Tumors of the perionychium
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Most tumors of the perionychium are benign. Local tumor cell lines of origin include bone, cartilage, periocyte, nerve, fibrous connective tissue, and skin. The nailbed itself is made up of epithelium and a rich supply of blood vessels and nerves. The nail plate is intimately adherent to the underlying nailbed. Any deformity of the nailbed therefore translates to an irregularity of the nail plate. In the absence of trauma, changes in the nail plate such as grooving, splitting, decreased sheen, or nonadherence may be indicative of underlying tumor growth. In this article, perionychial tumors are divided into benign or malignant and primary or metastatic, and described in terms of their cell line of origin.

Tumors of vascular origin
Vascular tumors include hemangiomas, arterial venous malformations, aneurysms, pericytomas, pyogenic granulomas, and glomus tumors. In general, vascular tumors of the perionychium are uncommon. Pyogenic granulomas and glomus tumors, however, are the most common. Hemangiomas, arterial venous malformations, and aneurysms are usually extensions from surrounding tissues of the digit, which encroach upon the perionychium rather than primary in this area.

Glomus tumor
Typically, glomus tumors have been described as presenting with a distinct triad of symptoms. Paroxysmal pain, cold sensitivity, and pinpoint tenderness at the site in question are characteristic of glomus tumors [1–3]. It is extremely uncommon to be able to palpate a mass or see any nail or skin changes because the tumor is so small and deep in the subcutaneous tissue or underneath the nailbed. Occasionally, a bluish hue or nail plate irregularity is observed. The symptoms progress over months to years. The etiology of glomus tumors is poorly understood; however, it does appear to arise from the vascular network of the glomus body. Although trauma has not been clearly linked to the growth of the tumor, patients will occasionally give a history of some preceding trauma and then subsequently notice symptoms [3]. The diagnosis of glomus tumor is usually made by clinical findings in the history and physical examination. Direct pressure over the site will reproduce the pain. The application of an ice cube over the tumor generally reproduces the exquisite pain as well. Magnetic resonance imaging (MRI) has also proven to be a valuable tool in detecting glomus tumor especially if multiple glomus tumors are suspected [4,5]. Although MRI is useful, clinical suspicion and working diagnosis of the glomus tumor alone is usually enough to merit surgical exploration. Plain radiographs rarely reveal any evidence of tumors, but occasionally cortical erosions from the mass effect can be visualized [6].

Glomus tumors make up 1–5% of the soft tissue tumors of the hand [7]. They arise from the glomus body, which is a specialized arterial venous anastomosis, which regulates blood flow and temperature at the fingertip. The glomus body itself is made up of five units. There is an afferent vessel and an efferent vessel, a capillary network known as a Suqet-Hoyer shunt, and a fibrous capsule [8]. Glomus tumors are usually located within a confined space between the nailbed and distal...
phalanx but may, on occasion, be in the soft tissue of the perionychium. These tumors typically measure 2–3 mm in diameter and are well-defined oval or circular, firm masses. Symptomatic glomus tumors of the nailbed require removal of the nail plate, a longitudinal incision in the nailbed directly over the mass and direct excision of the underlying tumor (Fig. 1A–E). The tumor is well circumscribed and is easily freed from the local tissue upon removal. A wedge or a bilenticular excision of sterile matrix is not required [9]. The nailbed, therefore, can be closed primarily under loop magnification and a 7-0 chromic suture. The nail plate is replaced under the eponychial fold and sutured in place. Short longitudinal incisions in the nailbed decrease the risk for abnormalities of the nail plate after its regeneration (Fig. 2A–F). On rare occasion, the nailbed may be compromised because of the pressure of the underlying tumor preventing primary closure. A split thickness sterile matrix graft can be employed to fill the defect in the sterile matrix [9].

The recurrence rate is rare following excision of the glomus. There have been reports of malignant degeneration of this tumor, although this is extremely rare. Occasionally, glomus tumors present in multiples within the perionychium requiring multiple local incisions to remove them.

**Pyogenic granuloma**

The term pyogenic granuloma is actually a misnomer, because the etiology of the term has little to do with infection (pyogen) but more with chronic inflammation. Originally described as “Sailors lesion,” pyogenic granulomas are a vascular overgrowth made up of hypergranulation tissue. The hands of sea-faring sailors were often fraught with repeated trauma from pulling the ropes and sheets on sailboats. Pyogenic granulomas arose from these chronic wounds.

