

Chapter 2

What Is Dysplastic Nevus?

Figure 2.1 illustrates cases with disorders discussed in this chapter.

Definitions

What Is Dysplastic?

The dictionary defines dysplastic as an “abnormal growth or development of cells, tissue, bone, or organ.”

What Is a Nevus?

The dictionary defines nevus as “any congenital anomaly of the skin, including moles and various types of birthmarks,” or “any congenital growth or pigmented blemish on the skin; birthmark, or mole.”

So What Is Dysplastic Nevus?

In 1978, the first report on what was named dysplastic nevus DN was published in the archives of dermatology by Wallace Clark and colleagues. The neoplasm was defined as a unique type of melanocytic nevus that occurred in two families with melanoma and multiple such nevi. Because the histological and clinical findings in the nevi of these patients were different from banal nevi, it was called DN, implying with unusual, atypical (or dysplastic) features. Soon after, similar lesions, clinically and histologically, were reported outside the familial melanoma setting, and called sporadic dysplastic nevi. Even in this nonfamilial setting, dysplastic nevi were found to increase a person’s risk for melanoma independent of other melanoma risk factors.

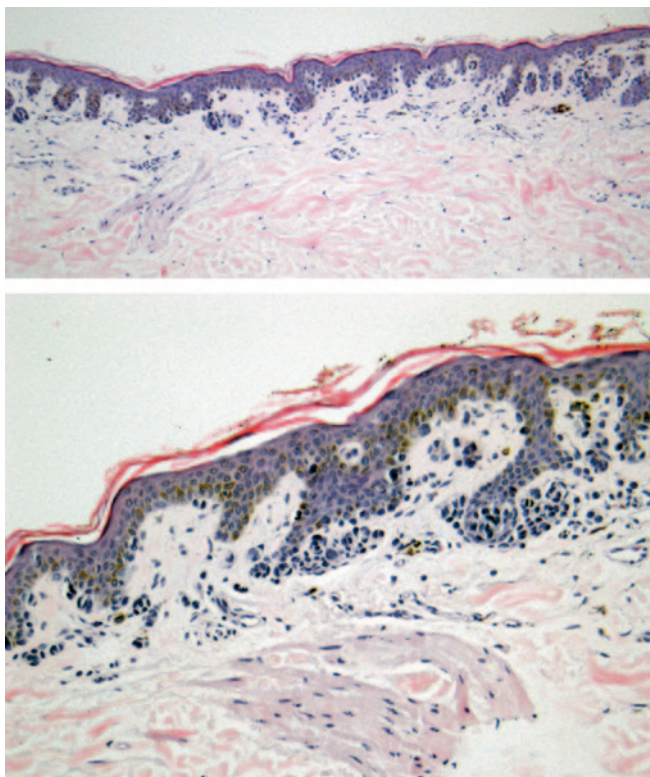


Fig. 2.1 *Top panel* reveals junctional proliferation of melanocytes in a predominantly nested pattern characteristic of junctional nevus. *Bottom panel* reveals that many melanocytes are hyperchromatic and that some nevus cells are seen along the shoulders of the rete, characteristic of the dysplastic nevus

Thus, the epidemic of the dysplastic nevus was born. Around 10% of the pathology material the author reads on a daily basis is to rule out dysplastic or atypical nevus. The neoplasm has progressively penetrated the awareness of the general public, in some cases producing fear in affected individuals, their friends, and relatives. Many times, a patient is told she had pre-melanoma or even early melanoma when the pathology report says “dysplastic nevus.” So how is this nevus diagnosed? What are the defining characteristics of dysplastic nevus? Are they reliable? Are they valid? Are they clinical or histological?

What Are the Defining Characteristics of Dysplastic Nevus?

Clinically, DN has features of the ABCD of melanoma, well known to dermatologists, but are generally stable (hence with very little or no E). Histologically, the dysplasia or atypia is recognized both in the architecture of the proliferation as well as cytology of the nevus cells.

Architecturally, the proliferation is asymmetric and the junctional component of a compound dysplastic nevus extends beyond the dermal component laterally, resulting in the macular and papular components of a lesion. Additionally, junctional nevus nests occupy the sides or shoulders of the rete in addition to the tips which occurs in a banal nevus. Unlike in a banal nevus, nevus cells are also likely to be present as single units among the basal layer, so-called lentiginous melanocytic hyperplasia. A nevus cell may be present in the spinous layer but prominent pagetoid spread is not seen and should raise suspicion for melanoma. In a DN, the papillary dermis also reveals some changes. These include fibrosis, dilated capillaries, melanophages, and lymphocytes, altogether contributing to the pinkish appearance of many dysplastic nevi, especially those that are not pigmented.

In addition to the above architectural features of the dysplastic nevus, there may be cytological or nuclear atypia as well. Unlike in melanoma, the nuclear atypia is random, that is, not uniform among all the proliferating junctional nevus cells. In other words, the atypia is sporadic, affecting scattered individual nevus cells among otherwise unremarkable ones. The degree of atypia has been arbitrarily divided into mild, moderate, and severe.

Using the above criteria, the 1991 NIH consensus conference on dysplastic nevus declared that a non-banal junctional or compound nevus would be interpreted as:

1. With architectural disorder only.
2. With architectural disorder and cytological atypia. Then, the cytological atypia be graded as mild, moderate, or severe.

