

presented a case and defined 3 histologic variants of this entity: type I, a combination of blue nevus and cellular nevus; type II, a combination of blue nevus with nevus spilus; and type III, a combination of blue nevus and fibromatous or myomatous nevoid formation. There is controversy regarding the cause of the association. Some authors consider that this is a random phenomenon,<sup>5</sup> while others believe that nevus spilus provides a favorable environment for the growth of other nevi.<sup>3</sup> The risk of malignant change is low in nevus spilus, although cases of melanoma arising on this pigmented lesion have been reported in the literature.<sup>6–8</sup> Our case presents the rare combination of nevus spilus with agminated blue nevi and, despite the clinical and dermoscopic diagnosis, it was necessary to perform several biopsies of the pigmented plaque to exclude a diagnosis of melanoma.

We have presented a case of Kawamura type II blue nevus, highlighting the need to perform periodic clinical control of pigmented lesions and suggesting the possibility of monitoring this type of lesion using digital dermoscopy.

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- D. Ayala,<sup>a,\*</sup> M.D. Ramón,<sup>a</sup> M. Cabezas,<sup>b</sup> E. Jordá<sup>a</sup>
- <sup>a</sup> Servicio de Dermatología, Hospital Clínico Universitario de Valencia, Valencia, Spain
- <sup>b</sup> Servicio de Anatomía Patológica, Hospital Clínico Universitario de Valencia, Valencia, Spain

\* Corresponding author.

E-mail address: dayalca83@hotmail.com (D. Ayala).

## Initial Evaluation of Patients with Pigmented Skin Lesions<sup>☆</sup>



## Valoración inicial del paciente con lesiones cutáneas pigmentadas

To the Editor:

The incidence of melanoma has increased significantly in Spain in recent decades, and this tumor is now a public health problem.<sup>1</sup> The role of the dermatologist is fundamental to the identification of high-risk patients and to the adoption of appropriate primary and secondary preventive measures for the early detection of skin cancer.<sup>1–3</sup>

Several well-standardized international clinical guidelines have been drawn up on the management of patients with melanoma,<sup>4–9</sup> but no protocols exist on how to take an appropriate medical history and physical examination. The Australian guideline proposes an initial evaluation to determine the future risk of melanoma (grade B recommendation) looking at various factors, including age, sex, past history of melanoma or nonmelanoma skin cancer, family history of melanoma, number of melanocytic and atypical nevi, skin and hair color, skin phototype, and actinic damage.<sup>4</sup>

When a patient attends dermatology outpatients for an initial clinical evaluation of a nevus or suspected melanoma,

an exhaustive medical history must be taken to detect the main risk factors (Table 1) and a complete physical examination performed.<sup>1–3</sup> In the literature, we have found no descriptions of a protocol for the initial clinical evaluation of this type of patient. We therefore present our standard approach to the first consultation in the melanoma unit of Hospital Clínic in Barcelona:

- Personal history of drug allergy, drinking and smoking, current and previous occupations, known diseases, surgical history, noncutaneous tumors, and usual medical treatments.
- Past dermatologic history: known dermatoses, history of skin tumors, dysplastic nevus syndrome, treatments performed, previously excised nevi, and risk classification (Table 2).
- Family history of dysplastic nevus syndrome and of skin and other tumors.
- History of sun exposure, UV protection (Table 3), pattern of intermittent or chronic exposure, and time dedicated to outdoor occupational or leisure activities.
- Detailed history of the suspected melanoma lesion<sup>3,10</sup>: site, time since onset, presence of a precursor lesion, symptom or sign that prompted consultation, pruritus, pain, bleeding, erosion, ulceration, suppuration, changes in color (multiple shades of dark brown or black, or appearance of various colors, including light brown, dark brown, black, red, blue, gray, and white), changes in morphology (appearance of irregular margins), change in size (rapid or continuous growth), elevation of the lesion, changes in the surrounding skin (erythema, edema,

<sup>☆</sup> Please cite this article as: Imbernón-Moya A, Podlipnik S, Malvehy J, Puig S. Valoración inicial del paciente con lesiones cutáneas pigmentadas. *Actas Dermosifiliogr*. 2016;107:616–618.

