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Baron Constantin von Economo and encephalitis lethargica

Although younger neurologists may not have encountered postencephalitic parkinsonism, encephalitis lethargica merits mention in contemporary reviews of Parkinson's disease. The name of von Economo is inseparably and justifiably attached to the disease since his was the major description¹ of an illness that raged in epidemic form in Europe and North America between 1916 and 1926. Ivy McKenzie provided a scholarly account of the illness in Glasgow.² Longer texts were written by Jelliffe,³ and by Wimmer.⁴ At first, others confused encephalitis lethargica with the pandemic of influenza (Spanish gripe). Cruchet had first noted encephalitis lethargica in the winter of 1915-16 in French soldiers in Verdun,⁵ and a few cases were seen in the spring of 1915 in Rumania.

von Economo published 27 papers including a book⁶ on encephalitis lethargica.

Excerpts from a translation of his classic description¹

"We are dealing with a kind of sleeping sickness, having an unusually prolonged course. The first symptoms are usually acute, with headaches and malaise. Then a state of somnolence appears, often associated with active delirium from which the patient can be awakened easily. He is able to give appropriate answers, and to comprehend the situation. This delirious somnolence can lead to death, rapidly, or over the course of a few weeks. On the other hand it can persist unchanged for weeks or even months with periods lasting hours or days or even longer, of fluctuation of the depth of unconsciousness extending from simple sleepiness to deepest stupor or coma During the first days of the illness, isolated signs of meningeal irritation appear The appearance of fever and its intensity do not seem to have any effect upon the course and signs of the disease

As a rule these general symptoms are joined by paralysis of the cranial nerves as well as in the extremities . . . a paralytic ptosis often combined with partial or total paralysis of other branches of the oculomotor nerve Paresis of the other cranial nerves and paralysis of the extremities with reflex disturbances can occur also"

[seven case reports and an account of the clinical features are then given]

"The spinal fluid . . . showed increased pressure at the beginning; it later decreased in spite of persistent somnolence. The total protein was below the normal upper limit; . . . In case 1, 43 cells/mm³, in case 2, 19 cells/mm³ . . . strong predominance of polymorphonuclear leukocytes. Repeated examina-

tions for microorganisms was negative in every case. The Wassermann reaction in serum and spinal fluid was always negative

As the residual of such a paresis (upper and lower extremities), spasms, increased reflexes, or the Babinski phenomenon can persist for a long time, even after the ocular muscle paralysis and somnolence have receded . . . one often finds a peculiar rigidity of the extremities (cases 1, 2 and 4). Like most of the signs this rigidity disappears later We frequently find (cases 1, 2, 3, 4, 7) ataxic disturbances that make one think of involvement of the cerebellum. Case 7 demonstrated severe ataxic tremor With reference to pontine encephalitis I describe briefly several cases seen in the emergency room during the same period"

[six further case reports and a long discussion on differential diagnosis and pathology follow]

von Economo concludes: "Therefore we have the histological picture of a *polioencephalitis cerebri, pontis et medullae oblongatae, with a slight poliomyelitis of a perivascular, inflammatory and diffusely infiltrative but not haemorrhagic and only slightly neuronophagic character.*"

Born in Braila, Rumania, of an aristocratic Greek family he studied engineering before reading medicine in Vienna. Early training was with Nothnagel, Pierre Marie, and Kraepelin before he returned to the psychiatric clinic in Vienna.

His work centred on neuroanatomy, head injuries in the first world war, and investigations on sleep, cortical architecture, and on Wilson's disease.

He married the daughter of Prince Alois von Schönburg-Hartenstein, and being of independent means, rejected the chair of psychiatry on von Jauregg's retirement in 1928. He died suddenly of heart disease in 1931.

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