ΑΝΑΣΚΟΠΗΣΗ

Diagnosis and treatment of nail tumours

II. Malignant tumours

ABSTRACT: Malignant tumours of the nail organ are rare. Among them, ungual melanoma and squamous cell carcinoma with its variants are the most important.

Bowen's disease of the nail organ is certainly underdiagnosed. A warty lesion occurring around or under the nail of an individual older than 35 to 40 years of age and not responding to common wart treatment should be biopsied, as this is the most frequent clinical appearance of an early lesion. Histopathologically, it represents carcinoma in situ. When not treated it will become invasive and ultimately metastatic, although this is very rare. Squamous cell carcinoma may also develop de novo in the nail organ. It may look like a hyperkeratotic wart or oozing tumour. Even carcinoma cuniculatum has been observed in the nail organ.

Melanomas of the nail apparatus are pigmented in about two-thirds to three quarters of the cases. They usually start with a longitudinal melanonychia that grows wider and turns darker until nail dystrophy and/or periungual pigmentation develop. Treatment of choice is radical local excision without amputation in early lesions. Defect repair with a free full-thickness skin graft or a cross-finger flap will yield an excellent functional result with acceptable cosmesis. All other malignant tumours including metastases are exceptional.

*Key words:* *Malignant nail tumours, diagnosis, therapy, prognosis.*

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INTRODUCTION

The tip of the finger contains almost all tissues found elsewhere in the body: skin, soft tissue, bone, nerves, vessels, etc. All these tissues may give rise to development of a malignant tumour in addition to metastases from internal cancers. However, in practice, only epidermoid carcinoma of the nail and ungual melanoma are important, as all other malignancies are exceedingly rare.

Of the 121 different tumours and pseudotumours of the nail organ mentioned, in the latest edition of Baran and Dawber's book "Diseases of the Nails and their Management"[1], 24 are malignant, however nail
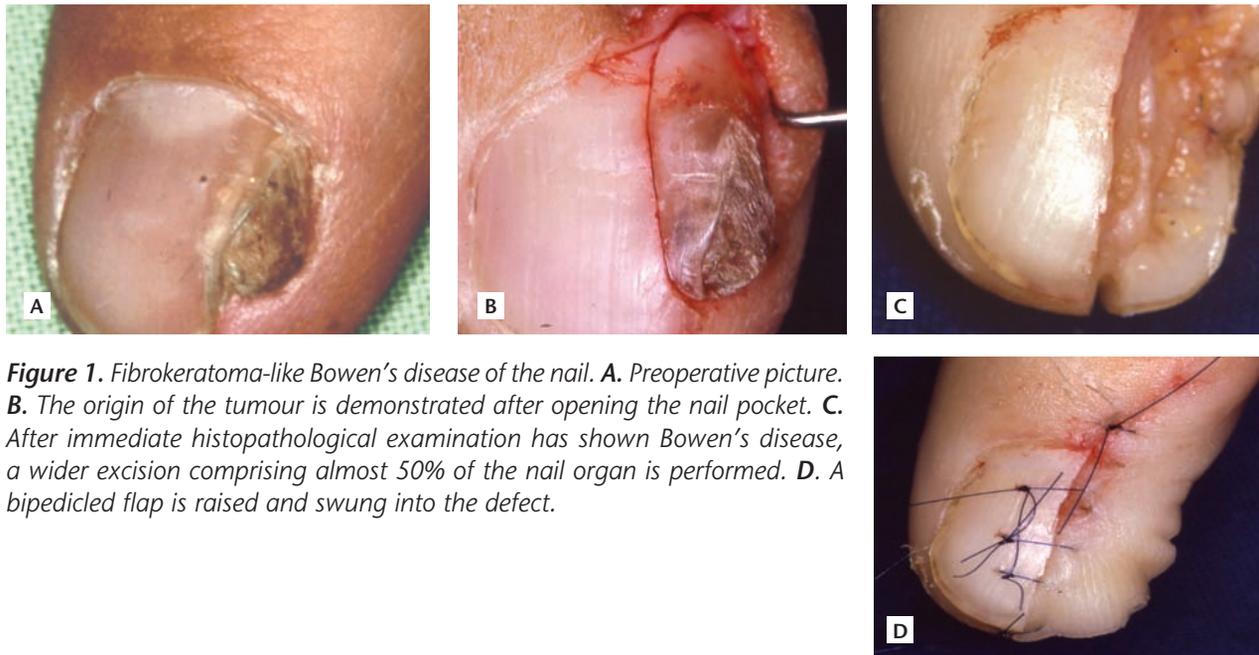
Figure 1. *Fibrokeratoma-like Bowen's disease of the nail.* **A.** *Preoperative picture.* **B.** *The origin of the tumour is demonstrated after opening the nail pocket.* **C.** *After immediate histopathological examination has shown Bowen's disease, a wider excision comprising almost 50% of the nail organ is performed.* **D.** *A bipedicled flap is raised and swung into the defect.*

involvement of malignant lymphomas, other malignant systemic diseases and metastases of internal cancers are not counted. Most of the malignancies are very rare and often only once described[1].