**Table 1** Main Risk Factors for Melanoma.

Phenotype	White race Fitzpatrick phototypes I and II Advanced age (> 65 years) Male sex Fair or red hair Blue or green eyes
External factors	Prolonged and intense sun exposure Sunburn in childhood Artificial UV-A tanning beds Residence in equatorial countries
Past history	Past history of melanoma and/or nonmelanoma skin cancer (basal cell carcinoma, squamous cell carcinoma, actinic keratosis) History of surgical excision of a dysplastic nevus Parkinson disease Acquired or iatrogenic immunosuppression (solid-organ or hematologic tumors, transplant recipients, immunosuppressant treatments, HIV infection) Albinism
Genetic syndromes	Xeroderma pigmentosum
Family history	First or second degree relative with a history of melanoma, dysplastic nevus syndrome, or other melanoma-associated tumor (pancreas, renal, breast, lung, colon, central nervous system)
Clinical characteristics of the patient	More than 100 clinically typical or more than 5 clinically atypical acquired melanocytic nevi (ABCDE rule). Dysplastic nevus syndrome (more than 100 acquired melanocytic nevi with 1 or more larger than 6 mm and 1 or more with histology of dysplastic nevus), multiple freckles or solar lentigines Giant congenital melanocytic nevus (> 20 cm) Mutations in genes <i>BRAF</i> , <i>MITF</i> , <i>BAP1</i> , <i>CDK4</i> , <i>CDKN2A</i> . <i>MC1R</i> gene polymorphism
Genetic factors	

**Table 2** Classification of Risk of Melanoma.

0	No history of melanoma
1	Personal past history of melanoma
2	First or second degree relative with melanoma
3	Personal past history of 2 or more melanomas or personal or family history of melanoma

**Table 3** Clinical Data to Be Considered in the History of Sun Exposure.

<i>Cumulative sun exposure:</i>	<10 y	10-18 y	>18 y	Current
Intense (> 120 h/y)				
Moderate (50-120 h/y)				
Mild (< 50 h/y)				
None (0 h/y)				
<i>Adequate sun protection:</i>	<10 y	10-18 y	>18 y	Current
Always				
Usually				
Occasionally				
Never/hardly ever				
<i>Level of exposure to artificial UV:</i>	UV-A	PUV-A	UV-AB	
Intense (> 40 h/y)				
Moderate (20-40 h/y)				
Mild (< 20 h/y)				
<i>Sunburn/UV radiation burn:</i> (E= erythema. B= blisters)	<10 y	10-18 y	>18 y	Current
> 5 episodes/year				
3-5 episodes/year				
1-3 episodes/year				
None				

satellite pigmentation), changes in consistency (softening, friability), and previous treatment of the lesion.

- Systems review: constitutional, neurologic, otorhinolaryngologic, respiratory, musculoskeletal, hepatic, gastrointestinal, cutaneous, subcutaneous, and lymphatic signs and symptoms.

We also present our standard procedure for complete physical examination:

- Skin phototype (I-VI),<sup>2,3</sup> race (white, black, yellow), eye color (green, blue, gray, brown, black), hair color at 18 years of age (red, fair, brown, black).
- Complete dermatologic examination, including scalp, intertriginous areas, oral, ocular, genital, and anal mucosas, palms, soles, and nails.
- Presence of freckles, solar elastosis, solar lentigines in sun-exposed areas, actinic keratoses, basal cell carcinoma, and squamous cell carcinoma.
- Estimation of the total number of acquired melanocytic nevi, whatever their size (< 50, 50-100, 100-200, > 200). Evaluation of the number of these lesions and of the clinical type (junctional, compound, intradermal) and size (small, 2-6 mm; intermediate, 6-15 mm; or large, > 15 mm) on the trunk and on the upper limbs. Evaluation of the number of clinically atypical nevi and presence of congenital nevi.
- Description of the suspected lesion: site, diameter, palpation, presence of papules or nodules, size, pigmentation, clinical type, ulceration, areas of regression, adjacent nevi. Physical examination of the regional lymph nodes, presence of hepatomegaly, splenomegaly, and locoregional and/or distant subcutaneous masses or nodules.

