In Situ Amelanotic Melanoma of the Nail Unit Mimicking Lichen Planus

Report of 3 Cases

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Background: Nail apparatus melanoma is known to be associated with a poor prognosis, mainly because of a delay in diagnosis that is made at an invasive stage. This delay is particularly true in cases involving amelanotic melanoma. To our knowledge, only 1 case of in situ amelanotic melanoma of the nail unit has previously been described. We report 3 cases of in situ amelanotic melanoma with clinical lichenoid features.

Observations: We describe 3 cases of in situ amelanotic melanoma of the nail unit. The patients ranged in age from 39 to 60 years. The lesions were located on the thumb (2 cases) or on the index finger (1 case). The duration of evolution was 6 to 18 months. Nail alterations were characterized by lichenoid changes with longitudinal striation, distal splitting, and nail plate atrophy. Histologic examination revealed in situ amelanotic melanoma extending from the proximal matrix up to the distal part of the nail bed. Complete excision of the nail apparatus was performed. There has been no sign of recurrence after follow-up of 1, 5, and 6 years.

Conclusions: Monodactylic lichenoid nail changes should be added to the more conventional signs of incipient nail melanoma. Chronic unexplained monodactylic nail dystrophy, especially in adults, should always be investigated histologically.

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REPORT OF CASES

CASE 1

A 51-year-old woman presented with brittleness of her right thumbnail that had evolved over 1½ years. She had first consulted a dermatologist who had advised wearing the nail short and had prescribed an amorolfine-based nail lacquer. After 1 year, as the treatment was not effective, she consulted a second dermatologist, who took a nail sample for mycologic examination. The sample did not reveal any onychomycosis, and the patient was then referred to us with a possibility of Bowen disease. She had a medical history of scalp psoriasis but was otherwise in good health and was not following any regular treatment regimen. Clinical examination revealed superficial longitudinal striations with thinning of the distal two-thirds of the median nail plate (Figure 1). Slight distal onycholysis and rare splinter hemorrhages, as well as a thin crevice across the distal nail ridge, were also recorded. The nail unit had never been pigmented. A 3-mm punch biopsy was performed on the distal aspect of the nail bed, at the junction between the detached and attached nail area.

Nail apparatus melanoma (NAM) is known to be associated with a poor prognosis, mainly because of a late diagnosis, and a median Breslow thickness of approximately 3 mm compared with only 1 mm for cutaneous melanoma. One study found that 52% of NAM cases had been misdiagnosed by the first clinician who saw the patient, and this misdiagnosis was responsible for an 18-month median delay in diagnosis. Such delays in diagnosis are particularly true in cases of amelanotic melanoma. We report 3 cases of amelanotic NAM presenting with nonpigmented lichenoid changes that were diagnosed at an early, in situ stage.

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performed. The defect was covered with a skin graft. Examination of the excised specimen confirmed the diagnosis of in situ amelanotic ALM extending from the proximal matrix up to the distal nail bed; confluent atypical melanocytes were mainly observed in the basal cell layers, lying single but also in nests. The higher melanocyte density was observed in the nail bed. There was only rare pagetoid spread of single melanocytes and almost no inflammatory infiltrate. Five years later, the patient was free of disease.

CASE 2

A 39-year-old man presented with dystrophy of his left index fingernail that had evolved over 6 months and had begun with 2 longitudinal ridges. The nail plate had then become thin and brittle. Clinical examination revealed involvement of the median part of the nail, with severe nail plate atrophy and longitudinal thin lines converging toward the center, isolating two lateral parts of normal nail plate (Figure 2). The longitudinal ridges did not affect the most proximal part of the nail plate. The distal aspect of the nail bed was slightly hyperkeratotic. The digital pulp was normal. Monodactylic lichen planus was suspected, and two 3-mm punch biopsies were performed in the proximal area of the nail bed and in the distal matrix. Histologic examination of both biopsy specimens revealed a dense and haphazard intraepithelial pagetoid spread of atypical melanocytes aligned in solitary units with very rare nests. The melanocytes were large, with hyperchromatic nuclei. A few melanophages were seen in the superficial dermis, but there was no inflammatory infiltrate. The diagnosis of in situ ALM was made. Additional consultation with the patient revealed that the 2 longitudinal ridges were initially slightly gray. Total excision of the nail apparatus was performed, with secondary intention healing. Histologic examination of the excised specimen confirmed the diagnosis of in situ ALM with an increased density of atypical melanocytes in the basal and suprabasal layers of the matrix epithelium (Figure 3). The melanocytes were more numerous in the nail bed epithelium, with a few nests. There was no dermal invasion. Six years later, the patient was free of disease.