CARCINOMAS OF THE NAIL ORGAN

The term "epidermoid carcinoma" comprises both ungual Bowen's disease and invasive squamous cell carcinoma. It was coined because it was felt that it is not possible to absolutely safely rule out an invasive portion of squamous cell carcinoma in an ungual Bowen's disease[2].

Patients with ungual Bowen's disease are commonly adults in their second half of life, although it has also been observed in adolescents. It is a non-aggressive carcinoma in situ that usually remains non-invasive for many years if not decades until it becomes a frankly invasive squamous cell carcinoma. In recent years, more and more cases have been published due to increased awareness of this particular lesion. Most cases present as a warty lesion insidiously growing out from under the nail plate or proximal nail fold. However, the tumour may be almost entirely subungual and result in considerable nail dystrophy. Infection, inflammation, fissuring and oozing may develop with time. It may induce a longitudinal melanonychia[3,4] or irregular pigmentation[5-7] or may itself be pigmented in persons with dark complexion[1]. Hypertrophic Bo-

wen's disease may mimic an ungual fibrokeratoma (figure 1)[8,9] or even look like a nail of pachyonychia congenita (unpublished observation). Involvement of matrix may cause longitudinal white streaks[10] or red streaks[11] in the nail. Involvement of two or more digits has been described [12-15].

Human papillomavirus types 16, 18 and 35 have repeatedly been demonstrated in ungual Bowen's disease[14,16,17]. Inoculation of anogenital high-risk HPV types like 16 and 35 has been suggested[16].

Histopathology is mandatory for the diagnosis. Apart from the classical features of Bowen's disease, some variants, such as clear cell ones, may be observed[18].

Any warty lesion of the nail in an adult not responding to treatment within 3 to 4 weeks should raise suspicion of epidermoid carcinoma. A representative biopsy should be taken and sent for histopathological examination. Treatment of choice is Mohs' micrographic surgery with immediate defect repair. As many cases originate from the lateral nail groove, a longitudinal partial nail resection and defect repair with a bipedicled bridge flap gives excellent functional results with good cosmesis (figure 1B-C). Metastases of ungual epidermoid carcinoma are extremely rare.

SARCOMAS

Different sarcomas have been observed in the

Figure 2. A wide longitudinal streak in the nail of a 70-year-old woman exhibits a streaky asymmetrical appearance: melanoma in situ.

Figure 3. Widespread Hutchinson's sign (melanotic whitlow) of the index finger of a 48-year-old woman.

Figure 4. Widespread acral melanoma of the toe involving the nail organ in a 79-year-old man *A.* Before surgery, *B.* 6 months after complete removal and defect repair with a full-thickness skin graft.

terminal phalanx. Phalangeal sarcoma may appear as enlarged phalanx or paronychia[19,20]. Epithelioid sarcoma may arise from the synovia of the distal interphalangeal joint and cause swelling of the tip of the digit[20]. It was even observed in a child[21]. Different other types of sarcomas were described as single cases[1].

MELANOMA

Ungual melanoma is still the most serious diagnosis and continues to be a great challenge both diagnostically as well as therapeutically. The problem of a melanonychia has been dealt with in part I of this publication. A longitudinal melanotic streak in the nail of an adult that is wider than 5mm, darkens or widens proximally, started at the age of 40 to 60 years, has a history of a previous trauma, exhibits an irregular colour and has blurred borders, occurs in an individual with a personal or family history of melanoma is suspicious for melanoma (figure 2). Nail dystrophy is a sign of a large melanocyte nidus that occupies a bigger area of matrix epithelium. Thumb, index finger and great toe are the preferred localizations, but a melanoma may occur on any digit's nail. Periungual pigmentation, also known as Hutchinson's melanotic whitlow, is considered almost a proof of malignancy (figure 3). About one quarter to one third of ungual melanomas is amelanotic. An oozing tumour is not infrequently misdiagnosed as an ingrowing nail. In dark-complexioned individuals in

whom streaky brown nail pigmentation is normal the sudden darkening of a single band is suspicious.

