SUBACUTE SCLEROSING PANENCEPHALITIS

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SUBACUTE SCLEROSING PANENCEPHALITIS

A Reappraisal

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Subacute sclerosing panencephalitis: A reappraisal
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SUBACUTE SCLEROSING LEUCOENCEPHALITIS

Dr F.J. RADERMECKER

In our monography from 1956 " Systématique et E.E.G. des Encéphalites et Encéphalopathies " , we drew attention to the fact that Dawson, first in 1933, and later in 1934, mentioned two similar observations which he described as a " specific type of encephalitis ", to be distinguished from the group of epidemic encephalitis. As a matter of fact at that particular time, distinction was made between two types of encephalitis: lethargic encephalitis type A, as described by von Economo, and type B or Japanese encephalitis.

In the course of the year 1939, L. van Bogaert and J. De Busscher reported an anatomoclinical observation with the following characteristics: " A female child, untill then normally healthy, shows two months after an appendectomy, an abrupt decrease in its intellectual performance at school, together with behavioural disorders. The learning capacities at school show a rapid decline while at the same time nocturnal hallucinations start to appear. She becomes coarse and brutal. Later on difficulties in writing and in visual perception develop as well as a tendency to aphasia and apraxia. Eight months after the onset, an objective clinical examination shows crises of muscle rigidity and opisthotonos with conjugated deviation of the eyes, optic hallucinations and paroxystic crises of cephalea. The child succumbs and a histologic examination shows evidence of an inflammatory process, predominantly in the white matter and to a lesser degree in the grey matter.

From that time on, L. van Bogaert, in the Journal of Neurology (1945) as a result of three new observations, for the first time, stresses the fact that in this case we are confronted with either an authentic illness or a reactive disease of the allergic type.

The whole image is dominated by axial inflammation which deserves a separate mention because of its sclerosing properties and its particular pattern of extension. The initial phase of the disease is characterized by a plasmocytary and micro-oligoglial infiltration of the white matter, with a high density level of the subcortical regions. It is accompanied by perivascular plasmolymphocytary infiltrations and by a gliofibrillary structure,

striking not only by its early onset, but also by the fact that it surpasses amply the needs of restauration, since there is no demyelination, nor necrosis or oedema.

As for the type of extension, the primary cortical involvment (dominated by an infiltration of rod cells and plasmolymphocytary perivascular reactions), is given a particular individuality with regard to the other diffuse scleroses, by localizations in the extracortical grey matter of the thelamostriated mesencephalic and bulbar regions.

Without running ahead of things, L. van Bogaert concludes as follows: " We believe that these observations truly represent a cerebral inflammatory disease which, from a histological point of view, displays a subacute sclerosing leucoencephalitis, with little or no demyelination."

Of all things, L. van Bogaert wanted to distinguish subacute sclerosing leucoencephalitis from diffuse multi-lobular sclerosis (or Schilder's encephalitis), and multiple sclerosis. This initial observation made in 1939, was published under the name of " sclérose diffuse inflammatoire de la substance blanche des hémisphères " (diffuse inflammatory sclerosis of the white matter of the hemispheres).

This denomination giving rise to confusion, L. van Bogaert from 1945 onwards, substituted the name of subacute sclerosing leucoencephalitis, clearly stressing its inflammatory properties, its axial predilection, its early sclerotic organization and the subacute character of its evolution.

To L. van Bogaert, this axial component together with the grey matter component, which he stressed from the beginning, constitutes the most particular element of the disease. The axial involvment on the one hand, and the constant clinical features on the other hand have, at the time of the first descriptions, postponed the idea of an interrelation between these cases, viz. of Pette and of Döring on the one side, and Dawson's on the other.

Especially in Dawson's encephalitis, the pathologists have been mesmerized by the presence of the inclusions, at the expense of the general pathological process. The personal contribution of van Bogaert has been to demonstrate the general processus of axial sclerosis. The American authors who did not completely agree on this matter, have changed the original name of subacute sclerosing