Typically, pyogenic granulomas are purple to red raised friable, benign tumors of the skin. Because of the repeated trauma to the fingertip,
hyponychium, and perionychium, these sites represent the most common areas of pyogenic granuloma growth. The friable nature of these vascular tumors makes them susceptible to bleeding, either spontaneously or from minor trauma. Histologically, pyogenic granulomas resemble a capillary hemangioma with a proliferative capillary network in a collagen matrix in conjunction with a myriad of inflammatory cells [8]. These tumors will generally grow up to 1 cm in diameter and may have a somewhat pedunculated base. Apart from the other vascular tumors, the differential diagnosis of pyogenic granuloma should also include spitz nevus, foreign body reaction, and amelanotic melanoma (Fig. 3A and B). The treatment modalities that have been used for pyogenic granulomas include silver nitrate application, cauterization, curettage, and if all else fails, excision with a rim of normal tissue to prevent recurrence. Because of the rare but possible inclusion of amelanotic melanoma in the differential diagnosis it is wise to send the specimen for pathologic analysis. The base of the defect is cauterized and the wound closed primarily, allowed to heal-in by secondary intention, or closed with a small split thickness sterile matrix skin graft. Recurrence has been reported, but this rare if a rim of normal tissue has been removed. Pyogenic granulomas do not undergo malignant degeneration.

**Tumors of synovial origin**

**Ganglion cysts**

Ganglion cysts of the perionychium consist of a gel-filled sac originating from the distal interphalangeal (DIP) joint [9]. The pseudocapsule-lined cysts are almost always associated with osteoar-
arthritic changes of the DIP joint. The cyst is more commonly found radial or ulnar to the midline on the dorsum of the finger and often encroaches onto the eponychial fold. The cyst generally ranges from 3–10 mm in size. The mass is often asymptomatic but may be tender, especially with repeated trauma. Occasionally, there is a history of clear drainage from the cyst or from the side of the nail fold. This has led to the inappropriate and outdated use of the term degenerative mucous cyst for these lesions. The ganglion may be seen as a firm, immobile mass overlying the DIP joint or as a more distal mass in the eponychial region. A ganglion above or below the germinal matrix may produce a ridge, groove, or split within the nail plate. Lateral irregularities of the DIP joint confirm arthritic osteophytes (Fig. 4A–D). Radiographs will also confirm arthritic changes within the joint.

Surgical treatment is recommended for ganglions associated with discomfort, nail changes, or pressure necrosis of the overlying skin. Many patients will present with cysts that have undergone aspirations on multiple occasions. Aspiration, however, is doomed to recurrence because the stalk of the cyst and the arthritic osteophyte at the DIP joint have not been eradicated. The past surgical treatment of ganglion cysts of the perionychium consisted of simple cyst excision. Currently, however, the most appropriate treatment
for ganglion cysts in this area should include excision of the inciting DIP osteophyte and simple cyst decompression (Fig. 5A and B) [9,10]. The close proximity of the cyst to the nailbed may result in injury to the germinal matrix during the resection. This results in a permanent nail deformity. With excision of the osteophyte alone, however, a decreased nail deformity rate from 36%–10%, and a 0% recurrence has been observed [11]. It is not necessary to excise the cyst if the osteophyte is removed because the stalk of the ganglion will be removed with this procedure permitting simple drainage of the remaining cyst to eradicate the ganglion.

Fig. 5. (A) A typical distal interphalangeal joint ganglion. The mass is present in the dorsum of the distal finger. (B) The nail deformity in the result of pressure on the germinal matrix. (C) The cyst is visualized through a T-incision. (D) The osteophyte is removed with a rongeur to remove the stock and source of the ganglion. (E) One-year follow-up with resolution of the cyst and nail deformity.
Tumors of connective tissue origin

Fibrous tumors of the perionychium can occur sporadically, or can be associated with some systemic conditions such as tuberous sclerosis. Fibromas present most commonly around the eponychial fold as firm masses. These tumors generally are between 2–6 mm in size and are observed as small growths within the underlying skin. They are generally asymptomatic, but may become tender on occasion. Subungual fibromas are embedded within the sterile or germinal matrix and often result in local pain as well as new grooving of the nail plate (Fig. 6A–C). Fibromas are fibroepithelial tumors that have a fibrous core surrounded by epidermis and a varying degree of acanthosis and orthohyperkeratosis [12,13].