Most pathologists use either these criteria or personal modifications on them.

Are the Criteria Reliable? And Valid?

To the extent that the above features were used to define DN, they are valid, but not 100% reliable. Interobserver agreement is far from 100%, even among pathologists who practice together, and if read by the same pathologist at different times. Agreement improves, if the group of pathologists agrees in advance on strict diagnostic criteria to be used.

If criteria have been published, why is inter-observer agreement not 100%? Why does the diagnosis of such an important lesion seem subjective?

Architectural Disorder

How many of the above architectural disorder features should be present for a pathologist to diagnose architectural disorder? And how abnormal should each feature be in order to qualify?

Unfortunately, there are no answers to these questions, and it is left to the personal interpretation of each pathologist, contributing to the large degree of variabil-

ity in diagnosis. Additionally, the so-called architectural disorder features are not unique to dysplastic nevi but some, such as melanophages, lymphocytes, and papillary dermal fibrosis, may be seen in early, otherwise unremarkable junctional nevus and lentigo simplex. Nevi in children and early-onset nevi in general often reveal features of architectural disorder. In my experience, general pathologists without training in skin pathology are more likely to exaggerate the degree of atypia.

Cytological Atypia

How many junctional nevus cells, or what percentage of the junctional nevus cells, should reveal nuclear atypia in order for a pathologist to call a nevus dysplastic? Is nuclear atypia in few nevus cells enough?

Unfortunately, there is no answer to this question. For the obvious reason of extreme biologic variability of neoplasms, it has been difficult to address this question.

How About Grading the Atypia?

As mentioned above, there are no reliably measurable criteria to determine whether atypia is mild, moderate, or severe. It is no different from determining whether epidermal hyperplasia, spongiosis, or a lymphocytic infiltrate is mild, moderate, or severe. Thus, it is common for two pathologists reading the same specimen (or the same pathologist evaluating the same specimen at two different times) to differ by at least one grade.

What if the Clinical Features of a Nevus Are Dysplastic but the Histological Findings Are Not?

This situation must happen often. At least one-half of the specimens submitted to rule out dysplastic nevus in the author's experience are banal junctional or compound nevi. Assuming that they looked clinically atypical, what is a clinician to do?

It has been observed that clinically atypical nevi without histological atypia are a risk factor for melanoma, just as nevi defined by the traditional histological criteria as dysplastic.

With this information in mind, why then continue to remove atypically appearing nevi and ask the pathologist to rule out or in dysplastic nevus? The answer to this question is in the mind of each dermatology practitioner and likely varies among practitioners.

What Should the Name Be? What's in a Name?

A few years following the first publication on dysplastic nevi in patients with familial melanoma debate as to the nature of these nevi ensued. Some, championed by Dr. A. B. Ackerman, expressed their disagreement with the term “dysplastic” based on the dictionary meaning of the word and its lack of specificity (see definition of the word “dysplastic” above).

It was stated by these authors that dysplastic nevus is extremely common, and may be the most common type of junctional or compound nevus. Many expressed the view that the dysplastic nevus is one step along the way of a progression of melanocytic proliferations, and that it often evolves into a common junctional or compound nevus. This view had strong support in the observation that the elderly rarely manifest dysplastic nevi.

So to honor Dr. Clark, Dr. Ackerman recommended that the neoplasm in question be referred to as Clark nevus akin to nevus of Reed and Spitz nevus. This term is used in some parts of the USA, especially the East Coast where Dr. Clark did his work. Others prefer the term “atypical nevus.”

Whether dysplastic, atypical, or Clark's, there is a nevus with some clinical and histological characteristics that confers on its patient an increased risk for melanoma. These patients tend to be young, fair-skinned, and of northern European ancestry. The nevus tends to favor the trunk, especially the back and occasionally sun-protected skin of the buttocks, unlike banal nevi. Patients with these neoplasms, especially if multiple, are at higher risk for the development of melanoma and should be screened according to their degree of risk, which includes other well-known melanoma risk factors.

Should All Clinically Atypical-Appearing Nevi Be Biopsied?

There is a wide variation in practice among dermatologists.

On one hand, some (that may be called conservative) make the diagnosis of dysplastic nevus on clinical grounds, inform the patient that they are at increased risk for melanoma and recommend a screening schedule. They may obtain one or two biopsies at the initial visit to help support their clinical diagnosis. Many are skilled at using the dermatoscope, which helps them to suspect melanoma only in rare instances then performing biopsies.

On the other hand, some practitioners seem to remove every atypical appearing nevus. They probably base their practice on the fact that a dysplastic nevus is a potential precursor for melanoma, albeit very rarely.

How Should an Incompletely Excised DN Be Followed?

There are no universal recommendations to answer this question.

Studies based on questionnaires on the behavior of dermatologists in the USA show that the majority re-excise a nevus with cytologic atypia, especially if it is moderate or severe.

As accurate histological evaluation of a melanocytic neoplasm requires submitting the whole lesion, it may be a better clinical practice to ensure removal of the whole lesion at the time of biopsy.

Conclusions

For the foreseeable future, dermatologists will continue to face the questions about what to do when they see an atypical appearing nevus, and when they get a pathology report of dysplastic nevus (or other names implying the same).

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