Nail melanoma makes up for about 1.5 to 2% of all melanomas. Whether or not trauma is an aetiological or precipitating factor is not yet clear[23].

The treatment of choice is complete extirpation of the affected nail organ. Amputation of the digit or only the distal phalanx was shown not to improve survival in patients with localized disease, i.e. early invasive melanoma[24-27].

There are a number of options to treat ungual melanoma. There is no dispute that the entire nail organ should be removed. This is done by starting an incision over the middle dorsal crease of the interphalangeal joint and carrying it down to the lateral aspects of the digit from where an incision is made down to the bone all around the distal phalanx, approximately 5mm below the level of the nail plate. The dorsal half of the

phalanx is then meticulously dissected from the terminal phalangeal bone with particular emphasis on not leaving any matrix tissue behind. In case of periungual pigmentation a wider excision has to be performed. Delayed Mohs' surgery with formalin-fixed paraffin sections has proven to be superior to determine the exact extension of this in situ melanoma[28]. We have performed several methods of defect repair that will be briefly described.

A full thickness skin graft is taken and sutured into the defect. The free graft is particularly useful for defects that are even bigger and can be used as wrap-around grafts to cover defects of the whole circumference of the digit (figure 4).

A reverse dermal graft is a good option if the blood supply to the recipient site is not optimal. A very thin split thickness skin graft is first raised from the thigh or buttock but left attached to its pedicle. From the underlying dermis, a piece of de-epidermised dermis is taken in same manner as a full-thickness skin graft. It is turned upside down and put on the defect and sutured. This graft has a very high take rate as it does not contain many cells, but it is mechanically resistant as it contains viable dermis with collagen and elastic fibres. It is laid on the defect upside down because the fine capillaries are used during the process of graft take and re-vascularisation; the deep side of this graft contains only few sectioned vessel lumina of the deep dermal plexus. Care has to be taken not to let this graft dry out. Under occlusive dressings, fine granulation tissue will develop on the surface with re-epidermises from the margins or may even be overgrafted with split thickness skin increasing the mechanical resistance of the grafted tip of the digit.

After tumour removal, a pocket is formed under the abdominal skin and the finger inserted and sutured to the abdomen. This must be left in place for approximately 18 to 21 days until the overlaying skin is cut to form a free flap on the distal phalanx. It is then sutured to the fingertip and modelled to form a new distal phalanx[24]. Though this is a safe method, it is inconvenient for the patient to have the hand tightly fixed to the abdomen for almost 3 weeks.

In recent years, we have used the cross-finger flap instead of abdominal skin. A flap is incised from the neighbouring finger, but not yet completely raised, as its random blood supply has to be trained for 8 to 10
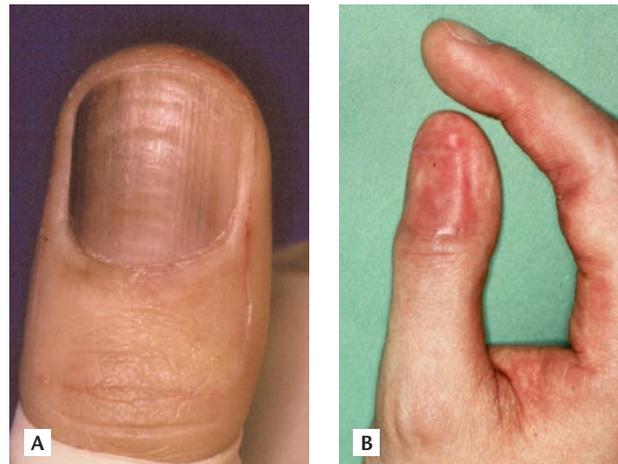
Figure 5*. In-situ nail melanoma of the thumb of a 40-year-old man **A.** Before surgery, **B.** 12 months after resection and defect repair with a cross-finger flap from the proximal phalanx of the index finger.*

days. It is then raised and sutured into the defect with its pedicle left in place for roughly 3 weeks until there is sufficient blood supply from the base of the defect. The donor defect is closed with a full-thickness skin graft. The pedicle is severed and the flap modelled to the fingertip. This gives very good functional and cosmetic results (figure 5)[29].

A controlled study has shown that survival of patients with early invasive ungual melanoma undergoing functional surgery was better than that of patients who had undergone digit amputation[30]. However, far advanced melanomas may still require amputation. The value of sentinel lymph node extirpation in thick ungual melanomas is not yet fully established.

CONCLUSION

Surgery of malignant nail tumours can usually achieve good functional results with acceptable cosmesis[29]. A correct diagnosis and treatment without delay as well as in-depth knowledge of the nail's anatomy and pathology are required.

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