Comment on Subungual Vascular Malformation with Unusual Presentation

Nail lesions often have a deceptive clinical appearance that mimics a variety of conditions. The diagnostic problems result from the inaccessibility of the lesions to direct inspection, to limited space available for growth, the unique reaction of the nail matrix or nail bed to chronic irritation, the symptom or absence of pain uncommonly seen in other locations, and last but not least from their rarity.

Among the virtually innumerable different vascular lesions–inflammatory, reactive, neoplastic benign or malignant, congenital or acquired–many do not occur or have not yet been observed in the nail apparatus. Vascular malformations form a particular group, which has been defined thanks to the groups of Mulliken and Glowacki[1] and Enjolras[2] and accepted by the International Society for the Study of Vascular Anomalies (ISSVA) in the last 3 decades. Their exact diagnosis is often not easy and many of them were incorrectly classified as "angiomas" in the past. Still now there is often a dispute on whether a particular lesion is of blood or lymphatic vessel origin.

Vascular malformations rarely affect the nail,[3] but this may be seen in association with Klippel-Trenaunay's syndrome and Maffucci-Kast syndrome.

A rare but well-known acquired angiomatoid lesion is intravascular papillary endothelial hyperplasia that commonly develops in a traumatized dilated vein and was also called pseudo-angiosarcoma of Masson. Only one case was described in subungual localization.[4] It had presented as a blue single-digit clubbing of an index finger.

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Acral arteriovenous shunting develops post-traumatically, after autonomic nerve damage as the result of luxury blood supply with high pressure[5] or as a chronic stasis dermatitis associated with venous insufficiency.[6] Clinically, it usually results in a pseudo-Kaposi like appearance with dark red to almost blackish papules and plaques that may also develop keloid-like areas. Acroangiodermatitis of Mali[7] and Stewart-Bluefarb syndrome[8] are variants. The toes and perionychium may be involved; however, the nails are rarely directly affected, but may show a purple hue or be leukonychotic. Some cases were observed in association with an arteriovenous fistula, particularly for chronic haemodialysis.[9]

Lymphangioma circumscriptum is a developmental anomaly usually involving the deep lymph collectors.[10] This leads to dilatation of the superficial lymph capillaries clinically simulating frog spawn. Bleeding into the lymphangioma is common. Involvement of the tip of the digit is rare, but may lead to nail deformation and gross enlargement of the digit. It may pose a problem in the differential diagnosis of Kaposi sarcoma-like conditions.[11]

Infantile haemangioma is the most frequent angioma, but it is extremely rare in the tip of the digit, particularly in the nail organ.

Cirsoid angiomas are arteriovenous tumours most commonly found in the fronto-temporal area as a solitary, dark red nodule. They may be observed in acral[12] and subungual location.[13] Clinically, they appear as a periungual or subungual mass not allowing to make the diagnosis of an angioma. In the published nail cases, either a nodular lesion was observed in the lateral nail fold or under the nail. In another patient, both thumb nails showed an overcurvature and a central wide split (Haneke, unpublished observation). The lesion presented

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as subungual vascular malformation by U Wollina bears some histopathological resemblance, but the presence of many wide, very thin-walled vessels is not characteristic of cirsoid angioma.

Pyogenic or telangiectatic granuloma is an eruptive lobular angioma.[14] It is commonly seen in association with a previous trauma. In the nail organ, it may be located in the lateral nail groove, on the proximal and lateral nail fold, at the hyponychium and in the nail bed and matrix; the latter cause a transungual growth. A rapidly growing, bright red papule of 1 to 2 cm in diameter is seen that often breaks through the horny layer of the skin giving the typical aspect of a collarette. The surface then becomes eroded, crusted and even ulcerated. Some lesions may regress with time, but if this is not the case removal is advocated.

