BEAUTY (AND DEATH) IS ONLY SKIN DEEP?

By

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Color Atlas and Synopsis of Clinical Dermatology: Common and Serious Diseases,
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TABLE OF CONTENTS

INTRODUCTION

There are only a small number of errors committed by dermatologists that ever result in professional negligence litigation. With the exception of metastatic melanomas, basil and squamous cell carcinomas, very few dermatologic problems result in injuries to a patient sufficient to justify the expense of a medical negligence lawsuit. For this reason, there are very few reported cases of dermatology malpractice. As such dermatologists enjoy some of the lowest insurance premiums available to physicians and other healthcare providers.

Unlike most other medical problems, skin disorders are usually visible to the unaided eye of both the physician and patient. No x-rays, MRI's or invasive procedures are required to identify a skin lesion and arrive at a differential diagnosis. In an approach to dermatologic diagnosis there are usually two distinct clinical situations present in a patient complaining of a skin disorder:

1. **Skin changes in a patient that are *incidental* findings in patients complaining of other problems during a routine general physical examination.** These incidental findings include lesions that are present in well people, but not troubling enough to the patient to have resulted in a specific visit. These incidental findings also include important skin lesions which are not noted by the patient but should be recognized by the physician. The most important of these are the cancers consisting of melanoma and carcinomas, dysplastic nevia and café-au-lait macules in Von Recklinghausen's disease. These lesions cannot be overlooked and must be treated aggressively, given their potential for increased morbidity and mortality, if ignored.

2. **Skin changes that are the chief complaint of the patient.** These conditions include minor problems such as rashes, warts, moles, caritosis and others. There are also generalized rashes and lesions associated with underlying diseases such as measles, rubella, herpes, lupus, vasulitis and others.

As in most other medical specialties, there are literally thousands of dermatologic disorders for which dermatologists are trained to recognize and treat. Those disorders may be placed in the following categories:

1. Disorders of sebaceous and apocrine glands
2. Dermatitis
3. Psoriasis
4. Bullous diseases
5. Inflammatory disorders
6. Erythroderma
7. Benign neoplasms
8. Photosensitivity and photo-induced disorders
9. Pre-cancerous lesions and cutaneous carcenomas

10. Melanomas and their precursors
11. Pigmentary disorders
12. Autoimmune diseases
13. Genetic, metabolic endocrine and nutritional diseases
14. Skin signs of vascular insufficiency
15. Skin signs of stomach cancers
16. Skin signs of hemotologic diseases
17. Cutaneous lymphomas
18. Adverse cutaneous drug reactions
19. Disorders of a psychiatric ideology
20. Bacterial infections involving the skin
21. Fungal infections
22. Rickets
23. Viral infections
24. Sexually transmitted diseases
25. Mucocutaneous manifestations of the human immunodeficiency virus disease

While this paper is intended to discuss the standards of care required for an adequate dermatologic diagnosis, the discussion will be limited to those disorders of the skin and mucus membranes consisting of cutaneous carcinomas and metastatic melanomas.

PRINCIPLES OF DERMATOLOGIC DIAGNOSIS

The following principles of dermatologic diagnosis appear in Dr. Thomas V. Fitzpatrick's text: *Color Atlas and Synopsis of Clinical Dermatology: Common and Serious Diseases*, 4th Edition, 2001. These principles appear in other texts and peer review literature regarding dermatologic diagnosis.

I. EPIDEMIOLOGY

Upon arrival in a physicians office a patient's age, race, sex and occupation must be obtained. This information is essential in assisting the physician and obtaining a rapid and accurate diagnosis of a patient's skin disorder.

II. CLINICAL HISTORY

A. Constitutional symptoms

1. "Acute illness" syndrome: headaches, chills, feverishness, weakness

2. "Chronic illness" syndrome: fatigue, weakness, anorexia, weight loss, malay

B. History of skin lesions.

Key questions:

1. When onset?
2. Where onset?
3. Symptoms: does it itch or hurt?
4. Evolution: has it spread?
5. Evolution: has the lesion changed?
6. Provocative factors: heat, cold, sun, exercise, travel history, drug ingestion, pregnancy, season?
7. Previous treatments: topical and systemic?

C. General history of present illness as indicated by clinical situation with particular attention to constitutional and prodromal symptoms.

D. Review of systems as indicated by clinical situation with particular attention to possible connections between signs and diseases of other organ systems.

E. Past medical history:

1. Operations
2. Illnesses
3. Allergies
4. Medications
5. Habits (such as smoking, alcohol, drug use)
6. Atopic history (such as asthma, hay fever, eczema)

F. Family medical history (particularly of psoriasis, melanoma, xanthomas, or tuberous sclerosis).

G. Social history with particular reference to occupation, hobbies, exposures and travel.

H. Sexual history: history of risk factors of HIV, blood transfusions, IV drugs, sexual activity, multiple partners, and sexually transmitted diseases.