Fibroblast and collagen bundles make up most of the scaffolding. These tumors are devoid of adnexal structures, nerves, and inflammatory cells. The fibromas can be classified as either simple fibromas-acquired digital fibrokeratomas or angiofibromas. Angiofibromas often associated with tuberous sclerosis are similar to simple fibromas but differ by the presence of the stellate type fibroblast, instead of fusiform in their central core.

Digital fibrokeratomas are rare, with less that 100 reported within the literature. These tumors are seen to appear spontaneously, have a rapid growth phase and have a characteristic hyperkeratotic surface [12,14]. The elongated hyperkeratosis often catches on objects and may become painful. Fibrokeratomas within the nailbed may cause onycholysis nonadherence of the nail plate or result in nail grooving.

Dermatofibromas are a subtype of fibromas that arise from the dermis of the skin in the perionychium [15]. Dermatofibromas are also approximately 1–4 mm in size. These tumors are mildly elevated, and are often slightly purplish in color.

The treatment of the connective tissue fibromas depends on the presenting signs and symptoms. Most are asymptomatic, and do not require further surgical intervention. Those fibromas that

Fig. 5 (continued)

Fig. 6. (A) A subungual fibroma resulting in significant deformity to the nail. (B) The nail plate is removed and a radial incision in the eponychium allows exposure of the tumor. (C) The fibroma is removed and the overlying nail bed is preserved.
are painful or are deforming the nail plate will require excision. It is usually possible to excise the lesion with primary closure of the skin or nailbed. If, however, the defect on the skin cannot be closed, the wounds can be allowed to heal via secondary intention, or alternatively, with local flaps.

Subungual fibromas (Koenan’s tumors) are benign, and usually appear during adolescence. Many of these tumors are associated with tuberous sclerosis [16]. Subungual fibromas are reddish brown to flesh colored, and may grow to be up to 1 cm in size, deforming the nail or protruding from the nail folds.

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exostosis, osteochondroma, enchondroma, and osteoid osteomas are benign bony tumors that can present as symptomatic lesions around the perionychium [23,24].

Exostosis

A subungual exostosis is a benign osteocartilaginous overgrowth that usually arises from the ulnar or radial aspect of the distal phalanx. The majority of exostoses are located on the great toe (77%), likely secondary to trauma of wearing shoes or from repeated pressure [7]. The primary symptom is pain, but these lesions can present as firm masses and/or nail deformities. Occasionally the underlying skin of the lateral nail fold or eponychium are affected, and appear as dry, red, scaly, or hyperkeratotic patches. The skin changes can act as a red herring and delay the proper diagnosis of the underlying lesion [13]. The radiologic exam of a chronic exostosis typically shows a sessile or pedunculated expansion of trabecular bone with radiolucent cartilage [13]. The treatment involves nail plate removal, incision in the nail bed or fishmouth incision and elevation of the nailbed to visualize the mass and excision of this osteocartilaginous tumor including involved cortex of the phalanx. The recurrence has been estimated from 6–12% after excision [25].

Osteochondroma

Less common than exostosis, osteochondromas are osteocartilaginous tumors that present with
similar symptoms of pain and possible nail deformity. Osteochondromas are thought to have a congenital rather than traumatic origin and commonly involve the proximal metaphyses rather than the distal bone as with exostoses [26]. Histologically, however, exostosis have a fibrocartilage cap compared to osteochondroma’s hyaline cartilage cap [13]. The treatment of osteochondromas is also excision.

**Enchondroma**

Enchondromas are benign cartilaginous tumors that are often asymptomatic and found inadvertently [27]. Patients may, however, present with a slow growing, painful deformity of the fingertip and/or nail. Occasionally, the intramedullary tumor causes significant weakening of the phalangeal cortex resulting in a pathologic fracture at this site. The classic radiograph reveals a radiolucent cavity and a thin cortex. The tumor cavity should be evacuated and bone grafted as needed. If the patient presents with a pathologic fracture, tumor curettage and bone grafting are delayed until the bone has healed [28]. This may take 1–2 months.

