Ganglioneuroblastoma of the Cerebellum
Case Report

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Neuroblastomas and ganglieneuromas of adrenal or sympathetic origin are
common tumors. Olfactory neuroblastomas, presumed to arise from the neuro-
epithelial cells of the nasal mucosa, are being diagnosed with increasing frequency. By con-
trast, nerve cell neoplasms of the brain, if one excludes the medulloblastomas, are ex-
tremely rare. Indeed, Willis’ questions their very existence. This report describes a tumor,
composed of primitive neuroblasts and mature neurones, which developed in the cere-
bellum of a child.

Case Report

A 3-year-old girl was admitted to the hospital on October 19, 1966. At 1½ years of age, she
had had four episodes of awakening at night and screaming for a few minutes over a period
of 2 months. At 2½ years, these attacks recurred, and the child became lethargic and irritable. She developed vomiting and occipital headaches, and later incoordination and a
tilt of the head to the right. She was admitted when the vomiting became almost unmanage-
able.

Examination. The patient was hypertonic and hyperreflexic and had bilateral Babinski
responses. Motor power was good and sensation intact. She had marked dysmetria and in-
tention tremor of all limbs. The gait was ataxic and she could neither stand still nor sit
upright without support. Bilateral papilledema was present. Radiographs of the skull re-
vealed separated sutures, and a ventriculogram confirmed the presence of a posterior fossa
tumor.

Operation. Suboccipital craniotomy showed an encapsulated tumor in the right cerebellar
hemisphere which was removed without difficulty. Following this, the skull and spinal axis
were irradiated (tumor dose, 3,500 rads). An intravenous pyelogram was performed post-
operatively and was entirely normal; a 24-
hour urine catecholamine excretion was also
within normal limits (27 mg/24 hrs).

Postoperative Course. Immediately after the operation the child was irritable, but at the
time of discharge on November 25, 1966, she was tractable, could walk without help, and
had better coordination of the limbs. This improvement was continuing at the time of this
report.

Pathological Specimen. The tumor weighed 61 gm, measured 6.8 cm in diameter, had a
roughly spherical, irregular, lobulated surface, and appeared encapsulated. It was rubbery
and, on sectioning, homogeneously pinkish-grey with a few scattered white flecks.

Microscopic Features. Multiple areas of the
tumor were sampled and stained with hematoxylin and eosin, cresyl violet, Luxol-fast-
blue-Holmes’ silver carbonate for axis cylinders and myelin sheaths, Bielschowsky and
reticulin stains. Two distinct neoplastic elements, one mature, the other immature, were
noted. Although they were occasionally mixed with each other, in general they existed in
microscopically discrete areas, and usually their borders were quite sharp.

The mature foci consisted of clearly recog-
nizable, albeit abnormal, adult nerve cells with a vesicular, often eccentric nucleus, a promi-
nent nucleolus, and Nissl substance at the periphery of the cytoplasm, confirmed by
cresyl violet stains (Fig. 1). Binucleate and, occasionally trinucleate forms, were not in-
frequent. An occasional mitosis was noted in
the adult cells (Fig. 2). The cells lay in hap-
hazard fashion, without orderly orientation. Abundant axons lay between the cells (Fig. 3).
These were unmyelinated and were undoubt-
edly processes of the neoplastic neurones. There were also foci of calcification.

The immature portions were densely cellu-
lar and consisted of regular round to oval cells
with little or no cytoplasm, and somewhat
hyperchromatic nuclei, with fairly frequent mitotic figures (Fig. 4). The cells showed a
tendency to be gathered into islands (Fig. 5),

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Fig. 1. Fully and partly mature nerve cells, some with peripheral Nissl substance, other multinucleate. H. & E., ×820.

separated by a reticulum, and there was invasion of the subarachnoid space in some areas. There was often a close resemblance to a medulloblastoma (a differentiation that is not always easy).  

Intermediate forms between neuroblasts and mature neurones were present. These were recognizable as nerve cells, but were smaller, lacked Nissl substance, and sometimes cytoplasm (Fig. 1).

Cresyl violet and Holmes’ stains quite clearly demonstrated the presence of Nissl substance only in the differentiated cells, and of axis cylinders only in the mature foci. Bielschowsky stains demonstrated processes arising from mature cells only. A few astrocytes and oligodendrogial cells were present, but did not appear to participate in the tumor formation.

Discussion

The early literature on central nerve cell tumors is difficult to survey. Not only was a wide variety of names used to describe similar and dissimilar tumors, but often normal nerve cells surviving in an infiltrating glioma were interpreted as being a part of the tumor, and the so-called “ganglioid” astrocytes were erroneously thought to be neo-

Fig. 2. Mitosis in a mature nerve cell. H. & E., ×820

Fig. 3. Unmyelinated axons lying between neurons. The large dark areas represent calcium. Luxol-fast-blue-Holmes’ silver stain, ×820.