The Turkish Journal of Pediatrics 1999; 41: 545-549 Case

PSORIASIS IN A PATIENT WITH NEUROFIBROMATOSIS*

Solmaz Çelebi MD**, S. Şebnem Kılıç MD***, Mehmet Okan MD****

SUMMARY: Çelebi S, Kılıç SŞ, Okan M. (Department of Pediatrics, Uludağ University Faculty of Medicine, Bursa, Turkey). Psoriasis in a patient with neurofibromatosis. Turk J Pediatr 1999; 41: 545-549.

A seven-year-old boy with neurofibromatosis who developed scalp psoriasis is presented. The clinical evaluation of the patient revealed multiple café au lait spots, axillary freckling, bilateral Lisch nodules and a psoriatic plaque on his scalp. Though there is no known direct relationship between neurofibromatosis and psoriasis, one is suggested in this patient, possibly related to a common genetic defect. *Key words: neurofibromatosis, psoriasis.*

Neurofibromatosis is a multisystem disease that is transmitted in an autosomal dominant pattern and seen with a frequency of about 1:3,000[1]. Approximately 50 percent of cases represent new mutations. There are seven distinct forms of neurofibromatosis. Neurofibromatosis type 1 (NF-1) is the most prevalent. The abnormal gene of NF-1 is the band q11,2 on the proximal long arm of chromosome 17[2,3]. Psoriasis is a common chronic hyperproliferative inflammatory papulosquamous dermatosis of unknown etiology. It tends to occur in families, although genetic transmission has not been clearly delineated[4]. When the onset occurs during childhood, about 50 percent have a positive family history of the disease[5].

It is rare for patients to have both disorders; we could find only two previous reports with this combination in the literature[6,7]. In this paper we report a male patient with neurofibromatosis who developed scalp psoriasis.

Case Report

A seven-year-old boy was admitted to emergency service because of a partial seizure attack. He had multiple brown papules on his trunk and extremities, reportedly present since birth. He had some learning problems. There was no family history of neurofibromatosis, psoriasis or consanguinity. On physical examination there were multiple café au lait spots and axillary freckling (Figs. 1-2). He had a dysmorphic facial appearance and hypertelorism. Examination

* From the Department of Pediatrics, Uludağ University Faculty of Medicine, Bursa.

** Research Assistant in Pediatrics, Uludağ University Faculty of Medicine.

*** Pediatrician and Fellow in Pediatric Immunology, Hacettepe University Faculty of Medicine, Ankara.

**** Associate Professor of Pediatrics, Uludağ University Faculty of Medicine.

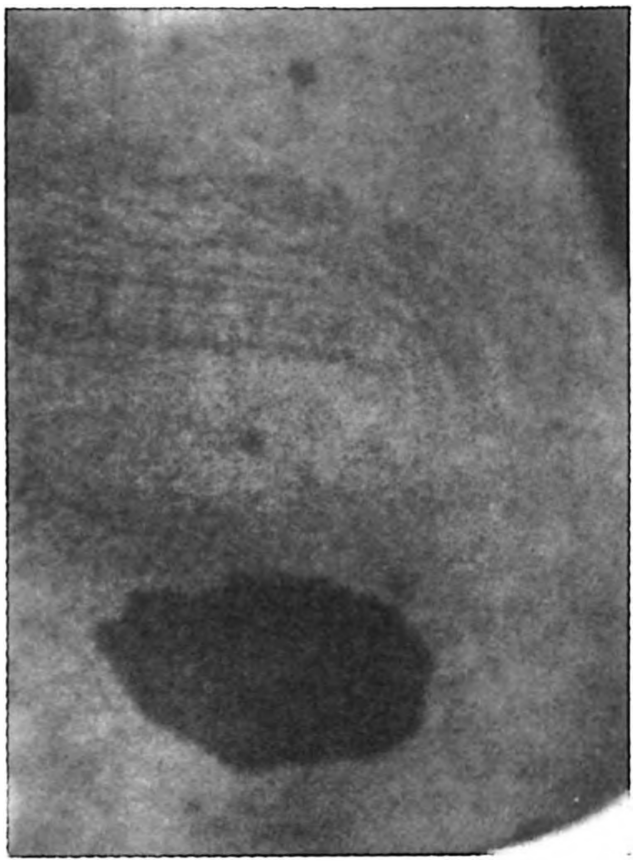

Fig. 1: Multiple café au lait macules in the patient with neurofibromatosis type 1.