**Osteoid osteoma**

Osteoid osteomas are benign bony tumors of osteoid woven bone and fibrovascular tissue [29]. Around the perionychium these tumors commonly present as a painful swollen fingertip [30–33]. Night pain is a common symptom. Classically, the pain is relieved with nonsteroidal anti-inflammatories or aspirin. Bednar reported that 1.6–5% of osteoid osteomas, however, are not painful [34,35]. The pain is likely related to the release of prostland E2 and local nerve irritation [36,37].

Radiographs are also useful in making the correct diagnosis of osteoid osteomas. Classically there is an area of relative sclerosis with an inner lucency. Bone scans, tomography, angiography, CT scans, and MRI have also been employed to identify this tumor [38–43]. The history and physical findings and plain X-rays, however, are usually adequate to diagnosis osteoid osteomas (Fig. 8a–d). The treatment of osteoid osteomas of the perionychium is usually surgical excision with or without bone grafting [44]. The symptoms and swelling generally completely resolve if the central nidus of the tumor has been removed.

**Tumors of nerve origin**

Schwannoma neuromas or neurofibromas are uncommon tumors of neural origin that may affect the perionychium. Neuromas usually present as point tenderness in areas previously traumatized. Neurofibromas are often associated Von Rechlinghausen’s disease [7]. The treatment of these tender lesions is surgical excision. The other diagnostic studies reported to be of value in determining the diagram of nerve tumors include ultra sound, CT scans, and MRI [19,45–47].

Schwannomas are well encapsulated tumors of Schwann-cell origin that are often asymptomatic. They are histologically organized into hypercellular (Antoni A) and hypocellular (Antoni B) and are generally extraneural growths [48]. Neurofibromas, on the other hand, are often initially localized within the nerve. Histologically, the neurofibromas consist of fibroblasts, Schwann cells, and perineural cells [8,48].

**Tumors of neural crest origin**

**Nevi**

Nevi are pigmented benign tumors of neural crest cell origin [8,13]. The exact diagnosis may be difficult because of their location under the nail plate (Fig. 9A–D). The differential diagnosis of subungual pigmentation includes hematomas, melanonychia striata, petechia benign nevi, or malignant melanoma.

Hematomas are usually diagnosed by a history of trauma. If the patient cannot recall a traumatic event and the diagnosis of hematoma is in question, the nail can be scored transversely at the distal and proximal extent of the pigmented area. The pigment of a hematoma will travel distally with the scored line. If the scored nail proceeds distally as the nail grows but the pigment remains in place, the lesion is not a hematoma [10]. Melanonychia striata are benign strips of tan, brown, or black pigmentation of the nail bed. These striata are thought to be caused by a local increase in number of pigmented production of melanocytes [49]. Melanonychia striata are seen commonly in African Americans but uncommonly in Caucasians.

Melanonychia striata present as small, elongated, pigmented lesions under and within the nail plate. As the nail matrix adds cells to the nail, the pigmented cells of the nevus are added as well. This leads to formation of a pigmented stripe on the nail distal to the tumor. Such lesions can be treated with simple observation.
Subungual nevi offer a challenge to manage because of their potential similar appearance to malignant melanomas. Whether or not the pigmented area has changed in size, shape, or color is valuable in determining the possibility of malignancy. However, the definitive diagnosis is obtained through an incisional or excisional biopsy. The decision to biopsy the lesion is made on an individual basis depending on the patient and the presentation of the nevi. In general, a biopsy with possible deformity of the nail is a small tradeoff to rule out malignant melanoma. If the lesion is less than 2–3 mm, primary closure after the biopsy site is possible, with the promise of the greatest chance for normal nail growth. A lesion larger than 2–3 mm will likely require split thickness or full thickness nail bed grafting, and may be best managed by incisional biopsy to make a diagnosis followed by appropriate treatment. Split-thickness nail bed graft can be used to cover a defect of the sterile matrix, and can be taken from nail bed adjacent to the lesion or from a toe. Because subungual nevi have a high chance of malignant degeneration, complete excision of the nevi with clear margins is recommended for lesions with atypia or dysplasia.

Tumors of inflammatory origin

Warts

Warts are the most common nail tumor. Fingernails are involved more often than toenails, and the prevalent population is children and teenagers (Fig. 10). Warts are caused by infestation of the abraded or macerated skin with the human papilloma virus [50]. These verrucous lesions present as a 1 mm to 10 cm nodular or linear tumor of the hyponychium, or proximal/lateral nail folds. Local nailbed destruction can result in significant deformities. Warts are often present for years, and have an insidious growth.

