

Table 1 Summary of the reported cases of papulotranslucent acrokeratoderma

Author	Family history	Localization	Age at onset	Sex	Associated features	Associated symptoms	Persistence
Onwukwe ²	Present	Palmoplantar	2nd decade	F	Sparse hair, atopia	None	Persistent
English ¹	Present	Palmar	2nd decade	F	Palmar hyperhidrosis, Foot odour	Tightening sensation	Transient
Lowes ⁶	Present	Palmar	(N/S)	F	None	None	Transient
Koster ⁷	Present	Palmoplantar	1st	F			
Heymann ⁸	Present	Palmoplantar	2nd decade	F	None	Painful	Persistent
Lee ³	Absent	Palmar	1st decade	F	None	None	Persistent
Yan ⁴	Absent	Palmar	1st decade	F	Atopia	Burning sensation	Persistent
	Absent	Palmoplantar	3rd decade	F	Atopia	Painful	
	Absent	Palmar	2nd decade	F	None	Burning sensation	
MacCormack ⁵	Absent	Palmar	2nd decade	F	None	Pruritus, pain	Persistent
	Absent	Palmar	2nd decade	F	None	Burning, bubbling	
Present case	Absent	Palmar	4th decade	M	None	None	Transient

N/S: not stated.

Because the review of the literature has disclosed that this particular form of keratoderma could be hereditary or acquired, genetic or sporadic, transient or persistent and that hair abnormality and atopic diathesis could be accompanying features, we believe that the plethora of appellations only perplex this entity (Table 1). Because the only constant feature of all the reported patients including ours is the typical whitish papulotranslucent lesions located on the palmoplantar region that accentuates after a short period of water exposure, the term aquagenic papulotranslucent acrokeratoderma (APA) would be a more descriptive and accurate nosology for this entity.

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A single nodule on the leg as a cutaneous manifestation of myelogenous leukaemia

To the Editor

Leukaemias are a group of disorders characterized by the presence of white blood neoplastic cells in the bloodstream and in bone marrow. Rarely, widespread infiltration of internal organs and skin may be observed. When the skin is involved the prognosis is usually poor. If present, skin infiltration usually develops a few months after the onset of the disease. Occasionally, it may precede haematological manifestations or may be concomitant with the diagnosis of systemic leukaemia.

A 74-year-old man presented with an asymptomatic cutaneous nodule that had appeared 3 months earlier on his right leg and gradually increased in size. Physical examination revealed a painless nodule measuring 5 × 4 cm in diameter with a central necrotic area surrounded by a violaceous and moderately infiltrated halo located on the extensor surface of his right leg (fig. 1).

Routine laboratory tests showed the following abnormalities: haemoglobin, 9.6 g/dL (n.v. 14–18); white blood cells, 20.3 × 10³/μL (n.v. 4.2 × 10³/μL) with a 4.6% haemoblast rate; erythrocyte sedimentation rate, 35 mm/h (n.v. < 15 mm/h); serum fibrinogen, 520 mg/dL (n.v. 180–450 mg/dL). Repeated cytological smears and microbiological swabs were non-contributory.

Histological examination showed a dense collection of neoplastic and benign inflammatory cells located in the dermis



fig. 1 Infiltrated nodule with a central necrotic area located on the leg.

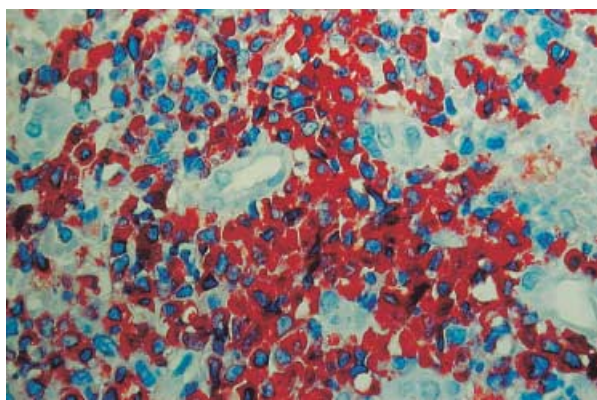


fig. 2 Positive immunoperoxidase staining of neoplastic cells.