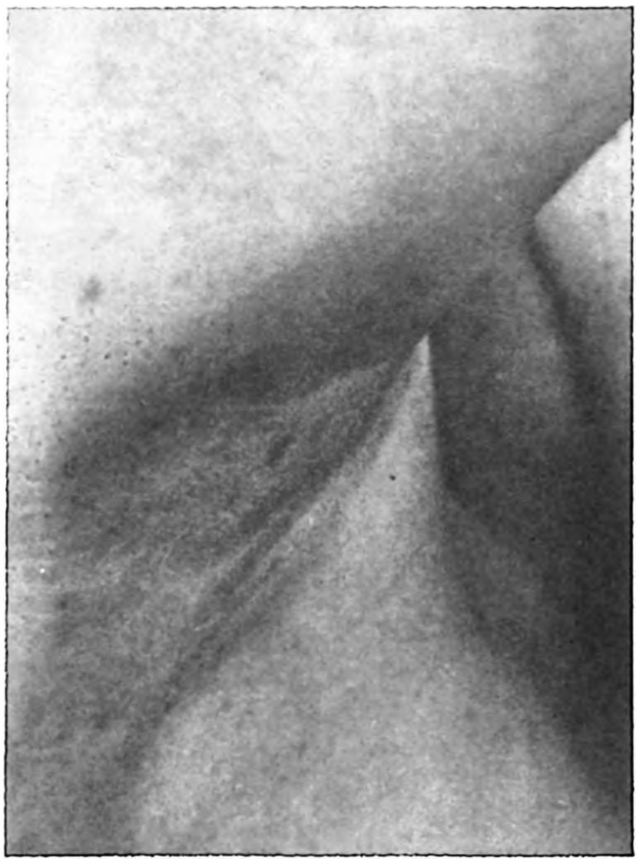

Fig. 2: Axillary freckling in the patient with neurofibromatosis type 1.

by an ophthalmologist revealed bilateral Lisch nodules. He had a large plaque of psoriasis on his scalp, varying in size from 2 to 3 cm in diameter and had erythema and scale. There was hair loss on the lesion. This plaque had been present on his scalp since the age of 6½.

Laboratory values were as follow: Magnetic resonance imaging (MRI) of the head demonstrated hyperintense fields and hamartomatous changes of basal ganglions, cerebellum and medulla oblongata. On electroencephalogram (EEG) there was active epileptiform abnormality originating from the right hemisphere. A biopsy specimen taken from the plaque on his scalp revealed hyperkeratosis with parakeratosis, acanthosis and neutrophilic infiltrates forming microabscesses, all characteristic for psoriasis. The chromosome analysis was normal. Treatment with topical steroid for his scalp lesion and with carbamazepine therapy for his convulsion was started.

Discussion

Neurofibromatosis (NF) is the consequence of an abnormality of neural crest differentiation and migration during the early stages of embryogenesis[8]. NF-1, von Recklinghausen's neurofibromatosis, includes the characteristic skin lesions

which occur in several forms, chiefly neurofibromas, but also café au lait macules, axillary freckles and giant pigmented hairy nevi. The café au lait macule is a hallmark of this disease[8]. The National Institutes of Health Consensus Development Conference[9] identified the following seven important components of the syndrome, two or more which must be present to establish the diagnosis of NF-1: 1) six or more café au lait macules, the greatest diameter of which is more than 5 mm in prepubertal patients and more than 15 mm in postpubertal patients (Crowe's sign), 2) two or more neurofibromas of any type, or one plexiform neurofibroma, 3) freckling in the axillary or inguinal regino, 4) optic glioma, 5) two or more Lisch nodules, 6) a distinctive osseous lesion such as sphenoid dysplasia or pseudoarthrasia, 7) a first-degree relative with NF-1 according to the preceding criteria. Our patient met these criteria for diagnosis of NF-1.

Children with NF-1 are susceptible to neurological complications. Neurological involvement may be focal or diffuse, central or peripheral. Mental retardation, epilepsy, brain tumors, and tumors of cranial nerves may occur[10]. MRI studies in selected children have shown abnormal signals in the globus pallidus, thalamus, and internal capsule, which probably represent low grade glioma or hamartoma that is not detected by computed tomography (CT) scanning[8]. Cranial MRI of our patient showed similar changes as above. These findings may account for the high incidence of learning disabilities. Complex partial and generalized tonic-clonic seizures are a frequent complication[10]. Our patient had some learning difficulties and partial seizure.

The Lisch nodules (hamartomas of iris) present as grayish-white spots, and are visible by direct examination. The optic nerve gliomas may be unilateral or bilateral and are observed in about 15 percent of patients; only about a third are associated with loss of vision[11]. Ophthalmologic examination of our patient disclosed bilateral Lisch nodules, but optic glioma was not detected.

