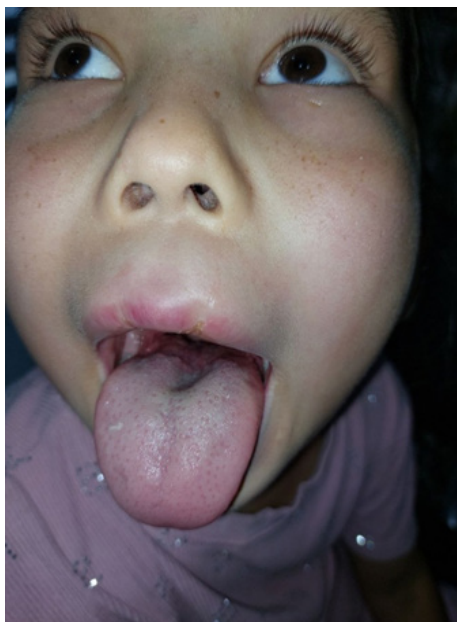
Qo'llanilgan terapiya fonida shishni sezilarli kamayishi va labning qizil xoshiyasini zichligini so'rilishi ko'rinishida ijobiy dinamika kuzatildi.

Melkersson-Rozental sindromini ilk marta tariflangan vaqtdan beri bu kasallik haqida juda ko'plab tadqiqotlar chop etilgan. Bunga asosan MRSning turli xil tarixiy atamaları ("Misherning granulematoz xeyliti", "orofacial granulematoz", "MRS") mavjud bo'lib, ular doimiy mavjud adabiyotlarda sinonimlari sifatida muntazam qo'llab kelinadi [1-20]. Hozirgi kunda MRS istisno tashxisi sifatida keladi, bu amaliy sog'liqni saqlash amaliyotida munozarali diagnozlarni qo'yish bilan birga keladi [6].

Biz keltirgan misolda ko'rinib turibdiki, kasallik bolaligidan o'spirinlikka o'tish davrida boshlanmoqda va kasallik rivojlanishi davodan keyin sustlashgan, ammo to'xtamagan. Bemorni tilida ham patologik o'zgarishlar boshlanganligi kuzatilmoqda. Kam uchraydigan bunday kasalliklar pediatrlar, stomatologlar hamda dermatologlarning amaliyotida kuzatilishi mumkinligini, va bu mutaxassislar MRS ni uchrab turishini unitmasliklari kerak.



Rasm-1. Yuqori labning shishi.



Rasm-2. Burmali (skrotal) til shakllanishi boshlangan holati.



Rasm-3. Burmalı (skrotal) til shakllanishi boshlangan holati.

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