CASE 3

A 60-year-old woman presented with lateral longitudinal splitting of her right thumbnail, which had disturbed her for several months. There was a red spot in the lunula, with no associated pain. Magnetic resonance imaging did not reveal any pathologic process and especially no glomus tumor. Six months later (Figure 4), there was complete longitudinal nail splitting, isolating a lateral spicule. The whole nail plate was flattened, with thin longitudinal ridges. Two
Amelanotic NAM represents 20% to 30% of ungual melanoma cases compared with less than 7% of the other cutaneous melanomas.1 It usually presents as a chronic paronychia, a torpid granulomatous ulceration, a wartlike keratotic tumor, or a pyogenic granuloma.4,5 It is usually located in the periungual folds or in the nail bed. Clinical misdiagnosis, which is particularly frequent in amelanotic melanoma,3 is responsible for a delay in diagnosis as well as a poor prognosis.

In situ NAM usually starts in the nail matrix and presents as a slowly widening longitudinal melanonychia with possible extension to the periungual skin (Hutchinson sign).6-10 It may also start as multiple longitudinal melanonychias on a single nail. From a histologic point of view, it corresponds to ALM. Histologic diagnosis may be difficult in early cases, especially when small incisional biopsies are involved. To the best of our knowledge, only 1 case of in situ amelanotic NAM has been reported.11 The clinical aspect was a longitudinal erythronychia with mild distal onycholysis. This aspect is different from that observed in our cases.

Our 3 cases were seen by dermatologists who are skilled in nail diseases. However, the clinical diagnosis of melanoma was missed. The first case was diagnosed as psoriasis because of a medical history of scalp psoriasis and the presence of onycholysis with splinter hemorrhages. However, the clinical aspect with longitudinal ridges and nail plate atrophy was more lichenoid than psoriasiform. In both other cases, the clinical diagnosis was monodactylic, isolated lichen planus. Indeed, longitudinal ridging and splitting, nail plate thinning, and focal redness of the lunula are classic signs of nail lichen planus.12 Confronted with the atypical monodactylic presentation that did not affect the whole nail plate, biopsies were performed in all 3 cases to assess the diagnosis and to exclude another process. Partial biopsies were performed because a melanoma was not expected. A total excision of the lesion would otherwise have been performed, thus allowing an accurate histologic diagnosis.

Superficial ridging is sometimes observed in junctional nevus and in situ melanoma. It is probably the result of a dysfunction of the proximal nail matrix that is related to a florid melanocytic hyperplasia. Nail brittleness is more frequent and manifests as distal splitting or onychoschizia, also reflecting nail matrix dysfunction. The severe median nail plate atrophy observed in cases 1 and 2 could be related to the severe nail bed involvement. Whether the nail bed may contribute up to 20% of the nail plate formation is debatable.13 It has been suggested that the matrix involved by the in situ melanoma produces a brittle nail. When the nail plate grows further, the dysfunction of the nail bed then creates the atrophy because its contribution to the nail plate is impaired.

In our cases, the histologic diagnosis was obvious in all biopsy specimens. This obvious diagnosis contrasts with the known difficult diagnosis in early nail melanoma presenting as longitudinal melanonychia. The lesions of our patients were amelanotic and might have remained unnoticed for a long time. This long evolution would also explain the severe histologic involvement of the nail matrix and nail bed by the in situ melanoma.

In conclusion, we diagnosed 3 amelanotic ALMs at an early, in situ stage. In contrast, most invasive cases of amelanotic NAM are nodular melanoma, which explains a completely different clinical presentation. Monodactylic longitudinal splitting, lichenoid nail changes with nail plate atrophy, and longitudinal ridges should be added to the more conventional signs of incipient nail melanoma. To the best of our knowledge, these clinical features have not been reported to date. Chronic unexplained monodactylic nail dystrophy, especially in adults, should always be histologically investigated. Monodactylic nail lichen planus should be confirmed histologically before treatment.

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Figure 4. Right thumb of a 60-year-old woman. Complete longitudinal nail splitting, isolating a lateral spicule. Flattening of the whole nail plate with thin longitudinal ridges and 2 splinter hemorrhages.
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