Endocrine disorders such as acromegaly, cretinism, hyperparathyroidism, pheochromocytoma, or precocious puberty may be present. Bone changes (usually erosive) may produce lordosis, kyphosis, pseudoarthrosis, dislocations, and atraumatic fractures[11]. We did not detect any endocrinological abnormality or bone changes in our patient.

In most children within psoriasis of the scalp, other areas of the skin also are involved, showing the typical plaques set on an erythematous base. Occasionally, however, only the scalp may be affected. The base of a scalp lesion is always inflammatory, thus erythema underlies the whitish scale. The scale sometimes forms large plaques, resulting in hair loss[12]. Psoriatic plaque of our patient was only present on his scalp, with similar properties as above.

Although a variety of clinical presentations of NF-1 may occur and the disease can affect many organ systems, there has been suggestion of a direct relationship between NF and psoriasis. Previously there were two reported cases in which neurofibromatosis and psoriasis coexisted. In 1985, Roenigk et al.[6] reported a case with NF-1 and psoriasis. In 1990, Nishimura et al.[7] reported neurofibromas developed in a patient with psoriasis vulgaris during PUVA treatment. Whether or not there is a causal relationship between neurofibromatosis and psoriasis in an intriguing question. Neurofibromas with large numbers of mast cells may occur as a cardinal feature of a wide spectrum of disorders referred to collectively as neurofibromatosis. Mast cells directly or indirectly contribute to the origin and/or growth of neurofibromas; interference with neurofibroma growth might be possible though the use of agents that block mast-cell secretion[13]. Furthermore, mast cell densities are increased in lesional psoriatic skin compared with normal or involved psoriatic skin. If non-involved skin is traumatized, mast cell numbers increase before psoriatic lesinos appear. Whether the presence of neurofibromatosis might predispose a patient to flares of psoriasis is only speculative. In any case, neurofibroma was not present in our patient. Although neurofibromatosis and psoriasis are genetically transmitted diseases in which skin involvement is of primary importance, there have been only two cases in the literature to date. With this case report, we want to draw attention to this association, which may even occur more often than is reported in the literature.

REFERENCES

1. Riccardi VM. Von Recklinghausen's neurofibromatosis. N Engl J Med 1987; 305: 1617.
2. Schmidt MA, Michels VV, Deward W. Cases of neurofibromatosis with rearrangements of chromosome 17 involving band 17 q11.2. Am J Med Genet 1987; 28: 771-777.
3. Ledbetter DH, Rich DC, O'Conell P, et al. Precise localization of N1 to 17 q11.2 by balanced translocation. Am J Hum Genet 1989; 44: 20-24.
4. Bohm M, Vorhees J. White paper on hospitalization for psoriasis care. J Am Acad Dermatol 1984; 10: 842-851.
5. Darmstadt GL, Lane A. Diseases of the epidermis. In: Behrman RE, Kliegman RM, Arvin AM (eds). Textbook of Pediatrics (15th ed). Philadelphia: WB Saunders Company; 1996: 1863-1865.
6. Roenigk RK, Manick KP. Neurofibromatosis and psoriasis. Cutis 1985; 35: 479-480.
7. Nishimura M, Hori Y. Late-onset neurofibromas developed in a patient with psoriasis vulgaris during PUVA treatment. Arch Dermatol 1990; 126: 541-542.
8. Haslam RHA. Neurocutaneous syndromes. In: Behrman RE, Kliegman RM, Arvin AM (eds). Textbook of Pediatrics (15th ed). Philadelphia: WB Saunders Company; 1996: 1705-1707.
9. National Institutes of Health. Neurofibromatosis. National Institutes Consensus Development Conference Statement. Bethesda, Md. The Institute, 1987: 6.
10. Charrow J, Listernick R. Neurofibromatosis type 1 in childhood. J Pediatr 1990; 116: 845-852.

11. Arnold HL, Odom RB, James WD. Some genodermatoses. In: Lamsback W (ed). Andrews' Diseases of the Skin Clinical Dermatology (8th ed). Philadelphia: WB Saunders Company; 1990: 643-645.

12. Tunnessen WW. Pediatric dermatology. In: Oski FA, DeAngelis CD, McMillan JA, et al. (eds). Principles and Practice of Pediatrics (2nd ed) Vol. 1. Philadelphia: Lippincott Co; 1994: 898-948.

13. Riccardi VM. Mast cell stabilization to decrease neurofibroma growth. Preliminary experience with ketotigen. Arch Dermatol 1987; 123: 1011-1016.

14. Cristophers E, Sterry W. Psoriasis. In: Fitzpatrick TB, Eisen AZ, Wolff K, et al. (eds). Dermatology in General Medicine (4th ed) Vol. 1. New York: McGraw Hill, Inc.; 1993: 489-514.