Introduction

The goal of this CME is to present an introduction to the examination of pigmented nails streaks, which are often overlooked and poorly understood in everyday clinical practice. These seemingly benign lesions can present a difficult clinical challenge because subungual melanoma must always be involved in the differential diagnosis, and quite often a biopsy becomes necessary to confirm what the lesion is. Similarities and differences between melanonychia striata and subungual melanoma will be addressed and clues to the diagnosis of subungual melanoma are emphasized. Important biopsy techniques applicable are considered and the indication for different surgical ap-
Melanonychia Striata

Melanonychia striata is characterized by a tan, brown, or black longitudinal streak within the nail plate and is a relatively uncommon occurrence. Melanonychia striata results from increased melanin deposits in the nail plate. There are many causes and simulators of melanonychia striata, and it is often impossible to differentiate one another by history and clinical inspection alone. More importantly, subungal malignant melanoma must always be included in the differential diagnosis (Figure 1).

If the causes of melanonychia striata are not apparent, then biopsy is necessary. However, this maxim poses a dilemma. In order to get accurate nail biopsy, tissue must be excised that is both adequate for diagnosis and representative of the underlying pathologic process. In order to minimize the risk of post-operative nail dystrophy, enough matrix must be preserved to permit regeneration of a nail plate that is functionally and cosmetically acceptable. Above all, an accurate interpretation of the biopsy specimen requires broad experience in the evaluation of disorders of the nail unit and its melanocyte system by the histopathologist.

The Nail Apparatus and Its Melanocyte System

The nail plate is derived from the nail matrix, a specialized germinative epithelium that lies beneath the proximal nail fold and cuticle. The proximal matrix produces the superior (upper) portion of the nail plate; the distal matrix produces the inferior (lower) portion of the nail plate (Figure 2).

Melanonychia striata originates in the nail matrix and results from increased deposition of the melanin from melanocytes which migrate distally as they transform into nail plate corneocytes. The linear band of the melanized corneocytes is visible in the nail plate.

Melanocytes are normal con-

Fortunately, the majority of these melanocytes are located in the distal portion of the nail matrix, and because of their greater density and activity, pigmented streaks are more likely to originate in the distal matrix.

Figure 1: Nevus. Fine melanonychia striata. Reflection of the posterior nail fold permits an easy visualization of the lesion, which is then excised.1

Figure 2: Formation of nail plate. The proximal portion of the nail matrix forms the upper third of the nail plate; the distal matrix forms the lower two thirds of the nail plate. The level of pigment (dorsal or ventral) within the band corresponds to the origin (proximal or distal) within the matrix.1
bottom half, post-operative dystrophy is less likely.

The level of pigment within the clipped nail specimen may be more accurately ascertained microscopically with Fontana-Masson staining of the nail clipping. This gives an important clue to the pathologist as to precise origin of pigment production and to the appropriate surgical procedure to be selected. But the single most important reason is to determine whether there is a primary melanoma present.

**Distribution of Melanonychia Striata**

Melanonychia striata occurs in 77% of African-Americans more than 20 years of age and in almost 100% who are more than 50 years old. The thumbs and index fingers are common sites as well as the great toes. The more frequently used digits are subject to more traumatic melanoma and have a higher incidence of melanonychia striata. Some authors have linked trauma and friction in both the causes of melanonychia striata and subungual melanoma. But to distinguish the small numbers of patients with subungual melanoma from the larger group of patients with non-specific melanonychia striata is difficult. Both are alike in several ways. In the hand, each arises most often in the thumb, index fingers, or both. Melanonychia striata has been reported to precede the onset of subungual melanoma and may be an early sign. Both occur commonly in dark pigmented people. By some estimates, 40% to 55% of subungual melanoma arise in the foot, and the majority occur in the great toes. It is not known if it is because of trauma or because the great toe offers a greater surface area. Approximately 3% of malignant melanoma in Caucasians are subungual melanoma. Subungual melanoma occurs mainly in older individuals over more than 50 years of age and appears with equal frequency in both sexes.

**Clues to Diagnosing Subungual Melanoma**

A thorough history and physical exam can help distinguish the exogenous causes of a single band of subungual melanoma. Common simulators include subungual hematoma which usually migrates distally. For-
than sharp lateral borders. • Occur in a patient with a history of malignant melanoma. • Occur in a patient when the risk for melanoma is increased (dysplastic nevus syndrome).• Are accompanied by nail dystrophy such as partial nail destruction or disappearances.