The management of warts is a challenge because they are known to frequently recur. Fortunately, in children, the natural course is often spontaneous resolution, with 25% of the tumors gone by 6 months and 65% by 2 years [51]. Therefore, aggressive measures are not recommended. Topical keratolytic and virucidal agents may help control the growth of the warts. Systemic therapy, such as intralesional injection of the antimitotic bleomycin, has been effective [13,52]. However, in our institution we have seen several patients who have sustained nail destruction by this chemotherapeutic agent. Surgical treatment of warts includes cryotherapy, excision, and laser therapy. Excision is less favorable due to high rate of recurrence and resultant deformity. Skin grafts or matrix grafts may be necessary for larger areas if the lesions were excised. Wide excision is usually reserved for those cases where the warts are quite symptomatic with pain or nail deformity. More recently, the CO₂, pulsed dye and Yag lasers have been used to obliterate these viral lesions, Yag have been shown to offer the best results with the least morbidity [53].

Fig. 8. (A) An osteoid osteoma of the distal phalanx presents as a swollen tender finger tip. (B) The X-ray reveals a central nidus surrounded by a relative lucency at the most distal aspect of the phalanx. (C) Following nail plate removal, a fish mouth incision permits access to the dorsal distal phalanx. (D) The 1-year follow-up of the asymptomatic finger tip.
Nodular tenosynovitis (giant cell tumor)

Localized nodular tenosynovitis, otherwise known as giant cell tumor, is the second most common tumor seen in the hand (Fig. 11). These tumors are seen in patients around 45 years of age, and have a predilection for females (3:1) [54]. Although the cause of nodular tenosynovitis is unknown, various etiologies have been suggested including trauma, neoplasia, inflammation, and metabolic abnormalities [54]. The tumor presents as a firm, lobulated, gray-yellow-orange, usually nontender mass. The tumor is frequently fixed to the underlying tendon sheath, capsular

Fig. 9. (A,B) A benign nevus of the nailbed. (C,D) An excisional biopsy removes the entire lesion usually with primary closure. Larger lesions may require split nail bed grafts for closure.

Fig. 10. (A,B) The common wart present as verrucous lesion usually isolated to the lateral nail folds.
ligament, bone, or joint. Cortical bone loss may be seen on X-ray from pressure of the mass [55]. Histologically, the tumor is composed of collagenized stroma, hemosiderin pigmentation, multinuclear giant cells, and polyhedral histiocytes [8,56]. These tumors are not malignant but have a very high chance of local recurrence (8–44%) [54]. Therefore, complete meticulous excision of the tumor including deep attachments is necessary.

Gout

Differential diagnosis for tumors around the DIP joint should include gouty tophi. Gout most commonly affects middle-aged to elderly males at a 95 to 5 predominance over females [57]. A family history of gout is present in 85% of patients [55]. The patient presents with hyperuricemia and acute, palpable, multilobular swelling at the DIP joint, frequently associated with swelling of the great toe metatarsophalangeal joint. Gout is diagnosed 50% of the time by clinical evaluation alone because of the diffuse, erythematous tender swelling around the DIP joint [58].

The swelling and gouty calcium deposits can encroach upon the perionychium. Synovial fluid evaluate often reveals the monosodium urate crystals that are needle-shaped, yellow, and negatively birefringent. Hyperuricemia is present in only 5% of the patients with gout [57,59].

The initial treatment of gout includes anti-inflammatory agents and colchicine for the reduction of the hyperuricemia. Surgical treatment is indicated for the excision of painful tophi, the drainage of infection, the relief of compressed nerves, and the improved cosmesis including the excision of tophi to permit the wearing of rings and gloves.

Rheumatoid nodules

A brief mention of rheumatoid nodules is included in the differential diagnosis of subcutaneous nodules of the perionychium. Nodules in this area are often seen with multiple other rheumatoid nodules of the hand and elbow. Symptoms include pain and tenderness upon pressure to the nodules located over bony prominences or weight-bearing surfaces. Steroid injection of the nodules may produce regression, but can also cause ulceration. Surgical excision is generally offered for pain control, erosion of overlying skin, limitation of motion of the DIP joint, or improved cosmesis. Upon excision, the nodule must be carefully dissected from the overlying skin and/or nail matrix.

