On Pigmented Macules and Butterfly Collection

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When we propose the individualization of a new disease, we should always be very careful; first of all, ask whether it is really something new and whether it has already been observed.

Furthermore, ask the question: Is it solely a trivial addition to what my old Master Pautrier called, with raillery, a 'butterfly collection'? We need to assess how useful the naming of a new disease is in solving the clinical problems and, above of all, to believe that this will remain true in years to come. Particularly, if similar or comparable observations are subsequently made by others, one can be convinced that the earlier study was not completely useless.

As A. Dupré points out in this issue, the etiology of the pigmented macules in question is not entirely known. Stable and without evidence of degeneration, they are not of racial origin and are not induced by drugs.

In our original report, the first patient was a 60-year-old Franc-comtoise woman who was in the hospital for bronchitis. Pigmentations of the oral mucous membrane and lips, strictly lenticular, were seen during systemic investigation. The second patient, seen at Geneva with N. Hunziker, was a British woman who consulted us for some important pigmented lesions, especially those on the lips. Facing her strong request for surgical ablation, we made a large excision of the most important macule on the lower lip. This provided us with the material for the histopathologic study and the ultrastructural investigation kindly made by Dr. Cesarini. Shortly after that, and to our surprise, we encountered another patient who had pigmented macules on the lips and mouth, as well as linear melanonychia on one or several nails. A few weeks later, Pellerat et al. [1] presented a similar case. At a later

period, during the 'patient presentation' at the Clinique de Dermatologie de Strasbourg, the debate was focused on a patient with a longitudinal pigmentation of one nail. The point in question was: Is it a malignant melanoma? The examination of the oral mucous membrane showed many pigmented macules. In our opinion, these were helpful clinical clues and permitted us to eliminate the diagnosis of melanoma.

Since then, the knowledge we learned from these pigmented lesions has allowed us to diagnose them as benign lesions and to assure the patients and their physicians. That is also the case for the genital pigmented macules of the vulva [2] or glans penis [3]. They can be isolated genital lesions, associated with oral macules as well as cutaneous lesions as observed by Sartoris et al. [4] and frequently found in patients of A. Dupré (this issue).

In what framework do we classify these epidermal hypermelanoses? Some authors consider hyperactivity of melanocytes, but what is the etiology? Ultrastructurally, we observed either small melanosomes in clusters or large-shaped melanosomes, but always without abnormality. We also think that an enzymatic hyperactivity in tyrosine-melanin biosynthesis is possible, but once again, it is not possible to explain the pathogenesis.

Thus, when patients are concerned about these pigmented macules, strictly for cosmetic reasons, one can confidently proceed to the surgical removal and prove its benignity by histologic study. When the lesions are discovered incidently, we can assure ourselves as well as the patient that luckily these are benign macules; perhaps this is the reason why this syndrome is more than just another 'butterfly' in the collection.

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Editorial

Acknowledgement

I thank Prof. Hu for his help in the writing of this editorial.

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