Some other important considerations include:

• Black bands may be important consideration in Caucasians for subungual melanoma.
• However, in African-Americans jet-black bands are not unusual.
• Color variation and streaks within streaks may suggest subungual melanoma, but they can be common in multiple benign melanonychia striata.
• Theoretically, wide bands suggest subungual melanoma, but the critical width that signifies melanoma has never been established.
• Bands that do not extend all the way to the free end of the nail are unlikely to be melanomas because they do not take their origin from the nail matrix.
• Bands which are progressively wider indicate rapid growth and are a constant feature of subungual melanoma (Figure 3).
• Multiple pigmented nail streaks are usually not neoplastic in origin.9 A drug history and complete system review can help rule out systemic disorders as the underlying cause of multiple melanonychia striata (Table 1).

Pre-operative Consideration for Melanonychia Striata

Despite meticulous evaluations, too often the cause of melanonychia striata is obscure, and a biopsy becomes necessary. There is no general consensus among pathologists as to the melanocytic causes for melanonychia striata, and therefore, the communication between the clinician and pathologist is critical. The skin pathologist must be provided with a complete history and precise clinical description of the lesion. Photographs are always helpful. It is up to the clinician to provide the pathologist with adequate tissue samples. Nail biopsy interpretation can be difficult, and inadequate tissue sample makes interpretation even more so.

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<td>Chronic: self-inflicted, onychomycosis</td>
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A biopsy has to be performed correctly the first time; patients should not be subjected to mutilating surgical procedures for a condition that may prove totally benign. But at the same time, the pathologist must be provided with adequate tissue that represents the disorder and adequate for the pigmented nail streak within the nail plate.

- Band width and matrix origin (proximal or distal).
- Periungual pigmentation when present with melanonychia striata the likelihood of subungual melanoma is greater.

The big advantage is that the pathologist is able to study the lesion in its entirety, render a precise diagnosis, and draw salient conclusions regarding prognosis.

Imaging studies, both x-rays and/or MRI’s, should be obtained, and the patient examined for lymphadenopathy. All infected portions of the nail apparatus (proximal and lateral nail folds, nail plate, nail bed, hyponychium and skin) are removed en bloc down to bone with relative disregard for cosmetic appearance to ensure complete biopsy and excision. The big advantage is that the pathologist is able to study the lesion in its entirety, render a precise diagnosis, and draw salient conclusions regarding prognosis.

The conspicuous disadvantage is significant post-operative deformity.

**Lateral Portion of Nail Plate Involvement**

The preferred surgical technique is a lateral longitudinal biopsy when melanonychia striata involves past the lateral third of the nail plate. The big advantage is that all affected tissue of the nail apparatus are completely removed, and the dermatopathologist can examine the lesion in its entirety. Post-operatively, the patient is left with a narrowed nail and a good to excellent cosmetic result.

**Mid-portion of the Nail Plate Involvement**

When melanonychia striata lie within the mid-portion of the nail plate, the chance for post-operative dystrophy is greater, and the selection of optimal biopsy technique is more difficult. It is important to establish pre-operatively the matrix origin (proximal or distal) of the streak. Because the more proximal the origin, the greater the risk of nail dystrophy. As noted previously, the origin of the longitudinal streak may sometimes be determined by clinical inspection of the nail plate tips or by microscopic examination of Fontana-Masson stained clippings from the distal free edge of the nail. But a more accurate method for establishing the origin of the pigmented streak is by making surgical relaxing incisions in the proximal nail fold and ascertaining the exact location by direct visualization (Figure 1). These biopsies are best carried out under ring block anesthesia at the base of the digit. The biopsy is then performed with a punch or scalpel. Punch defects smaller than 3mm in diameter need no suture. The punch is run through the soft plate and matrix down to the bone, and the entire specimen is transferred to the fixative. Care should be taken because the matrix tissue is very friable. An antibiotic gauze dressing can then be applied.

**Wide Plate Nail Involvement**

When a wide plate portion of the nail is pigmented, a large portion of the matrix would necessarily be involved. Under these circumstances, the underlying disease process is
Malignant Melanoma of the Nail Apparatus

Early diagnosis and surgical removal of the malignant melanoma of the nail and surrounding tissue is necessary to improve currently poor survival rates. The initial assessment staging and follow-up are similar to that for melanomas on other skin sites. Wide local incision of the lesion is recommended. There are no clear surgical guidelines. For malignant melanoma in situ, complete excision of the nail apparatus down to the underlying bone is recommended, followed by a full thickness skin graft. For invasive malignant melanoma, amputation of the digit is required. There seems to be little difference in survival between patients treated with local proximal interphalangeal joint amputation compared to more proximal amputations—provided that adequate excision of the lesion is performed. The level of amputation is chosen to obtain the best functional outcome. Therapeutic lymphadenopathy is advised where there is clinical evidence of metastatic disease in regional lymph nodes. These melanomas are known to metastasize very rapidly to the brain and eye tissue.

Summary and Conclusion

This article was written to help the clinician identify pigmented nail streaks which are commonly seen and often overlooked in everyday practice. These enigmatic benign lesions can present a difficult clinical challenge because subungual melanoma mimics melanonychia striata, and biopsies are often necessary to distinguish between the two. They should be performed for a single reason: to determine whether there is a primary melanoma.