Malignant tumors

Malignant lesions of the perionychium are frequently misdiagnosed, leading to significant delays in the correct treatment.

Malignant lesions can be mistaken for perionychial infections, pyogenic granulomas, keratoacanthomas, melanomas, warts; trauma, factitious insults, or benign nevi. A high level of suspicion is needed when these lesions appear atypical or do not respond to initial treatment. Biopsies are recommended for any suspicious lesion of the perionychium.

Squamous cell carcinoma

Squamous cell carcinoma (SCC) and SCC in situ (Bowen’s disease) are rare tumors of the perionychium. The average age of presentation is the sixth to seventh decades but has been noted as early as 10 years of age [7]. Male-to-female ratio is roughly 3:1 [60]. The most significant etiologic factor is radiation exposure, either inadvertent or therapeutic. In more recent years, with the knowledge of radiation-induced tumor formation, and avoidance of radiation exposure, this etiology is
much less likely. Human papilloma virus has been thought to play a role in predisposing patients to SCC in the perionychium [7]. The human papilloma virus has been shown to promote malignant degeneration in the urogenital tract, oral cavity, sinuses, larynx, esophagus, and lung [7]. Immunosuppressant drugs may also play a role in SCC transformation as an increasing number of transplant and HIV-infected patients develop SCCs.

SCC lesions of the lateral and proximal nail folds present similarly to other SCC of the skin on other areas of the body (Fig. 12). These tumors are often raised red lesion, with or without ulceration. However, the rate of growth of perionychial SCCs are often much slower than in other areas of the body. This slow growth may lead to their misdiagnosis. Subungual SCC may also present with pain or distortion of the overlying nail (Fig. 13).

Suspicious lesions should be biopsied for SCC in situ. Fleegler recommends removal of the nail plate and nailbed with preservation of the underlying periosteum. The defect is closed with a full thickness skin graft. For invasive squamous cell carcinoma, amputation through the unaffected joint proximal to the lesion is recommended [7, 60,61]. Therefore, an SCC of the perionychium requires amputation at the DIP joint. Carroll et al have shown a cure rate of essentially 100% with this method. Moh’s treatment has been advocated to preserve length and function of digits [60]; however, in a series by Mikhail, the 5-year cure rate was only 79%. In a more recent report by Mikhail, 2 of 24 patients demonstrated a local recurrence, and were successfully treated by further Mohs excision. Goldmitz reported a recurrence in 2 of 25 patients [62]. De Berker reported no recurrences in eight patients treated with Mohs excision and 10% formalin fixation of fresh tumor, hypothesizing that the fixation improved histologic definition [63]. The controversy, as to whether amputation or Moh’s excision is the better treatment for SCC continues to be debated [63]. Advocates of amputation argue that the recurrence rate with Moh’s is too high. Advocates of Mohs micrographic surgery argue that with a low chance of metastasis from subungual SCC, preservation of digit length with a decreased morbidity outweighs the chance for recurrence. When the underlying distal phalanx has been invaded by the tumor, both sides agree that amputation is required.

**Basal cell carcinoma**

Basal cell carcinoma is a very common form of skin cancer especially in the head and neck region. However basal cell carcinomas of the hand and
digit are very rare, with only a handful of cases reported (Fig. 14). The etiology of basal cell carcinoma, on the hand, is thought to be related to radiation exposure [64,65]. The treatment involves of excision of the tumor with free margins reported by frozen section. Distal phalanx amputation is reserved for bony involvement.

**Melanoma**

When evaluating a pigmented lesion of the perionychium the differential diagnosis includes subungual hematoma, foreign body, onychonycosis nigricans, junctional nevi, pyogenic granuloma, paronychia, vascular lesions, fibromas melanonychia striata, melanoma in situ, and melanoma. Most pigmented lesions of the nail are benign; however, such growths should be evaluated carefully to rule out melanoma.

Acral lentiginous melanoma is found beneath the nail, on the palm of the hand, or on the sole of the foot. These lesions represent approximately 3% of all cutaneous melanomas [66,67]. Subungual melanomas make up 1.5% of all melanomas, and are found as acral lentiginous and the nodular histologic type. The great toe and thumb are the most common locations [68]. Fifteen to 20% of melanomas in African Americans are found in the subungual region making the incidence of acral lentiginous melanoma similar in African Americans and Caucasians.

