s early stage sometimes and some patients of IAPP also have sis lesions on other parts of their bodies.

was reported that IAPP was associated with morphea, which sistent with our case report. Ru-Zhi Z *et al.*[3] reported that a ar-old woman presented with morphea-like atrophic lesions mbosacral portion and thought it might be a special IAPP rtive type or superficial type of morphea. In our case IAPP up after morphea, so we think that IAPP and scleroderma be the same disease spectrum. Mayo Clinic Classification fies 5 main morphea types: plaque, generalized, bullous, lind deep. On the basis of certain clinical and histopathological es, IAPP might be a variant of Morphea en plaque classified ubtype of localized scleroderma.

me scholars believed that IAPP might be a syndrome. Kopeć-ek *et al.*[4] reported that a female patient suffering from IAPP lso diagnosed as papillary thyroid cancer, so they thought APP might be a paraneoplastic syndrome. Kim *et al.*[5] reported 15-year-old patient suffering from juvenile idiopathic arthris also diagnosed as IAPP and thought it might be a Pasini-i-juvenile idiopathic arthritis overlap syndrome.

e disease is benign and may be self-healing after several months rs, but in some cases, the skin lesions may appear forever.

e disease has no effective treatment, topical and systemic corticoid, Vit E, retinoids, niacin, and phototherapy is option-ydroxychloroquine is suggested as the treatment of chronic tory IAPP.

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An unusual manifestation in a patient with Neurofibromatosis type 1

Dear Editor,

Neurofibromatosis type 1 (NF1) (OMIM 162200), also called von Recklinghausen disease, is an autosomal dominant condition with an incidence of 1:3000 and a prevalence of 1:4000 to 1:5000.

Diagnostic criteria include at least two of the following: six or more cafè-au-lait 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]

Patients with NF1 are at increased risk of developing many neoplasms, approximately four times higher than general population matched for age and gender, particularly neoplasia originated from the neural crest derivatives.[2]

Chronic lymphocytic leukemia (CLL) is the most prevalent leukemia in the Western world, with an incidence of approximately 1 out of 100.000 patients for year, and usually affects individuals older than the age of 65. The majority of cases are diagnosed in asymptomatic patients with an incidental finding of lymphocytosis or lymphadenopathy.[3]

We report herein the extraordinary occurrence of CLL in an adult patient with NF1.

A 48-year-old man was examined for the presence of axillary freckling, café-au-lait macules and hundreds of cutaneous and subcutaneous neurofibromas spread over the entire body (Figure 1). On the basis of these findings, the diagnosis of NF1 was made. In addition, on physical examination a generalized lymphadenopathy and a marked splenomegaly were present. The patient reported fatigue and weight loss over recent months.

Hematological studies revealed the following: hemoglobin 9.4 g/dL, WBC $97x10^3$ µL with 93% lymphocytes and platelets $82x10^3$ µL.

Bone marrow examination showed diffuse infiltration of the marrow with around 82% of the cells being lymphocytes. Flow cytometry performed on the peripheral blood showed the lymphocytes to be positive for CD5, CD19, CD20 and CD23 and negative for CD10 and CD7. Cytogenetic studies did not reveal any chromosomal abnormalities.

Based on CD5/CD19 positive lymphocytosis, lymphadenopathy and thrombocytopenia, a diagnosis of CLL, Rai stage IV was made.

He was elected to undergo chemotherapy with fludarabine, cyclophosphamide and rituximab (FCR), with clinical remission.

NF1 is a genetic condition which confers an increased risk of a wide range of cancers.

Although gliomas and neurofibrosarcomas are the most frequent malignant complication of NF1, it is well known that there is also an elevated risk of leukemia, particularly of chronic myeloid leukemia.