If there is a pigmented longitudinal nail streak on a toe, then the source of that streak is in the nail matrix, and that is where the biopsy must be performed. A biopsy done anywhere else will result in no information that will help. The nail apparatus can be a trap for both the patient and the clinician, and proof of diagnosis is necessary. PM

Disclaimer: Any application of the information in this article in a professional situation remains the professional responsibility of the practitioner. The primary purpose of the author, Dr. Vannucchi is educational. Information presented and surgical techniques discussed are intended to inform the reader about the knowledge, techniques, and experiences of the author. A diversity of professional opinions may exist, and the views of the author are solely his own.

Acknowledgments

My appreciation is extended to Patrick Vannucchi for editing and preparation of this manuscript.

Endnotes

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This article was written to help the clinician identify pigmented nail streaks which are commonly seen and often overlooked in everyday practice.
1) Which of the following statements regarding melanonychia striata is correct?
   A) Melanonychia striata is characterized by a tan, brown, or black longitudinal streak within the nail plate.
   B) There are many causes and simulators of melanonychia and it is often impossible to differentiate one another solely by history and clinical inspection.
   C) If the causes of melanonychia striata are not apparent then biopsy is necessary to rule out subungual malignant melanoma.
   D) All of the above

2) Systemic disease(s) that can cause pigmented nail streaks include:
   A) Addison’s Disease
   B) Hypertension
   C) Vitamin B12 Deficiency
   D) All of the above

3) Which of the following is more likely to produce a single digit melanonychia striata rather than a multiple digit melanonychia striata?
   A) Malignant subungual melanoma
   B) Nevus
   C) Foreign Body
   D) All of the above

4) Which of the following drugs does not cause multiple digit melanonychia striata?
   A) Ketoconazole
   B) Anti-malarials
   C) Retinoids
   D) Beta Blocking Agents

5) Which of the following is NOT a neoplastic cause for single band melanonychia striata?
   A) Onychomycosis
   B) Metastatic Melanoma
   C) Basal Cell Carcinoma
   D) Verruca Vulgaris

6) Common non-neoplastic causes of melanonychia striata include:
   A) Pregnancy
   B) Subungual Foreign Body
   C) Trauma and Friction
   D) All of the above

7) Which of the following statements regarding nail matrix is correct?
   A) The nail plate is derived from the nail matrix which lies beneath the proximal nail fold and cuticle.
   B) Melanonychia striata usually arises in the distal portion of the matrix and not the proximal portion.
   C) Both A and B
   D) Neither A or B

8) Melanonychia striata is more common in:
   A) African-Americans
   B) Hispanics
   C) Caucasians
   D) Native Americans

9) In the foot subungual melanoma arises more commonly on the ______.
   A) Great toe
   B) Second toe
   C) Third toe
   D) Fourth and fifth toe

10) Approximately what percentage of malignant melanomas are subungual in Caucasians?
    A) 1%
    B) 3%
    C) 5%
    D) 10%

11) The median age at which subungual melanoma is diagnosed is _______.
    A) 10 to 20 years
    B) 20 to 30 years
    C) 30 to 50 years
    D) More than 50 years

12) The male/female ratio of causes of subungual melanoma is approximately _______.
    A) 1 : 4
    B) 1 : 2
    C) 1 : 1
    D) 2 : 1

13) Of the following, the best stain for melanoma pigment is the _______.
    A) S-100 immunoperoxidase stain
    B) Fontana-masson stain
    C) Methenamine stain
    D) Colloidal iron stain

14) Hutchinson’s sign is _______.
    A) Pigment observed in periungual tissue.
    B) An important indicator of subungual melanoma but is not pathognomonic.
    C) Both A and B
    D) Neither A or B

15) When longitudinal nail streaks are dark brown and simulate pigmentation of the overlying cuticle and proximal nail fold because of the skin’s transparency, which of the following statements is correct?
    A) The sign is referred to as pseudo-Hutchinson’s sign
    B) Periungual pigmentation is salient, but always specific
    C) Malnutrition and certain drugs do not cause pigmented bands and hyper-pigmentation
    D) After nail surgery for a benign nevus, periungual pigmentation never occurs

16) When a nail biopsy is performed for melanonychia striata which of the following statements is true?
    A) Postoperative nail dystrophy is less likely with proximal matrix procedures than with distal matrix biopsies
    B) The entire source of the pigment production does not have to be removed
    C) Biopsy should be performed more aggressively in older patients because the likelihood for subungual melanoma is greater
    D) Appearance and functional integrity is more important in the toes than in the hands

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Melanonychia Striata and the Evaluation of Pigmented Nail Streaks (Vannucchi)

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