Bielschowsky Bodies: Lafora-Like Inclusions Associated with Atrophy of the Lateral Pallidum

Guillermo A. de León
Division of Neuropathology, Department of Pathology,
Temple University Health Sciences Center, Philadelphia, Pennsylvania

Received June 25, 1974; Accepted July 25, 1974

Summary. Peculiar amylaceous bodies were found within the lateral pallidum of the brain of a 93 year old patient with an athetoid form of cerebral palsy. The bodies were numerous and mostly confined to the neuropile. Occasional bodies were seen within the perikarya of pallidal neurons; many were shown inside nerve processes. The shape of the bodies was quite variable; some were round, ovoid or pear-shaped; others were elongated in the shape of sausages, rods or threads of variable width and length. They tended to form chains which often branched along nerve processes. In paraaffin embedded sections, the bodies presented a dense central core, and a paler peripheral zone which exhibited a characteristic foamy appearance. They seem identical to bodies previously described by Bielschowsky (1912) and Vanderhaeghen et al. (1967). Their nature and staining properties are very similar to those of Lafora bodies and corpora amylacea. However, in view of their location and striking morphologic characteristics they should be considered as a distinct type of structure. It is fair and convenient to name them after Bielschowsky, their original discoverer.

Key words: Athetosis — Bielschowsky Bodies — Corpora Amylacea — Lafora Bodies — Pallidal Atrophy.

Introduction

In 1912, Bielschowsky described some peculiar bodies within the lateral pallidum in a case of double athetosis. That case had been studied during life at the clinic of Barré in Paris, and the brain was made available to C. Vogt (1911) who briefly reported it as an example of état marbré. The important abnormalities in that brain were apparently limited to the basal ganglia which were atrophic and showed marbling of the putamen. In a small area of the lateral pallidum, near the external medullary lamina, Bielschowsky found amylaceous bodies which he identified as the just described Lafora bodies (Lafora, 1911; Lafora and Glueck, 1911).

Vanderhaeghen et al. (1967) described two cases of torsion dystonia and choreoathetosis in which the pertinent pathologic findings were limited to the lateral pallidum. In addition to nerve cell loss, numerous amylaceous bodies similar to those described by Bielschowsky were found in both cases. The bodies were also identified as Lafora bodies and the cases were otherwise compared to those of pure pallidal atrophy (van Bogaert, 1946).

A case of athetoid cerebral palsy will be described here. When the patient was 17 years old her condition was reported by Preston (1896) as an example of “congenital chorea”. In the atrophic basal ganglia, bodies of the type described by Bielschowsky (1912) and Vanderhaeghen et al. (1967), were found in the lateral
pallida. This finding will be specifically emphasized because it seems that these bodies are clearly distinguishable from Lafora bodies, corpora amylosea, and other similar structures.

Case Report

Anamnesis. This female patient died at age 93. She was born postmaturely by footling presentation. Her mother complained that fetal movements had been weaker than with previous pregnancies. There was no breathing impairment at birth but the baby presented extreme flaccidity and feeding difficulties. “Twitching” or “choreic” movements were noted since early life. Development of motor abilities was markedly delayed. She sat up and held up her head at five years. She was finally able to walk without support but stopped doing it after a fall in which she injured her hip. Thereafter, she learned to ride a special tricycle, and this became her principal means of locomotion. She never had articulate speech. In contrast with this, her intellectual development was far more advanced. She learned to communicate with sign language and to type with one finger. She read a lot, was remarkably well-informed, and became a sensitive writer. On examination, she was of short stature and presented continuous athetoid movements of face, tongue, and jaw, as well as arms and legs. During the last years of her life she was generally weak, particularly in extensor muscles, and developed flexor contractures of her legs and tremor of her arms.

Gross Brain Findings. The formalin fixed brain weighed 825 gm. Both cerebral hemispheres were small. The cerebellum was also small but proportionately to a lesser degree than the cerebrum. The cerebral vessels showed moderate atherosclerosis. On sections, the lateral and third ventricles were symmetrically enlarged; the cerebral cortex was thinned; the white matter was decreased in volume and firm. The thalami were pale and small. The basal ganglia were decreased in size; the medullary laminae, as well as the myelinated bundles of the lentiform nuclei, were indistinct. A few small lacunes were found in these grisea. The substantia nigra appeared normally pigmented; the bases pedunculi were narrow. The cortex and white matter of the cerebellar vermis were thinned and sclerotic.

Histological Findings

The basal ganglia were much reduced in size. The thinned medullary laminae, as well as the bundles of the striatum and the fibers of the pallidum, were poorly myelinated. The head of the caudate nucleus was somewhat shrunken. In the posterior third of the putamen a mesh of numerous myelinated abnormal fibers gave the nucleus the typical appearance of marbling. There was at this level moderately severe neuronal loss and gliosis. A few small lacunar infarctions were present in the putamen and caudate nucleus. The most striking finding in the basal ganglia was the presence of numerous peculiar bodies of variable size and shape which were disseminated throughout the lateral pallida of both cerebral hemispheres. These bodies will be referred to as Bielschowasky bodies. As these bodies were considerably more numerous than the neurons, they dominated the microscopic appearance of the pallidum. They were equally abundant at all levels of the lateral pallidum, but only rare bodies were present in the putamen and medial pallidum. Only a few were contained within the perikaryon of pallidal neurons. However, in section stained with Palmgren’s silver methods, many bodies were seen to be inside distended nerve processes. Bielschowasky bodies were strongly argyrophilic when stained with Gomori’s methenamine silver, and were also deeply stained with Alcian blue, PAS, iodine, and Best’s carmine. They gave negative reactions for amyloid, iron and calcium. Their shape was quite variable. Some were round, ovoid or pear-shaped (Fig. 1); others were elongated, sausage-like, or appeared as rods or threads. They were found either singly or grouped in chains, some stretching for up to about 160 microns in length (Fig. 2). Some of these chains were formed by bodies of tapering length and/or width; many chains presented dichotomies. In sections stained with silver, a sheath could be seen surrounding many of the chains or single bodies. At the end of the bodies the edges of such sheaths were often seen to come in contact and merge into a nerve process, revealing the intracellular nature of the bodies (Fig. 3). The smaller globoid bodies measured no more than 1 – 2 microns in diameter. Similarly, the thinner rod-like bodies were no more than 1 micron in width. The larger round bodies measured...