nventional melanoma.[2] We report our experience
.NB in a series of patients with ASTs, who have
r Department. In addition, the outcomes of these
ı retrieved.
4 to May 2011, 15 patients with ASTs underwent
ɔn and SLNB according to the standard proce-
ıa at the Department of Dermatology of Pistoia

ɔf AST was assessed by two or three experienced
sts from two different institutions, who utilized
al criteria for ASTs, as defined by Barnhill [3] (size
try, ulceration, poor circumscription and lateral
expansile dermal nodules, impaired maturation
ation, mitotic activity with deep and/or atypical

80%) women and 3 (20%) men with a mean age
ıge 10-52 years). Three patients were younger
Eleven ASTs (73%) were located on the extremi-
the trunk, and 2 (13%) on the cephalic region.
ranged from 0.75 to 3.5 mm with a mean thick-
Ulceration was present in one case. Nodal in-
ınd in only one case (7%). She was a 47-year-old
ı-ulcerated AST of 1.5 mm in thickness, located
During SLNB procedure a SLN was harvested
ı. Histopathology showed isolated atypical mela-
ɛ sub-capsular and trabecular region of the node.
d complete lymph node dissection (CLND). She
2 months later. All patients were alive and free of
ɛ of review. The follow-up ranged from 42 to 124
4 months). The patient data and SLNB results are
ble I.
ıce we detected only one patient (7%) with AST
This low SLN positivity rate might be related to
ɔf patients in our series. Furthermore, no sentinel
ıs observed in a report of 40 patients with ASTs,
B at the National Cancer Institute of Naples.[4] In
c review, including 24 studies from world-wide
, 541 patients with ASTs showed a good progno-
erall survival during a 5-year follow-up. SLNB
ɛd in 303 (56%) of these patients, who disclosed
119 (39%) cases.[2] Distant metastases with fatal
orted in 6 (1%) cases. Finally, only one of these
en submitted to SLNB (positive SLN) with no
).
clearly indicate that having a SLN positivity for
s does not mean poorer outcome, as happens for
nary melanomas. Consequently, the prognostic
ɔle of SLNB in these patients appears question-
tion of the favorable prognosis even in presence

st authors agree that complete excision with clear
lar clinical follow-up can be reasonable initial
ɔns for patients with ASTs.[2, 4]
seem to indicate a role for fluorescence in situ
SH) analysis to identify patients with ASTs at
gressive clinical behaviour.[2] Cases with homo-
tions have the greatest risk.[5] Therefore, SLNB
rapeutic procedures should be reserved only to these selected patients. Finally, efforts of pathologists to search further prognosticating factors for patient with ASTs should keep on.

Roberto CECCHI [1] *, Laura BARTOLI [1], Luigi BRUNETTI [1], Giovanni TROIANO [1], Cataldo De GAUDIO [2], Stefania INNOCENTI [3]

[1]Department of Dermatology, San Jacopo Hospital, Pistoia, Italy; [2]Department of Nuclear Medicine, Lucca Hospital, Lucca, Italy; [3]Department of Pathology, San Jacopo Hospital, Pistoia, Italy

*Corresponding author: Roberto Cecchi, Department of Dermatology, San Jacopo Hospital, V. Ciliegiole, 51100 Pistoia, Italy.
E-mail: r.cecchi@usl3.toscana.it

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Coexistence of neurofibromatosis type 1 and psoriasis: more than a simple association

Dear Editor,

Neurofibromatosis type 1 (NF1; OMIM 162200), also known as Von Recklinghausen's Disease, is an autosomal dominant condition with an incidence of 1:3000 and a prevalence of 1:4000 to 1:5000.