III. PHYSICAL EXAMINATION

A. Appearance of Patient: uncomfortable, "toxic", healthy, well.

B. Vital Signs: pulse, respiration and temperature.

C. Skin with special emphasis on four major skin types:

1. Type of Lesions:

    a. flat lesion
    b. elevated lesions
    c. depressed lesions

    The color of the lesions is also important as well as:

    a. consistency
    b. deviation in temperature
    c. mobility
    d. tenderness
    e. estimated depth in relation to depressed lesions

2. Shape of the individual lesions:

    Are the lesions:

    a. round
    b. oval
    c. polygonal
    d. polycyclic
    e. anular (ring shaped)
    f. iris
    g. serpiginous (snake like)
    h. umbilicated

3. Arrangement of multiple lesions:

    a. grouped
    b. disseminated

4. Distribution of lesions:

    The extent of the lesions, are they:

    a. isolated
    b. localized
    c. regional
    d. generalized
    e. universal

    Is there a pattern to the lesions, are they:

    a. symmetrical
    b. exposed areas

c. sites of pressure
d. follicular location
e. random

Characteristic patterns are important such as:

a. secondary syphilis
b. atopic dermatitis
c. acne
d. candidiasis
e. lupus arerthmatosis
f. santhomas
g. and others

D. Hair and nails. Here, texture and color are important, together with any changes, acute or chronic.

E. Mucus membranes.

F. General physical examination. As indicated by clinical presentation and differential diagnosis with particular attention to mucus membranes, a dermatologist must examine for neurologic changes, hepatomegaly joints, and other potential problem sites.

IV. LABORATORY EXAMINATIONS

A. Dermatopathology:

1. Light microscopy: site, process, and cell types;

2. Immunofluorescence;

3. Special techniques: stains, transmission electron mycroscaphy;

4. Microbiologic examination of the skin: scales, crust, exudate, or tissue.

B. Laboratory examination of blood: this examination includes bacterialogic cultures, serologic cultures, hemotologic studies and blood chemistries including fasting blood sugar, liver function and thyroid function tests (if indicated).

C. Imaging, including x-ray, CT scan, MRI and ultra sound, if appropriate.

D. Urinalysis, if appropriate.

E. Stool examination (for cult blood present in vasculities syndromes: parasites and other fecal findings).

F. Woods lamp examination of urine, hair, and skin.

G. Epiluminescence microscopy used for pigmented lesions.

FINAL DIAGNOSIS

Upon conclusion of the history, physical examination and laboratory studies, a clinical diagnosis should be made. Time is of the essence. If the diagnosis cannot be well established at the first visit, the patient must be reexamined in a few days, not a few weeks, as changes may occur and appear in the eruption of skin disorders with new sites of distribution and new types of lesions. A patient's entire clinical picture may become a lot clearer with the passage of only a few days.

DERMATOLOGY MALPRACTICES – BEAUTY (AND DEATH) IS ONLY SKIN DEEP?

SQUAMOUS CELL CARCINOMA IN SITU

Squamous cell carcinoma in situ (SCCIS) primarily results from exposure to ultraviolet radiation (UV) or to infections resulting from the human papilloma virus. Squamous cell carcinoma is also associated with exposure to arsenic, tar, chronic heat exposure, chronic radiation, dermatitis and scarring. Untreated, invasive squamous cell carcinoma (SCC) may arise within squamous cell carcinoma in situ, resulting in lymph node metastasis. The preferred course of treatment, if possible, is cryo surgery, however, the highest cure rate is associated with surgical excision.

INVASIVE SQUAMOUS CELL CARCINOMA

Invasive squamous cell carcinoma (SCC) is a malignant tumor of the keratinocytes, arising primarily in the epidermis, skin appendages and stratified squamous mucosa. Invasive squamous cell carcinoma varies in aggressiveness, with ultraviolet radiation-induced lesions appearing to have the lowest rate of distant metastasis. Squamous cell carcinoma occurring in immunosuppressed individuals can be rapidly metastatic. Onset of squamous cell carcinoma usually occurs in persons older than 55 years of age and occurs in males more frequently than females. Ultraviolet radiation-induced squamous cell carcinoma occurs more frequently in white skinned individuals, but brown or black skinned persons may develop SCC from numerous other etiologic sources. Laborers such as farmers, sailors, life guards, construction workers, and those spending a great amount of time outdoors are at the highest risk for developing SCC. Surgery is the primary method of treatment, with radiotherapy as an alternative, if surgery is not feasible. Overall, the metastatic rate for squamous cell carcinoma is relatively low, around 3-4%. Squamous cell carcinoma occurring in immunosuppressed patients seem to have a higher degree of metastasis.

