Pringle-Bourneville syndrome associated with tinea capitis – a case report

Overview. Bourneville tuberous sclerosis (TS) is a genetic disease with autosomal-dominant transmission with high penetrance, but incomplete and variable clinical expression. Recent studies have shown a prevalence of 1: 5000 under the age of 5 years and 1: 25000 for all ages due to the possibilities of diagnostic performance (P. Curatolo, 2003). Onset of illness is generally in childhood, but the diagnosis after the age of 20 years is not unusual. Frequently, the first signs of cutaneous manifestations of the disease are, in particular, achromic or hypopigmented spots. Classic triad syndrome includes distinct clinical expressions – epilepsy, mental retardation and facial angiofibromas (Roach S.E., Sparagana S.P., 2004). There are two genes responsible for the syndrome. TSC1 gene is located on chromosome 9 (9q34) and protein encoding hamartina; TSC2 gene is located on chromosome 16 (16p13.3) and protein encoding tuberin. TSC2 gene is associated with more severe forms of the disease and would be responsible for 55% to 80-90% of the cases. Mutation of one of the two genes leads to loss of control of cell division and tumor formation. Tumors facial feature arrangement with symmetrical on cheekbones (aspect of butterfly wings) suggests the possible involvement as predisposing factor, sun exposure in skin tumor. In the early weeks of gestation mutant genes cause disruptions in the germ cells of the matrix, and subsequently forming giant cells neuroglial resulting in three changes: cortical tubers, subependymal nodules and defective myelination (Kwiatkowski D.J. et al., 2004). Clinical manifestations are characterized by a pronounced polymorphism and are in accordance with the age. The most common type of skin patches are achromic spots(98%), others as angiofibromas – in 54% cases, stains „cafe au lait" – in 38.5%, Shagreen's spots – in 36.5% and periunghiale fibromas – in 1.9% of cases.

A case report. We present the case of a male child, age 6, from rural areas, hospitalized with Tinea capitis and associated with Bourneville disease. Clinical: on the scalp were present multiple erythematosquamous plaques, round-oval, 0,5-2,0 cm in diameter, hair was broken at a high level, with a pale green fluorescence in Wood's lamp. Tinea capitis caused by the Microsporum canis was confirmed by direct microscopic examination of hair and cultures. Also, 4 hypopigmented spots of 1-2 cm in diameter and 2 hyperpigmented hamartoma-like papilloma, were present on the patient's abdomen, thigh, legs; eruption occurred when the patient was 7 years old. Angiofibromas on the patient`s face (forehead, nasolabial folds, cheeks and chin) have been appearing since he was a toddler. The patient suffers from epilepsy and mild mental retardation. Histopathological report has shown vascular and fibroblast proliferations. Magnetic resonance imaging of the skull reveals nonspecific gliosis unique outbreaks. Ophthalmic pathology was excluded.

Conclusion. This case was presented for discussion because association of tuberous sclerosis Bourneville with tinea capitis is a rare manifestation.

Key words: Bourneville tuberous sclerosis, Tinea capitis, child.

Lupus vulgar la copil – prezentare de caz

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Prezentare de caz. Prezentăm cazul unui copil de sex masculin, în vârstă de 6 ani, internat în Spitalul Dermatologie și Maladii Comunicabile pentru o erupție cutanată diseminată pe față și trunchi, fără acuze subiective. Procesul cutanat a debutat cu 3 ani în urmă, când pentru prima dată, la nivelul obrazului stâng, mama copilului a observat o placă netedă, moale, de culoare roșie-maronie. Anterior, cu 6 luni mai devreme, după o traumă, erupția a crescut în dimensiuni și s-a extins pe obrazul drept, nas, menton și pe trunchi. Examenul tegumentelor a evidențiat multiple plăci netede, de culoare roșie-maronie, consistență moale, cu atrofie în centru. Cartilajul nazal treptat s-a deformat. Inspecția generală a depistat o limfadenită submandibulară. Examenul histopatologic a evidențiat focare de necroză cazeoasă, înconjurate la periferie de inflamație granulomatoasă. Hemoleucograma a pus în evidență o leucocitoză marcată din contul polimorfonuclearelor. Biochimia sanguină, imunograma, analiza sumară de urină, precum și roentgenografia cutiei toracice au fost în limitele normei, dar în testul cutanat Mantoux, leziunea rezultantă a depășit 20 mm în diametru. Tratamentul standardizat antituberculos a dus la o regresare remarcabilă a procesului cutanat.

Concluzii. Lupusul vulgar reprezintă o provocare în diagnostic și determină o abordare multilaterală. Tabloul histopatologic specific, testul Mantoux pozitiv și absența afectării pulmonare sunt reperele caracteristice ale acestui caz.

Cuvinte-cheie: lupus vulgar, copil, prezentare de caz.

Lupus vulgaris in a child – a case report

A case report. A 6-year old boy has been admitted to the pediatric department of Hospital of Dermatology and Communicable Diseases with complaints of skin lesions disseminated on his face and trunk, without any subjective sensations. Parents presented a 3 year history of flat, soft and red color skin eruption, which has occurred initially on patient's face (left cheek). 6 months prior to hospitalization, after a trauma, eruption increased significantly in size and extended to the right side of the patient's face, nose, chin and trunk. Clinical inspection has revealed multiple flat, soft, red infiltrative plaques with central atrophy. Nasal cartilage has mutilated gradually. A submandibular lymphadenopathy was observed. Histopathologic exam has revealed foci of caseous necrosis surrounded by a peripheral granulomatous inflammation. In blood smear a marked leukocytosis due to polymorphonuclear cells was seen. Biochemistry of the blood, immune cell count, a routine urine test, as well as, X-ray examination of the patient's chest haven't shown any significant deviations, with the exception of Mantoux test in which diameter of the papule was more than 20 mm. Obvious antituberculous therapy, which the patient received, resulted in spectacular regression of the eruption.

Key words: lupus vulgaris, child, case report.