Pseudopyogenic granuloma[15] is now called histiocytoid hemangioma.[16] Involvement of the distal phalanx and the nail was described several times,[17] both as multiple angiomatous nodules in the fingertip, nail bed and lateral nail folds,[18] as involvement of the skin and bone with distal onycholysis, subungual and periungual reddening, longitudinal splitting of the nail, swelling of the nail folds and purulent secretion,[19] or as multiple small painless lesions of a fingernail and a larger bright red, smooth, eroded, vegetating angiomatous nodule destroying the nail plate.[20]

Angiolymphoid hyperplasia with eosinophilia is seen by some authors as another type of histiocytoid angioma characterized by circumscribed proliferation of blood vessels and a chronic inflammatory infiltrate rich in eosinophils.[21-23] Clinically, bright red or violaceous nodules are seen, quite often in the head and neck region. Distal digital localization is rare. Nail bed involvement leads to a reddish or bluish-red discoloration or to nail splitting and nail deformity.[24] One case was seen in association with pachydermoperiostosis.[25]

Acral pseudolymphomatous angiokeratoma of children (APACHE) has also been observed in adolescents and adults.[26,27] Characteristically, multiple small hyperkeratotic angiomatoid lesions are seen on the tips of several digits with striking clinical similarity with angiokeratoma of Mibelli.[28] Single digit plus nail involvement is very rare.[29]

Angiokeratoma circumscriptum is a hyperkeratotic lesion with an angiomatous base. Involvement of the toes, fingers and perionychium has been observed.[30] Verrucous hemangioma and acral pseudolymphomatous angiokeratoma may be indistinguishable clinically. When it is black the major clinical differential diagnosis is melanoma.

Angioleiomyoma of the nail was described already in 1889.[31] Since then, more cases were reported.[32] Because of the pain some were mistaken for glomus tumour.[33,34] Angioleiomyomas of the nail may elevate the nail plate, distort the nail or appear as a small nodule at the tip of the digit just under the hyponychium. They do not clinically appear as an angiomatous lesion.

The glomus tumour was described 200 years ago as a painful subcutaneous nodule.[35] More tumours were later described as colloid sarcoma or angiosarcoma. Because of its highly characteristic symptoms, it is probably the best known nail tumour. Most of them occur in the fingers, particularly in the nail matrix and nail bed. Intense spontaneous pain but also elicited by minor trauma or cold may cause pain that can radiate up to the shoulder. A tourniquet at the base of the finger or a blood pressure cuff inflated to 300 mm Hg is able to alleviate the pain. Most patients are between 30 and 50 years old and women are more frequently affected than men. The glomus tumour is often seen as a bluish or violaceous round to oval spot of 3 to 8 mm in diameter with a reddish line extending distally. The nail may be gently elevated over the lesion or even split. Probing provokes intense pain, but can usually localize the tumour. Dermatoscopy, ultrasound, thermography, dynamic thermography, arteriography, magnetic resonance and particularly angio-MRI help to visualize the lesion, but are rarely more precise than probing. Roughly 10% of glomus tumours are multiple, which may be the cause for presumed recurrence.[36,37] Surgical removal is the treatment of choice. The clinical differential diagnosis comprises virtually all painful conditions of the nails such as subungual warts, keratoacanthoma, subungual exostosis, enchondroma, neuroma, Pacinian neuroma, caliber-persistent artery, leiomyoma, paronychia, osteitis terminalis, subungual felon, herpetic whitlow, causalgia, gout, melanoma[38] and several more. In contrast, glomangioma is often multiple, sometimes in linear distribution involving an extremity including the periungual skin, usually not painful or only tender on deep palpation, and the main clinical differential diagnosis is venous malformation or blue rubber bleb angiomatosis.[39]

Caliber-persistent artery is also known as Dieulafoy's lesion, cirsoid aneurysm or submucosal arterial malformation. It is an acquired or inborn lesion where the terminal artery caliber does not diminish with each branching but remains wide. It is common in the intestinal tract and may be the reason for dramatic or even fatal gastrointestinal bleeding[40] with a lethality of 60%,[41] but it is rare in skin. Its main cutaneous localization is the lower lip.[42] Two lesions causing a split nail were histologically diagnosed as subungual caliber persistent artery. This again bears some clinical resemblance to Wollina's case.