BASIL CELL CARCINOMA

Basil cell carcinoma (BCC) is the most common type of skin cancer. While basil cell carcinoma is a malignant tumor, it has a limited capacity to metastasize. Basil cell carcinoma is primarily a locally invasive, aggressive and destructive malignancy. The reason for this characteristic is the tumor's growth depends on its stroma, which upon invasion of tumor cells into the vessels, is not disseminated with the tumor cells. When tumor cells lodge at remote sites, further metastatis usually does not occur. Basil cell carcinoma usually occurs in persons older than 40 years of age and occurs more frequently in white skinned persons with poor tanning capability. Exposure to ultraviolet radiation is the primary etiology of basil cell carcinoma. Surgical excision or cryosurgery are the primary methods of managing basil cell carcinomas. Metastasis to lymph nodes, adjacent or remote organs, is extremely rare.

MERKEL CELL CARCINOMA

Merkel cell carcinoma (MCC) is a rare malignant solid tumor thought to be derived from a specialized epithelial cell, the merkel cell. The etiology of this carcinoma is unknown, but may be related to prolonged ultraviolet radiation exposure. This tumor may be solitary or multiple and primarily occurs on the head and on the extremities. Merkel cell carcinoma usually occurs in persons older than 50 years of age and when established, merkel cell carcinoma usually grows rapidly, within one year. Reoccurrence rates from merkei cell carcinoma are high, usually averaging a local reoccurrence within five months. This higher rate of reoccurrence following excision is significant since the disease may spread to regional lymph nodes in more than 50% of the patients and can be disseminated to the visera and central nervous system.

MELANOMA: IN GENERAL

The occurrence of melanoma in white skinned persons within the United States is approaching an epidemic proportion. Approximately one in ninety white skinned individuals will develop the disorder. In males between the ages of 30-49, melanoma of the skin is the second most prevalent cancer (following cancer of the testes) and in older males between the ages of 50-59, melanoma of the skin is the foremost prevalent cancer, (exceeded only by bladder, lung and rectal cancers). Primary melanomas of the skin affect all age groups. The ability to distinguish between melanomas and all other forms of skin cancers is extremely important since early accessibility to physicians is directly related to curability and metastasis. With early detection, the five year survival rate may exceed 90%. This figure is significantly better than the survival rate for melanomas in the early 1970's, where more than 40% of those affected would ultimately die from this metastatic disease.

The primary reason for the higher rate of success in identifying and treating melanomas at an early stage has been increased awareness of the general public in regard to the characteristics of this disorder. Cancer education programs in the 1970's stressed "Danger Signs of Cancer". As a result of this effort, surgical and excisional cures of primary melanomas are now common, however, advanced primary melanomas still have an extremely poor prognosis and survival rate. For this cancer, perhaps more than any other, early diagnosis is critical. The seriousness of this disease therefore has placed an increased responsibility on physicians and other healthcare providers to definitively distinguish melanomas from other forms of skin disorders. Detection and identification of suspicious lesions become the responsibility of the primary care physician, the nurse, the physical therapist or the healthcare provider who observed the total skin of the body. Dermatology textbooks warn that most melanomas are not visible in fully dressed persons because the most common site for the development of this disorder is on the back of both males and females. Medical textbooks and peer review journals unequivocally recommend that in any clinical practice, no matter what the patient's presenting complaint, a total examination of the body should be requested of all non-pigmented (white skinned) patients at the time of the first encounter.

RISK FACTORS ASSOCIATED WITH MELANOMAS

Current medical literature strongly recommends that all dermatology patients fill out a detailed questionnaire listing the risk factors associated with primary melanomas. In 1997, at the inaugural meeting of the Brendan Society, a mnemonic device was developed to assist in the identification of risk factors associated with melanoma. The mnemonic device was designated "MMRISK" and occurs as follows:

M MOLDS: A Typical (Displastic or Clark nevus) ($\geq$ 5)

M MOLDS: Common Molds (numerous) ($\geq$50)

R RED HAIR AND FRECKLING: (often these persons have few or no molds)

I INABILITY TO TAN: Skin Prototypes I and II

S SUNBURN: Severe sunburn especially before the age of 14 (relevant only in nervous-associated melanoma)

K KINDRED: Family history of melanoma (family history must be documented with photographs or clear history from a parent that the lesion was present before the age of 4 years). Many confuse "skin cancer" with melanoma when it is actually non-melanoma skin cancer that was recalled. Also, family history is irrelevant in certain forms of melanoma.

