Malignant nails tumors

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Malignant nail tumors

- Squamous cell carcinoma (Epidermoid carcinoma)
- Melanoma

- Metastasis
- Various bone sarcomas
- Kaposi sarcoma
- Basal cell carcinoma
- Lymphomas
- Merckel cell tumor
- Periungueal eccrine porocarcinoma
- Others
Take home message

• Malignant nail tumors are rare

• Even if diagnosis is suspected, tumors are only diagnosed by an experienced pathologist

• Squamous cell carcinoma is a malignant tumor due to a viral infection

• Melanoma is the most severe cancer but can be cured if diagnosed early
Squamous cell carcinoma

- Bowen’s disease is a Squamous cell carcinoma (SCC) in situ
- Without treatment, 3-5% will evolve to SCC
Etiologies of Squamous cell carcinoma

- Emerging evidence that Bowen’s disease is linked to oncogenic subtypes of HPV (*HPV16-34,35 responsible for genital tract neoplasms*)

- Immunosuppression (transplant recipient, AIDS...) is a high risk factor for HPV induced tumors

- Exposure to Xrays, arsenic, trauma and chronic skin diseases have been associated to digital SCC in the past
Epidemiology

- Long delay in diagnosis (5-7 years)
- Third to sixth decade (# warts)
- Immunosuppression (transplant recipient, AIDS ...)
- Fingers (thumbs especially) are more commonly affected than the toes
- Monodactylous or polydactylous lesions (Bowen’s disease)
Diagnosis

- Scalling and onycholysis that are disproportional to the verrucous changes,
- Periungueal pigmented scalling
- Lateral onycholysis with erosion of the nail bed

AIDS Patient
Diagnosis

- Longitudinal melanonychia
- Periungueal swelling
- Acropachy
- Hyperkeratotic tumors and plaques
- Crusts
- Nail plate dystrophy
- Paronychia
Diagnosis

- First, Think of it

- Biopsy of the lesion at different levels
  - Biopsies should be performed at the deepest portions of the lesion (ulceration, tumors) because
  - Some lesions may show intra-epidermal carcinoma in certain zones and invasive carcinoma in others
 Diagnosis

✓ Pathology requires an experimented dermatopathologist

- Acanthosis with marked hyperkeratosis.
- Anaplasia and disarray that involves its entire thickness.
- Many epidermal cells are atypicals, dyskeratotic, multinucleated, pycnotic, necrotic or with large hyperchromatic nuclei, mitoses.
Diagnosis

• “Bowen’s disease” is an intra-epidermal tumor that may evolve to an invasive squamous cell carcinoma

• It is extremely difficult to differentiate intra-epidermal SCC from invasive SCC or from viral warts

• Multiple biopsies followed by a complete excision are often necessary
Diagnosis

- Bowen's disease
- Invasive SCC
- Wart
- Invasive SCC

Images showing examples of each condition.
What to do?

- X-ray to rule out bone invasion

- Lymph node evaluation (epitrochleal, axillary)
What to do?
(with the help of a dermatologist)

- Complete cutaneous and digital examination to search for a polydactylyous process (rare)
- Investigation of the patient's immune status
- Genital examination of the patient and his or her partner
Treatment of Squamous Cell Carcinoma

- Treatment of squamous cell carcinoma disease should be surgical and conservative and rely on an accurate diagnosis:
  - intra-epidermal VS invasive SCC
Treatment of SCC

• Intra-epidermal squamous cell carcinoma ("Bowen’s disease")

• Early SCC may benefit from carbon dioxide laser vaporization

• Total excision of the lesion with 3 mm margins is considered adequate (pathological control of the margins is mandatory)

• Bleomycin intrallesional injections, topical imiquimod or photodynamic therapy are under investigations
Bleomycin injection
(experimental data)

- Immunosuppressed patient (heart transplant)
Surgical Treatment

- Complete excision of the lesions
In situ SCC
Lateral excision
Treatment of invasive Squamous Cell Carcinoma

- Moh's micrographic surgery is the treatment of choice

- Total excision of the lesion with 5 mm margins is considered adequate (pathological control of the margins is mandatory)

- SCC invasive to the bone require amputation of the distal phalanx

- Axillary lymph node dissection should be performed in the presence of a palpable node, sentinel node biopsy should be considered otherwise
Treatment

- SCC invasive to the bone require amputation of the distal phalanx

After surgery, patients should undergo follow-ups at regular intervals.
Evolution of the disease

- Prognosis of SCC is encouraging despite the frequent delay in diagnosis.
- Evolution is mostly local with a very low risk of distant metastasis (lymph nodes).
Melanomas and longitudinal melanonychia
Melanomas

• Malignant neoplasm derived from melanocytes

• Represent 2-3 % of all melanomas in caucasians (rare lesions)

• Represent around 20% in individuals of dark skin races
Epidemiology

- Sixth decade
- Women > men
- Thumb and great toe are more often concerned
- Amelanotic melanoma account for 15-25% of nail melanomas
Problem in practice

✓ Low frequency of melanomas
  ● 2/3 of Longitudinal melanonychia (LM) are secondary to melanocytic hyperactivity
  ● 1/3 are naevi or lentigos
  ● Only 5% are melanomas
Problem: Clinical diagnosis is very difficult even for experienced dermatologists
These three pictures represent:

- Lentigo
- Jonctional nævus
- Melanoma
The difficulty in practice

- Melanomas seen early carry a very good prognosis
- But the diagnosis may be difficult or impossible on a partial biopsy
- When the clinical picture is suggestive of a melanoma the biopsy SHOULD PREFERABLY INCLUDE ALL the lesion
• It means that
• Any excisional-biopsy of LM carries a high risk of permanent nail dystrophy
Histologic subtypes

- Acral lentiginous malignant melanoma
- Superficial spreading malignant melanoma
- Nodular malignant melanoma.

