Giant Nevus as an Aberrant Form of von Recklinghausen Neurofibromatosis

Sculptor: José Barta, Clínica del Dr Sáinz de Aja.
Comment*

At first glance, this model appears to represent a giant melanocytic nevus ("bathing suit" nevus), on which tumors like the one visible in the lumbar region may develop. These tumors may be histologically indistinguishable from neurofibromas, even though the patient may not have neurofibromatosis.

In any case, it is also known that there is a relationship between type 1 neurofibromatosis and giant melanocytic nevi. In fact, von Recklinghausen had already called attention to this association in 1 of the 28 patients he described in his monograph on the disease that bears his name, an association later confirmed in other, more extensive series of type 1 neurofibromatosis cases. Therefore, there exist both giant melanocytic nevi that develop neurofibromas and type 1 neurofibromatosis accompanied by giant melanocytic nevi. Which is the case for this model?

We believe that Dr Sáinz de Aja, who studied this case and ordered the sculpture, would have had evidence now unavailable to us on which to base his argument that this patient presented neurofibromatosis, and not simply this melanocytic nevus and the kind of tumors that sometimes accompany it. It seems clear that this is a congenital melanocytic nevus with multiple proximal lesions and paravertebral nodular masses, but comparison with classic neurofibromatosis is not unreasonable, since some plexiform neurofibromas have a similar appearance.

Between 1925 and 1936, a dermatological journal of considerable originality entitled Ecos Españoles de Dermatología y Sifiligráfía (Spanish Echoes of Dermatology and Venereology) was published. Among its stated aims was the publication of a small atlas of the wax models of the Olavide Museum; a photogravure of this very molding was published in the second issue of the journal in November 1925.

*Since this is a figure sculpted by J Barta, it lacks a clinical history, unlike the models made by E Zofío, which are all accompanied by the corresponding case histories.

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