If on examination, it is determined that a dermatology patient has one or more of the six (6) risk factors from melanoma, the standard of care requires a total body examination.

Another device used to assist in the diagnosis of malignant melanoma is the "ABCDE" or six signs of malignant melanoma;

A Asymmetry in shape – one half unlike the other half.

B Border is irregular – edges are irregularly scalloped.

C Color is modeled – haphazardly display of colors: shades of brown, black, grey, red and white.

D Diameter is usually large – greater than the tip of a pencil eraser (6.0 mm).

E Elevation is almost always present – surface distortion is assessed by side-lighting, however, melanoma in situ and acro lentiginous lesions may be flat.

Enlargement – a history of an increase in the size of a lesion is perhaps one of the most important signs of malignant melanoma.

CUTANEOUS MELANOMA

Cutanous melanoma in situ, the correct histological definition of cutanous melanoma in situ, is applicable only to atypical malano sites confined to the epidermis. This form of melanoma is further divided into superficial spreading melanomas, nodular melanomas, lentigo maligna melanoma and unclassified.

The most common form of cutanous melanoma in situ is the superficial spreading melanoma. This disorder occurs primarily in white skinned patients and occurs in females slightly more often than in males. The immediate onset for superficial spreading melanomas is 37 years. Risk factors for this disorder include precursor lesions, family history, light skin and excessive sun exposure. Left untreated, superficial spreading melanoma develops a deep vertical growth over a period of months to years. Surgical excision is the primary course of treatment. Superficial spreading melanomas are the most common type, representing approximately 70% of the cutaneous melanomas.

NODULAR MELANOMA

Nodular melanomas (NM) is second in frequency (14%), following superficial spreading melanomas. The median onset of this lesion is approximately 50 years of age and has an equal incident of occurrence in males and females. Nodular melanoma is unusual in that it also incurs in all races with only a slight preference to persons of light skin color. Predisposing risk factors for nodular melanoma include precursor lesions, a family history of melanoma and excessive sun exposure. This type of melanoma has a relatively rapid growth rate over several months and is often noted by a patient as being a new "mole" that was not present before. This lesion appears as a solid blue or blue-black lesion and is extremely distinctive. Again, surgical excision is the treatment of choice.

LENTIGO MALIGNA MELANOMA

Lentigo maligna melanoma is the least common melanoma with less than 5% of incidents among skin cancer patients. Lentigo maligna melanoma has a relatively delayed onset, occurring usually on or after 65 years of age. The incidence of lentigo maligna melanoma is equal between male and female with predisposing factors, including sun induced non-melanoma skin cancers such as squamous cell or basil cell carcinomas or persons with excessive exposure to UV light. Surgical excision is also recommended as a primary course of treatment.

Less common types of melanoma include acrial lentiginous melanoma (ALM), which is an unusual cutaneous melanoma arising primarily on the sole, palm, finger nail or toe bed. This disorder occurs most often in Asians and African-Americans, comprising approximately 50-70% of melanomas of the skin found in these populations. The median age for occurrence is 65 years and ALM occurs three times more often in males than in females. Predisposing factors for this disorder are unknown, but this

disorder may be in some way related to trauma. The appearance and location of this lesion make it difficult to diagnose, resulting in a 5-year survival rate of less than 50%. Surgical excision is the primary course of treatment.

HISTOLOGIC MICROSTAGING OF MELANOMAS

The survival rates and treatment protocols for melastatic melanomas are primarily based upon the histologic micro staging of the lesion:

Level I	– Melanoma Cells confined to the Epidermis
Level II	– Melanoma Cells invade the Dermal Papillae
Level III	– Melanoma Cells completely occupy Papillae Layer
Level IV	– Melanoma Cells invade Mid Reticular Dermis
Level V	– Melanoma Cells invade Subcutaneous Fat

METASTATIC MELANOMA

Metastatic melanoma occurs with a frequency between 15-26% in Stage I and Stage II melanomas. Metastacis of the disease from the primary site usually occurs in a linear sequence from local reoccurrence to regional metastacis to distant metastacis. Because of this relatively common manner of metastacis, *sentinel node biopsy* can be utilized to provide an accurate prediction of the presence of metastatic melanoma cells within the regional lymph nodes. The presence of micro-metastacis can be detected by sentinel node mapping, which involves specialties in surgery, nuclear medicine and pathology. The hypothesis behind central node mapping is that the first node draining the lymphatic basin, commonly referred to as a sentinel node, can predict the presence or absence of metasticis in all nodes further down the chain. Sentinel node mapping is essential in determining a existence or non-existence of metastatic disease. The overall survival rate at three years is estimated to be greater than 90%, if the sentinel node is free of metastacis and greater than 65%, if the sentinel nodes contains evidence of disease.