with sparing of the upper papillary dermis. Most infiltrating cells had abundant eosinophilic cytoplasm and irregular-shaped nuclei with prominent nucleoli. Some mitoses were observed. Immunoperoxidase staining confirmed the haematopoietic origin of the cells (fig. 2). Bone marrow aspiration revealed a high proportion of large blasts (45%). Immunohistochemistry showed that the majority of these cells expressed myeloid antigens namely, CD33, CD13, MPO, CD34, CD117, consistent with a diagnosis of acute myelomonocytic leukaemia (AML),

subtype M4 according to the French-American-British classification. A total body computerized tomographical scan disclosed hepatic enlargement but no lymphadenopathy or splenomegaly. On the basis of clinical examination, histology and haematological parameters, a diagnosis of leukaemia cutis presenting as a single necrotic nodular lesion in a patient with no systemic involvement was made.

Cutaneous manifestations of leukaemia are classified into two groups: non-specific lesions or *leukaemids*, in which inflammatory lesions contain no leukaemic cells, and specific lesions or *leukaemia cutis*, in which the skin is invaded by malignant haematopoietic cells.¹ *Aleukaemic leukaemia cutis* is a rarer condition characterized by invasion of the skin by leukaemic cells before their appearance in the peripheral blood.¹

Leukaemids are common, occurring in approximately 30% of patients with leukaemia.² Conversely, the incidence of leukaemia cutis changes considerably according to the leukaemia type, ranging from 10% of patients with M0, M1, M2, M3 FAB subtypes to 50% of patients with M4–M5 subtypes and to 20% of patients with granulocytic and lymphocytic leukaemias.^{3,4}

Clinical features of leukaemia cutis are variable and include macules, papules, nodules, plaques, ecchymoses, palpable purpura, and ulcers; erythroderma and bullae may also be observed.² Legs are involved most commonly, followed by arms, back, chest, scalp and face.² In general, the time between diagnosis of systemic leukaemia and leukaemia cutis ranges between 1 month and 4 years, and cutaneous involvement is usually associated with a poor prognosis.⁵

Our case is an example of a rare occurrence of leukaemia cutis presenting as a single necrotic nodular lesion in a patient with no systemic manifestation or complaints.

Clinical features of leukaemia cutis are in general not indicative and histology and immunocytological studies as well as a complete staging procedure are essential for a correct diagnosis. The pathogenesis of leukaemia cutis in myelogenous leukaemia is not well understood and is probably related to a tissue-selective homing of a unique subpopulation of malignant myeloid clones.⁴ The occurrence of necrotic lesions, as observed in our case, has been interpreted by low deformability of blasts with competition for the available oxygen supply.⁶

In conclusion, accurate management is important in patients with leukaemia, in particular in those where skin lesions are the first clinical sign of the disease, as observed in our case, or where skin lesions precede the onset of leukaemia, in order to plan a prompt, therapeutic approach.

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Development of multiple tumours arising in a nevus sebaceus of Jadassohn

To the Editor

Nevus sebaceus (NS) is an infrequent cutaneous malformation, secondary to a disorder in the development of the epithelial, pilar, sebaceous and apocrine structures. It emerges as a congenital plaque of alopecia, which is mainly located on the scalp. The development of a wide variety of neoplasms¹ can occur in the last stage.

We describe an unusual case of the development of four tumours arising in an NS on the scalp, which has been presented for years.