Aneurysmal bone cyst is also called arterio-venous fistula. It is a rare benign, but locally aggressive bone tumour that may also occur in the distal phalanx of young individuals. It grows rapidly, is painful and markedly enlarges the tip of the digit.[43-45] This leads to an enlarged nail. Radiographs show a distension of the bone resembling the secular protrusion of the walls of an aneurysm. The phalanx appears almost completely substituted by an osteolytic process.[46] The etiology is not clear; however, one case of an aneurysmal bone cyst of the distal phalanx was described after a crush trauma.[47]

Acquired fibrokeratoma is a relatively common lesion of the nail and periungual tissue. Histologically, an angiomatous type of fibrokeratoma exists, which is clinically not different from the common fibrotic type and does not have the clinical aspect of an angioma.

Sclerotic haemangioma is a histiocytoma, a very common skin lesion but quite infrequent in the nail organ. Again, it has no clinical resemblance with an angioma.

A lesion rarely recognized clinically as an angiomatous one is eccrine angiomatous hamartoma. It is an infrequent nevoid lesion composed of mature eccrine sweat glands and capillary vessels among and around the eccrine glands. Clinically, it appears as an induration, plaque or nodule, single or multiple. Tenderness or pain, itching, hypertrichosis and often hyperhidrosis are the most common symptoms and signs.[48] Nail involvement is rare[49] and may cause nail destruction.[50] Misdiagnosis has led to amputation.[51] We have observed a peculiar painful subungual neuro-arterio-syringeal hamartoma of the big toe consisting of a well circumscribed proliferation of densely packed eccrine sweat glands, thick-walled arterial vessels and thick myelinated nerves.[52]

Malignant vascular lesions are excessively rare in the nail organ. Depending on the time of their description they were occasionally reported under different terms. The group of angioendotheliomas once grouped between the frankly malignant sarcomas and the benign angiomas are know thought to represent low to medium grade angiosarcomas.[53] Most cases published before 1920 as subungual angiosarcoma[54,55] were probably glomus tumours.

Epithelioid haemangioendothelioma is an uncommon soft tissue tumour.[56] One case of epithelioid haemangioendothelioma presented clinically as a paronychia with a progressive tender swelling of her great toe.[57] A multifocal epithelioid haemangioendothelioma of the sole of the foot and tip of the toes was seen in a 63-year-old woman. MRI scan and digital subtraction angiography showed multifocal bone involvement. Treatment with interferon-α led to partial regression.[58]

Another case of a subungual haemangioendothelioma was thought to be due to prolonged professional contact with vinylchloride.[59]

Originally called sarcoma haemorrhagicum multiplex and mainly seen in the lower extremity of elderly men, Kaposi's sarcoma is now subdivided into the classical form, an epidemic form mainly seen in Africa, and a subtype seen in acquired immunodeficiency syndrome and other immunodepressions including drug-induced immunosuppression. Histopathologically, they are virtually identical. The classical form, but also the other types very often involve the lower legs and feet. Kaposi's sarcoma often affects the nail folds or even overgrows the nail. Subungual Kaposi's sarcoma was found to cause elevation and deformation of the nail plate.[60] A 61-year-old man with "angiosarcoma multiplex" in the distal phalanges of three toes was described. This patient later developed metastases in the calf.[61] Kaposi sarcoma of the nail region in AIDS patients often appears as a small bruise that may turn brown or violaceous. Clinical differential diagnosis comprises pseudo-Kaposi sarcoma in patients with acral hyperstomy syndrome and hyperplastic acroangiodermatitis.

Glomangiosarcoma is a very rare form of a malignant glomus tumour, of which only one case was observed in the distal phalanx of the thumb.[62]

There are certainly more ungual lesions either clinically or histopathologically resembling angiomas of various types. The arteriovenous malformation described by U Wollina adds one more to this interesting list.

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