The race of the patient is also important when considering the differential diagnosis. Multiple pigmented streaks of the nails in an African American will likely be melanonychia striata in comparison to a solitary pigmented streak in a Caucasian, which should raise more suspicion of a melanoma. Melanomas can present as either pigmented or amelanotic lesions. A subungual melanoma often starts as a pigmented distal streak under the nail.

Subungual melanomas have a worse prognosis than other cutaneous melanomas (Fig. 15). In comparison to a 72% 5-year survival rate for cutaneous melanomas, Park found a 41% 5-year survival for patients with subungual melanomas [69]. The poorer prognosis is likely due to a delay in the definitive diagnosis [70]. Melanomas were misdiagnosed 40–50% of the time [71,72]. Most common misdiagnoses included nevi, hematoma, or infection. The diagnosis is more difficult with
amelanotic melanomas, which represent 20–30% of all subungual melanomas [49]. Amelanotic melanomas are mistaken for fungal infections or inflammatory conditions. The delay in diagnosis commonly ranges from 1.5 to 2.5 years [67,72]. Because of this delay, 21–30% of patients have metastases at the time of diagnosis [66,73]. Krige reported a 10% regional nodal metastases and 10% systemic metastases [71]. This leads to the lower prognosis with subungual melanomas. Klausner reported a 5- and 10-year survival rate of 28–30% and 0–13%, respectively [74].

Tumor thickness is the most significant prognostic factor for subungual melanomas. Other factors that influence the prognosis include ulceration, mitotic activity, and vascular invasion [49,75]. A biopsy should be performed on any suspicious perionychial lesion. Briggs recommends biopsy of any pigmented subungual lesion that has not changed in 4 to 6 weeks [66]. Fleegler and Zeinowicz recommend biopsy for any dark streaks present at birth or shortly after birth [7]. A lesion with cell atypia or dysplasia should be completely excised with clear margins. Melanoma in situ also requires excision with clear margins of <5 mm. A thorough physical examination of the lymphatic basins is required. Liver function tests and chest X-ray evaluation for metastatic disease are recommended.

Treatment consists of amputation through the joint just proximal to the lesion. For lesions of the thumb, deepening of the first web space with local Z-plasty may improve function.

A lymph node dissection is recommended in the presence of palpable lymph nodes. Elective lymph node dissection in the absence of palpable nodes is controversial. Retrospective studies have shown improved survival in patients with intermediate thickness (1.0–4.0 mm) cutaneous melanomas who undergo elective lymph node dissection in addition to wide excision [76]. Glat recommends axillary lymph node dissection for lesions greater than 1 mm in thickness [77]. Sentinel node mapping has been used in recent years with success at evaluating the lymph node basins while avoiding the morbidity of a full axillary or groin dissection. Balch showed a less than 1% chance of regional lymph node metastasis when the sentinel node was negative [78].

Grover, studied the activity of c-myc oncogene in subungual tumors of 24 patients using flow cytometry [73]. Oncoproteins were found in all tumors at a level nearly twice that of cutaneous melanomas. Subungual oncprotein positivity was 82% compared to 47% in cutaneous melanomas. A significant relationship between high oncogene expression and shorter disease free interval (0.01) was reported. At 5 years, patients below the median onco-protein positively showed a 44% disease-free survival compared to 0% of patients with above median c-myc positively.

Metastatic lesions

Subungual metastatic tumors are rare. Metastatic skin lesions are the first sign of malignancy in only 1% of patients. However, in 44% of patients with subungual metastases, the subungual tumor was the first sign of malignancy. In a review of Henry Ford Hospital cancer cases, six metastatic hand lesions were reported out of 41,835 patients. A literature review by Cohen, demonstrated that subungual metastatic tumors most frequently occur in patients with primary tumors of the lung (41%), genitourinary tract (17%), and breast (9%) [79]. Other primary cancer sources include the oral cavity, colon, and melanoma. Histologically, the most common metastatic lesions to the perionychium are renal cell and squamous cell carcinoma. Bony metastases are often associated with extension into the soft tissue. The distal phalanx is likely to be involved twice as often as the proximal phalanx and three times as often as the middle phalanx. There is no predilection for certain digits.

References