TABLE I.—*Characteristics of the nine patients with concomitant NF1 and psoriasis.*

Case	Age	Gender	NF1 form	NF1 signs	Other manifestation	Form of psoriasis
1	65	M	Familial	Axillary freckles Café-au-lait spots Neurofibromas Iris Lish nodules	Hypertension, osteoporosis	Psoriatic arthritis
2	77	M	Sporadic	Axillary/inguinal freckles Café-au-lait spots Neurofibromas	Schwannoma	Psoriasis vulgaris
3	48	M	Familial	Axillary/inguinal freckles Café-au-lait spots Neurofibromas		Psoriasis vulgaris
4	29	M	Familial	Café-au-lait spots Neurofibromas Iris Lish nodules		Psoriasis vulgaris
5	64	F	Sporadic	Café-au-lait spots Neurofibromas Iris Lish nodules Scoliosis		Inverted psoriasis
6	53	F	Familial	Axillary freckles Café-au-lait spots Neurofibromas Iris Lish nodules	Thyroiditis, carcinoma of the breast	Psoriasis vulgaris
7	64	F	Familial	Axillary freckles Café-au-lait spots Iris Lish nodules Sphenoid dysplasia		Psoriasis of the scalp
8	18	M	Sporadic	Axillary/inguinal freckles Café-au-lait spots Neurofibromas Iris Lish nodules		Psoriasis of the scalp
9	68	F	Familial	Axillary/inguinal freckles Café-au-lait spots Neurofibromas Macrocrania	Hypertension	Psoriasis of the scalp

Diagnostic criteria include at least two of the following: six or more café-au-lait-colored spots, two neurofibromas or one plexiform neurofibroma, axillary or groin freckling, optic glioma, two Lisch nodules, bone dysplasia and first-degree relative with NF1.[1]

Psoriasis is an inflammatory, immune-mediated and genetically determined skin disease characterized by hyperproliferation of keratinocytes, impaired barrier function and pronounced infiltration of inflammatory cells. Its etiology remains unknown, but the polygenic and multifactorial nature of the disease is well-established, with triggering environmental factors, such as infections, trauma and medications also known to contribute to the development of disease.[2]

In this report, we present nine cases of psoriasis in patients with NF-1 who were diagnosed in a single dermatological unit during the last 15 years.

The mean age of the nine patients with psoriasis and NF1 was 54; there were four women (44.4%). Family history was recorded in six subjects (66.6%).

All presented café-au-lait spots. Some patients had neurofibromas (88.8%), freckles (77.7%) and Lisch nodules (66.6%). Three subjects experienced skeletal changes (33.3%): one had scoliosis, one had sphenoid dysplasia and one had macrocrania (Table I).

In all cases the diagnosis of psoriasis has been made after the diagnosis of NF1.

Three patients had a psoriasis of the scalp, one patient had an inverted psoriasis while five patients had a psoriasis vulgaris (Figure 1). One patient was also diagnosed as psoriatic arthritis, based on the clinical findings of psoriasis and the typical inflammatory arthritis, which was confirmed by a rheumatologist.

The association of NF1 and psoriasis has been rarely reported.

In literature, only other five cases of this association have been described. In 1985, Roenigk reported a 57-year-old man with this association. In 1990, Nishimura observed a 58-year-old man with psoriasis vulgaris and neurofibromastosis. In the patient, neurofibromas had developed during psoralen + ultraviolet A treatment. In 1999, Çelebi described a 7-year-old boy with neurofibromatosis who developed scalp psoriasis. In 2005, Arica reported a 20-year-old woman with NF1 who developed psoriasis bilaterally on the extremities. In 2012, Vasili a 22-year-old woman with NF1 and plaque-type psoriasis.[3]