- Dermoscopic features.
- Photographs of the lesions prior to the surgical excision of suspicious skin lesions (essential).

In conclusion, the objective of this article has been to present the guideline used in our center for the initial clinical evaluation of nevi or suspected melanoma in the hope that it will help dermatologists evaluating patients with such lesions. Finally, it is essential that we undertake patient education regarding periodic self-examination, the clinical characteristics of melanoma (ABCDE rule and ugly duckling sign), and appropriate measures of photoprotection.<sup>1-3,10</sup>

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- A. Imbernón-Moya,<sup>a,\*</sup> S. Podlipnik,<sup>b</sup> J. Malvehy,<sup>b</sup> S. Puig<sup>b</sup>
- <sup>a</sup> Servicio de Dermatología, Hospital Severo Ochoa, Leganés, Madrid, Spain
- <sup>b</sup> Servicio de Dermatología, Hospital Clínic de Barcelona, Barcelona, Spain
- \* Corresponding author.  
E-mail address: adrian\_imber88@hotmail.com  
(A. Imbernón-Moya).

## Bednar Tumor (Pigmented Dermatofibrosarcoma Protuberans)<sup>☆</sup>



### Tumor de Bednar (dermatofibrosarcoma protuberans pigmentado)

To the Editor:

Dermatofibrosarcoma protuberans (DFSP) is a dermal connective tissue tumor with low malignancy due to its slow growth and locally aggressive nature. It can be classified into several variants according to morphologic features, although there are no major differences in terms of prognosis. The pigmented variant of DFSP, also known as Bednar tumor,<sup>1,2</sup> is rare and is characterized by the presence of fibroblasts interlaced with melanin-containing dendritic cells. As with other variants of DFSP, fibrosarcomatous changes may occur; these are characterized by CD34 negativity, scarce melanin pigmentation, and increased cell prolifer-

ation and pleomorphism.<sup>3,4</sup> This transformation tends to occur in recurrent DFSP and is associated with poor prognosis and an increased risk of metastasis, although the real risk is low.

The identification of the translocation t(17;22)(q22;q13) and the detection of the COL1A1-PDGFB fusion protein in DFSP led to the synthesis of tyrosine kinase inhibitors, such as imatinib as an alternative treatment for unresectable locally advanced disease or metastatic disease<sup>5</sup> and sunitinib for nonresponders to imatinib.<sup>6</sup>

We report the case of a 93-year-old man with a pigmented stain, part of which contained a firm nodular lesion of 1.5 cm, located in the left preauricular region (Fig. 1). On observing that the lesion had grown, and based on a clinical suspicion of melanoma on lentigo maligna, the decision was taken to completely excise it. The histopathologic study showed focal atrophy of the epidermis, without ulceration, and a dense proliferation of monomorphic spindle cells with variable pleomorphism throughout the dermis and extending into the subcutaneous tissue (Fig. 2). The spindle cells presented areas with a histiocytic/xanthomatous appearance, abundant melanin pigmentation, and isolated mitotic figures (Fig. 3A). In the immunohistochemical study, the tumor cells showed diffuse positive staining for vimentin, focal staining for CD34 and CD68, and negative staining for

☆ Please cite this article as: Añón-Requena MJ, Pico-Valimaña M, Muñoz-Arias G. Tumor de Bednar (dermatofibrosarcoma protuberans pigmentado). *Actas Dermosifiliogr.* 2016;107:618-620.