A 68-year-old woman, without significant antecedents, was referred to our department with a congenital alopecia plaque on her scalp that had recently bled. Examination revealed a dark-yellowish verrucous plaque with multiple nodular lesions (fig. 1). Complete excision of the lesion and histological exam-

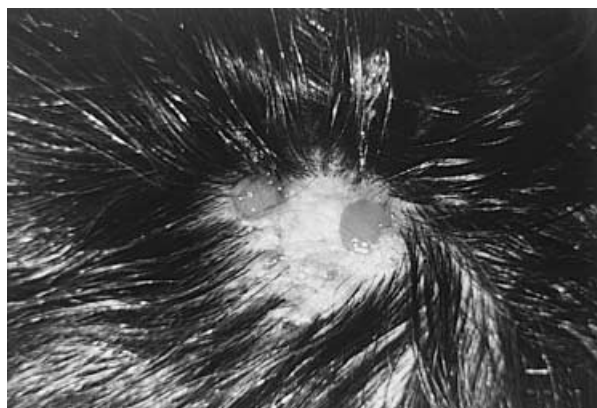


fig. 1 Dark-yellowish verrucous plaque with multiple nodular lesions.

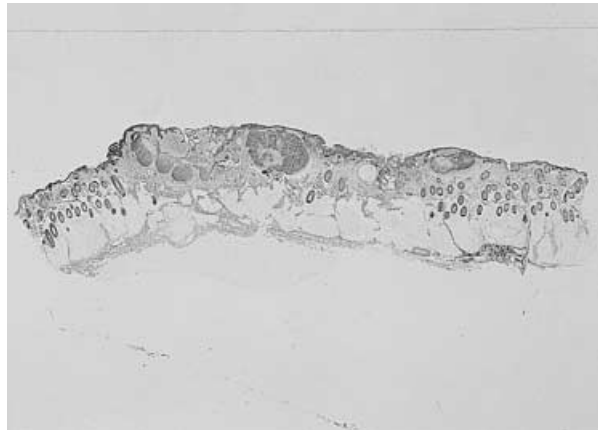


fig. 2 Panoramic view: coexistence of multiple tumours in the surgical specimen.

ination of the sample were carried out (fig. 2). The following tumours arising in the epidermis with hyperkeratosis, acanthosis and papillomatosis as well as in the dermis with many mature sebaceous glands and phenomena of follicular induction were observed. First, a tumour with a superficial papillar shape, whose papillar edges contained plasma cells and a double layer of epithelial cells. Second, a dermal lobuled lesion without connection to the epidermis, with two populations of undifferentiated eosinophilic cells, as well as vacuolated cells with the nucleus indented by such lipidic vacuoles. A third tumour was observed in contact with the epidermis, with a central desmoplastic area and surrounded by a crown of conjunctive tissue, with cells ordered at the periphery without peritumoural tisular retraction. Finally, a fourth multinodular and cystical tumour in contact with the epidermis, whose cavity had a CEA-positive epithelial lining. These findings revealed the coexistence of a papilliferum syringocystadenoma, a sebaceous adenoma, a desmoplastic trichilemmoma and a hidradenoma of clear cells arising in an NS.

The patient remained completely asymptomatic 1 year after the excision.

The neoplastic potential of the NS is well established. The neoplasms or epithelial proliferations, with adnexal differentiation, appeared in the postpubertal or late age.² There is a wide variety of neoplasms that can develop.³ The benign tumours, which have been associated with NS, can have a pilar, sebaceous, eccrine and apocrine origin.⁴

The development of more than one tumour arising in the same NS is not uncommon.⁵ Nevertheless, the coexistence of four tumours, as in our case, is exceptional. Mehregan and Pinkus,¹ in a series of 150 patients with NS, found only four patients who had developed three tumours and only one patient with four tumours. Wilson Jones and Heyl⁶ observed only one case of development of three tumours arising in NS among 140 patients selected. Stavrianeas *et al.*⁷ and Yooh *et al.*⁸ reported, respectively, a patient in whom three tumours arising in a NS had developed and finally, Lillis and Ceilley⁵ reported a case