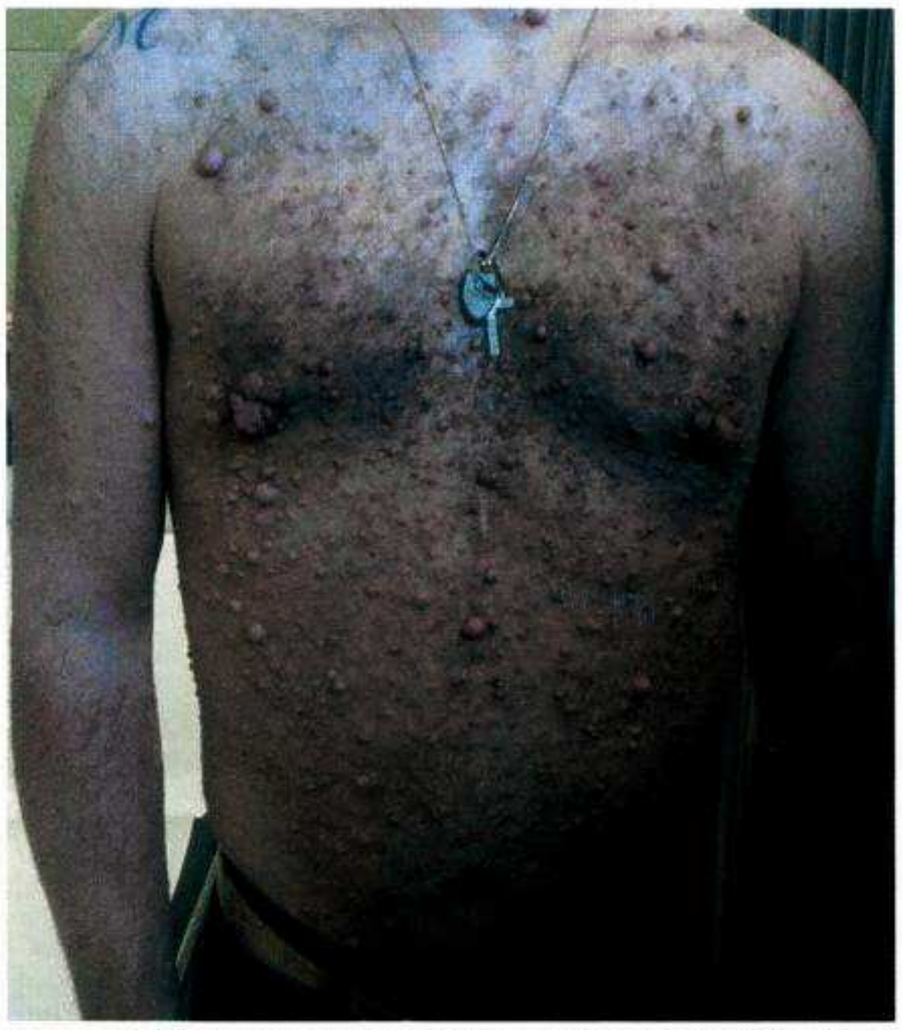

Figure 1.—Multiple neurofibromas of the trunk in a patient with NF1.

The NF1 gene is located on chromosome 17q11.2 and encodes for a protein called neurofibromin. This protein interacts with the ras p21 protein and may regulate ras activity.

Literature also suggests that activating ras mutations may result in increased T- or B-cell malignancies in animals.

The association between hematologic malignancies and germline mutations of NF1 gene has been established in the pediatric setting. Children with neurofibromatosis type 1 have a 500-fold increased risk of developing a rare form of leukemia, known as juvenile myelomonocytic leukemia; a higher incidence of non-Hodgkin's lymphoma and acute lymphoblastic leukemia has also been reported.[4]

In adults patients affected by NF1 the risk of malignancies is well known and increases with age, though the association between NF1 and malignant blood disorders has rarely been demonstrated.

Typical CLL is often found incidentally, on a routine laboratory evaluation. Common manifestations of the disease can be fatigue, autoimmune hemolytic anemia, frequent infections, splenomegaly, hepatomegaly, lymphadenopathy or extranodal infiltrations.[3]

Our digital database (from 1984 to 2013) contains 457 patients with NF1 who undergo regular multidisciplinary follow-up visits and none of them developed CLL so far.

To our knowledge, this is the second case of CLL in an adult patient with NF1.[5]

Although we cannot exclude that such association is a coincidental finding and therefore further studies are needed, we suggest to collect a thorough anamnesis, to perform an accurate skin examination with palpation of the main lymphnode stations and routine annual blood tests including complete blood count and liver function in all patients with NF1.

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Primary idiopathic anetoderma

Dear Editor,

A 32-year-old Italian woman has come to our clinic with a one-year history of progressive eruption of erythematous patches on the trunk and back.

Dermatologic examination revealed many well-defined pink patches disseminated on the trunk. On the back, older lesions became flattened and changed into wrinkled, atrophic skin. During chest bending, older lesions tended to herniate upward (Figure 1A). There were no cutaneous lesions elsewhere. The patient was otherwise in good health, and her past medical history was unremarkable.

Complete blood cell count, erythrocyte sedimentation rate, C3,