Less useful for nail melanomas.
Prognosis

- Breslow’s tumor thickness
  - < 0.75 mm
  - 0.75-1.5 mm
  - 1.5-4 mm
  - > 4 mm
Prognosis

- Clark’s level of invasion
  - I  in situ
  - II invades the papillary dermis
  - III invades the papillary reticular-dermal interface
  - IV invades the reticular dermis
  - V invades the subcutaneous tissue
Prognosis

• Diagnostic delay! It is most often the fault of the doctor...
Diagnostic (early)

- Large, progressive LM ($\geq 5$ mm),
- Pigmentation of the periungueal tissue (Hutchinson sign) is frequent but not pathognomonic

Two years natural evolution of nail melanoma in a 34 years old lady who refused treatment

*Breslow 0.66 mm / Clark Level II*
Diagnostic (early)

- Progressive widening
  - (A proximal width of the band superior to the distal indicates a rapid growth rate)
- Thumb, index, great toe
- Traumatism

Breslow 0.55 / Clark level II
Diagnosis (early)

- Light brown bands but more often dark, with variegated colors and multiple fine linear streaks of denser hyperpigmentation.
Amelanotic melanoma
15-25 % of nail melanomas !

♀, 54 yrs
Breslow 2,4 mm /Clark Level IV
Amelanotic melanoma
15-25 % of nail melanomas!

- Breslow 0.45 mm
- 7 yrs evolution

- 33 yrs old, female
  Breslow 2.7 mm / Clark Level IV
  7 yrs evolution

- 62 yrs old, male

- Destruction of the nail plate or fissure
- Tumor
- Granulation tissue - pigmented or not
- Ulceration of the nail bed associated with onycholysis or destruction of the nail plate
- Infection, bleeding or pain
Treatment

- Early diagnosis and surgical removal of NM is mandatory to improve currently poor survival rates.

- Surgical principles are similar to MM at other skin sites.

- Wide local surgical excision
Treatment

- Treatment guidelines of MM at other skin sites are well defined and rely upon Breslow thickness

- Recommended Surgical Margins for Melanoma*
  - T1 (<1.0 mm) 1 cm (radial)
  - T2 (1.1-2.0 mm) 1-2 cm, depending on location
  - T3 (2.1-4.0 mm) 2 cm
  - T4 (>4.0 mm) 3 cm

(*Primary surgical closure whenever possible)
Treatment, 1st step

- A well-done biopsy
- Given in one piece, with orientation, to an experienced pathologist
- If negative, nail dystrophy should be limited
Lateral biopsies

- Margin 0.5 to 1 mm
Lateral biopsies

- Margin 0.5 to 1 mm
- Take off the lateral wall
• Margin 0.5 to 1 mm
• Take off the lateral wall
• Remove the ventral and proximal part of the matrix
Lateral biopsies

- Reconstruct the lateral wall using Dubois’s flap
Central LM < 1-2 mm

- Remove proximal part of the nail plate
Central LM < 1-2 mm

- Transverse excision
- Closure with some tension
Central LM < 1-2 mm

- Longitudinal excision
- Close with Johnson’s flap
Central LM > 3 mm

- Excision will be large (3 mm + 1 mm on each side) and nail dystrophy cannot be avoided
Central LM > 3 mm

- Closure with 1 (2) Schernberg’s flap
- Leave it open (spontaneous healing) until the pathologist give the answer
Treatment (in situ melanoma)

- For malignant melanoma in situ we recommend complete excision of the nail apparatus to the underlying bone followed by a full thickness graft.

- No amputation (skin disease, not bone)
Our series

- 13 patients
- 9 Melanoma in situ
- 4 epidermoid carcinoma (2 Bowen’s)
Our series

- 4 years FU
- No nail regrowth
- 5 mm Weber Two-point discrimination
- Normal DIP mobility
- No recurrence
Our series

- 5 epidermal cysts
- 2 patients had some difficulties to accept their fingers
Literature

- « old » literature favors amputation through the MP joint
- Recent papers (decade) favor amputation through the DIP/IP joint
- Most « recent » papers favor nail apparatus excision with nail reconstruction using nail skin graft
Treatment (late stages)

• Amputation
  • DIP/IP joint amputation is enough
  • No benefit of proximal over distal amputations
  • Level of amputation is chosen in order to obtain the best functional outcome
Other treatment

- Sentinel node biopsy
- Interferon
- Other protocols
  - For melanomas research projects
  - I have no experience
Take home message

• Malignant nail tumors are rare

• Nail dermatologist is needed to eliminate differential diagnosis which are not tumors

• Most tumors are only diagnosed by an experienced pathologist

• Any surgeon must have a high suspicion of melanoma facing a patient with a melanonychia