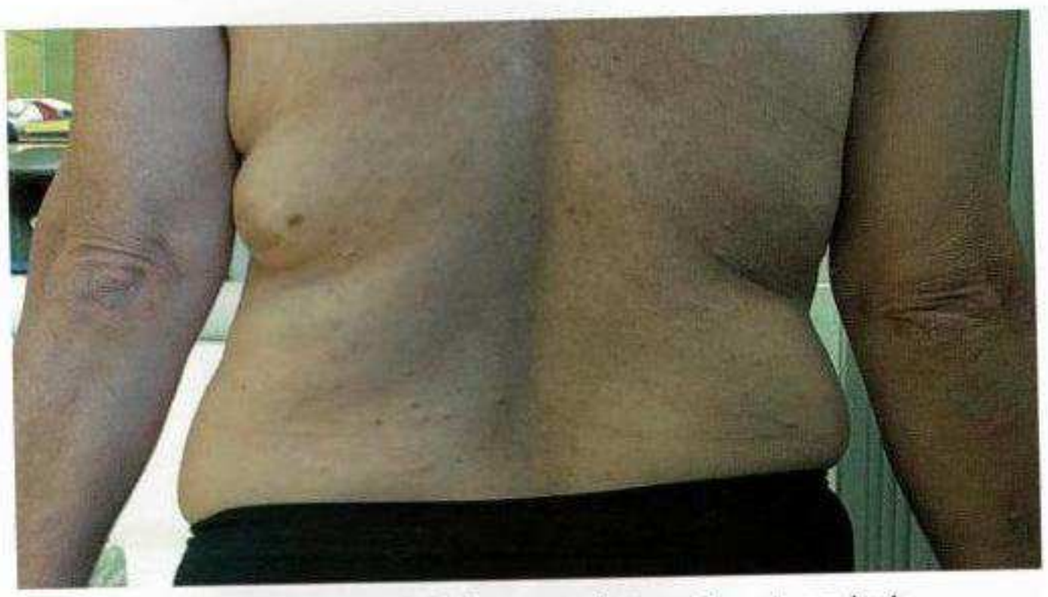

Figure 1.—Patient with neurofibromatosis type 1 and psoriasis.

Psoriasis and neurofibromatosis are both disorders that have a strong genetic basis.

NF1 is caused by a mutation in the NF1 gene located on chromosome 17q11.2 that encodes for neurofibromin, a protein with oncosuppressive activity.[1] Neurofibromin contains a domain related to the GTPase-activating protein (GAP) and accelerates the inactivation of the proto-oncogene RAS in various cell types.

RAS proteins function as molecular switches in many signal transduction pathways, causing alterations in cytoskeletal structure, gene expression and cell-cell interactions.

Reduced levels of neurofibromin and an increased activation of RAS were also demonstrated in psoriatic lesions, although the primary events leading to these alterations in psoriasis remain to be elucidated. Alterations in activity of RAS causes hyperproliferation, altered cytoskeletal organization and altered cell adhesion.[4]

Moreover, Endo found that defects in the regulation of the Hedgehog signaling pathway, due to deficiency of neurofibromin, contributed to the hyperproliferation of lesional keratinocytes in psoriasis.[5]

NF1 and psoriasis share tumor suppressor gene expression defects, which suggests common pathogenetic pathways that should be further and deeply investigated. According to these findings, the association between psoriasis and NF-1 would not seem a coincidental occurrence.

In patients with NF1, the deficit of neurofibromin could, at least in part, predispose to the development of psoriasis.

Emanuele MIRAGLIA *, Franca CANTORESI, Stefano CALVIERI, Sandra GIUSTINI

Department of Dermatology, "Sapienza" University, Umberto I Polyclinic, Rome, Italy

*Corresponding author: Emanuele Miraglia, Department of Dermatology, "Sapienza" University, Viale del Policlinico 155, 00161 Rome, Italy.
E-mail: emanuele.miraglia@hotmail.it

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Localized Darier's Disease

Dear Editor,

Darier disease (DD) is an uncommon genodermatosis which was first reported by Darier and White in 1889. The clinical features of the disease are characterized by the symmetric eruption of the hyperkeratotic, warty papules with red-brown coloration which especially occurring in the seborrheic regions of the body, nail dystrophy and mucosal changes. Lesions may be exacerbated by various factors such as sun light, occlusion, heat, sweating, and stress. The eruption is usually generalized but localized form may also occur. As we know, the localized form of the DD is very rare.[1, 2] We report one case of type 1 localized DD who presented with small, linearly distributed, red-brown papules on the back side of his trunk.

A 50-year-old man patient was admitted to our dermatology clinic due to pruritic red-brown colored lesions on the back side of his body. His lesions had been persisting for about twenty years. Reportedly, the lesions worsened with sweating in the summer months. On dermatological examination, he had multiple red-brown pruritic papules on the midline of the back side of the body in a linear pattern following Blaschko lines (Figure 1). No lesions were found elsewhere on the body, oral mucosa and nail. His medical history and systemic examination revealed nothing. There was no family history of a similar skin disease. Routine blood tests revealed no abnormality. Lesional skin biopsy showed focal vertical hyperkeratosis with parakeratosis, suprabasal clefts and acantholytic dyskeratotic cells. The upper dermis exhibited a slight perivascular inflammatory infiltrate consisting of lymphocytes and eosinophils (Figure 2). Our