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Polar spongioblastoma: an immunohistochemical and electron microscopical study

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Summary. A case is reported of a 9-year-old boy with a cerebral polar spongioblastoma. This neoplasm, first described by Russell and Cairns in 1947, is morphologically a distinct entity characterized by bipolar tumor cells with palisading nuclei. In the case under study immunoreactivity for neuron-specific enolase was found and ultrastructural features of developing neuronal elements were present. A neuro-endocrine nature was suggested by de Chadarévian et al. (1984) in a morphologically similar case. These findings are in contrast with the long-held view that the polar spongioblastoma is cytogenetically related to the embryonal radial glial cells.

Key words: Cerebral tumor – Polar spongioblastoma – Astrocytoma – Immunohistochemistry – Electron microscopy

The term polar spongioblastoma is used to designate two different tumor types. One of these, described by Bailey and Cushing [1], is at present more commonly known as the pilocytic astrocytoma of cerebellum, brain stem and optic nerve. The other tumor is a histopathological entity first described by Russel and Cairns [8] and characterized by bipolar cells with a parallel arrangement of cellular processes and palisading of nuclei. So far 11 cases of this latter tumor type have been reported. Morphologically this "true" polar spongioblastoma seems to be related to the embryonal spongioblasts or radial glia [7], but ultrastructural details of a case presented by de Chadarévian et al. [3] suggest a neuro-endocrine origin. We present immunohistochemical and ultrastructural data of a new case.

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Case report

A 9-year-old boy was referred to the Neurosurgical unit of the Academic Medical Center in Amsterdam. The boy had a 2-year history of frequent epileptic attacks, and more recently showed behavioral changes and forgetfulness. One year previously a small, hypodense lesion was present in the left frontal lobe on CT scanning, although neurological examination had revealed no signs of raised intracranial pressure, or motor or sensory disturbances. Renewed CT scanning showed that the hypodense lesion had grown and calcifications were present (Fig. 1). The patient underwent surgical resection of the lesion. Presently, 1 year later, the patient is doing well.

Materials and methods

For histology, paraffin-embedded sections (6 μm) were stained with hematoxylin and eosin (H&E), phosphotungstic acid hematoxylin (PTAH), van Gieson, Gomori's reticulin and Grimelius. For the immunohistochemical demonstration of cellular antigens, 5-μm paraffin slides were stained with either a three-step indirect immunoperoxidase procedure, a peroxidase-antiperoxidase procedure, or the avidin-biotin labeling method (antisera, suppliers, staining method used and results see Table 1). For electron microscopy surgically obtained tissue was fixed in Karnovsky medium and processed according to routine electron microscopical procedures.

Results

Histology

The tumor, which had a soft, grayish appearance, is composed of two different parts. The largest area shows bipolar cells aligned in parallel with their ovoid, monomorphic nuclei in palisading rows. This part is bordered by numerous proliferating small blood vessels. The processes of the tumor cells are directed to thin-walled capil-

Table 1. Summary of antisera, suppliers, staining methods used and results

Antisera raised against	Supplier	Method	Result
Glial fibrillary acidic protein (polyclonal)	Dakopatts Z334	PAP	–
Glial fibrillary acidic protein (monoclonal)	Dakopatts M761	3 step	–
S-100 protein	Dakopatts Z311	ABC	+++
Neuron-specific enolase	Dakopatts A589	ABC	+
Neurofilament – 68 kDa, non-phosphorylated	Boehringer 326	3 step	–
Neurofilament – 168 kDa, non-phosphorylated	Boehringer 334	3 step	–
Neurofilament – 200 kDa, non-phosphorylated	Boehringer 342	3 step	–
Neurofilament – 200 kDa, phosphorylated	Eurodiagnostics	3 step	–
Synaptofysin	Dakopatts M776	ABC	–
Vimentin	Dakopatts M725	ABC	Focal +
Desmin (polyclonal)	Eurodiagnostics	ABC	–
Desmin (monoclonal)	Monosan	ABC	–
Chromogranin	Camon E001	ABC	–
Growth hormone	Dakopatts A570	ABC	–

PAP: Peroxidase-antiperoxidase; 3 Step: three step indirect; ABC: avidin-biotin complex method; – : immunoreactivity absent; + : reactivity present; +++: strongly immunoreactive

Table 2. Summary of polar spongioblastoma cases

Reference	Sex	Age (years)	Localization	Therapy	Survival (years)
Cairns and Russell [2]	f	9	3rd ventricle	S + RT	0.3
Mittelbach [5]	f	8	Cerebellum	S + RT	0.3
Russell and Cairns [8]	m	15	Mesencephalon	–	1
	m	1.3	4th ventricle	–	1.2
	f	13	4th ventricle	–	1.5
Slooff et al. [11]	m	46	Spinal cord	–	0.5
de Chadarévian et al. [3]	f	5	Diencephalon	S + RT	> 4
Schochet et al. [10]	f	11.5	Spinal cord	S + RT	> 2.5
Steinberg et al. [12]	m	0.5	Cerebellum	S + RT	> 14
Present case	m	7	Frontal lobe	S	> 1

S: surgery; RT: radiotherapy; – : no therapy

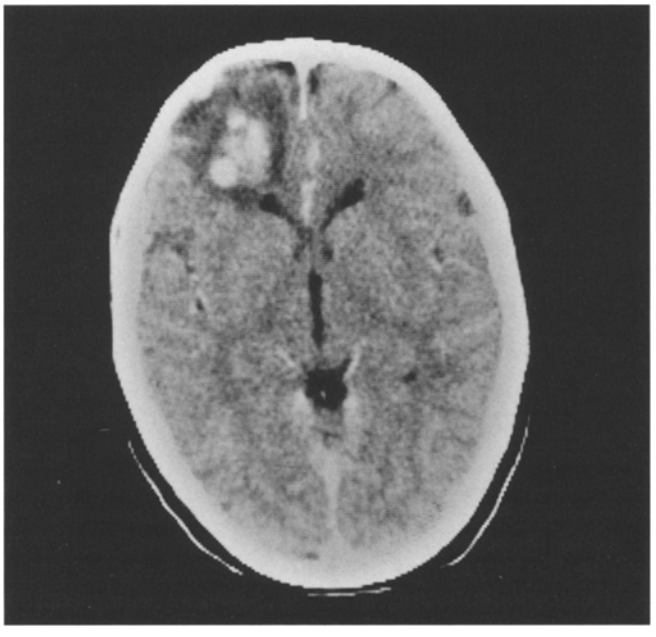

Fig. 1. CT scan: left frontal hypodense mass with calcifications

laries, which form the tumor stroma. These cellular processes are PTAH positive. Rosette formation is not present (Fig. 2a, b). A small part of the tumor is composed of astrocytic cells and granular bodies. Many calcospherities are embedded in the tumor. No mitoses or necrotic tumor tissue is present.

Immunohistochemistry

Of the antigens listed in Table 1, the typical tumor cells show a positivity for S-100 protein and a mild reactivity for neuron-specific enolase (NSE). Vimentin reactivity is only focally present in cellular processes (Fig. 2c). There is no immunoreactivity for neurofilaments.

Electron microscopy

The palisading nuclei of the bipolar tumor cells contain a distinct nucleolus. Endoplasmatic reticulum is well developed in most tumor cells. Within the thin tapering

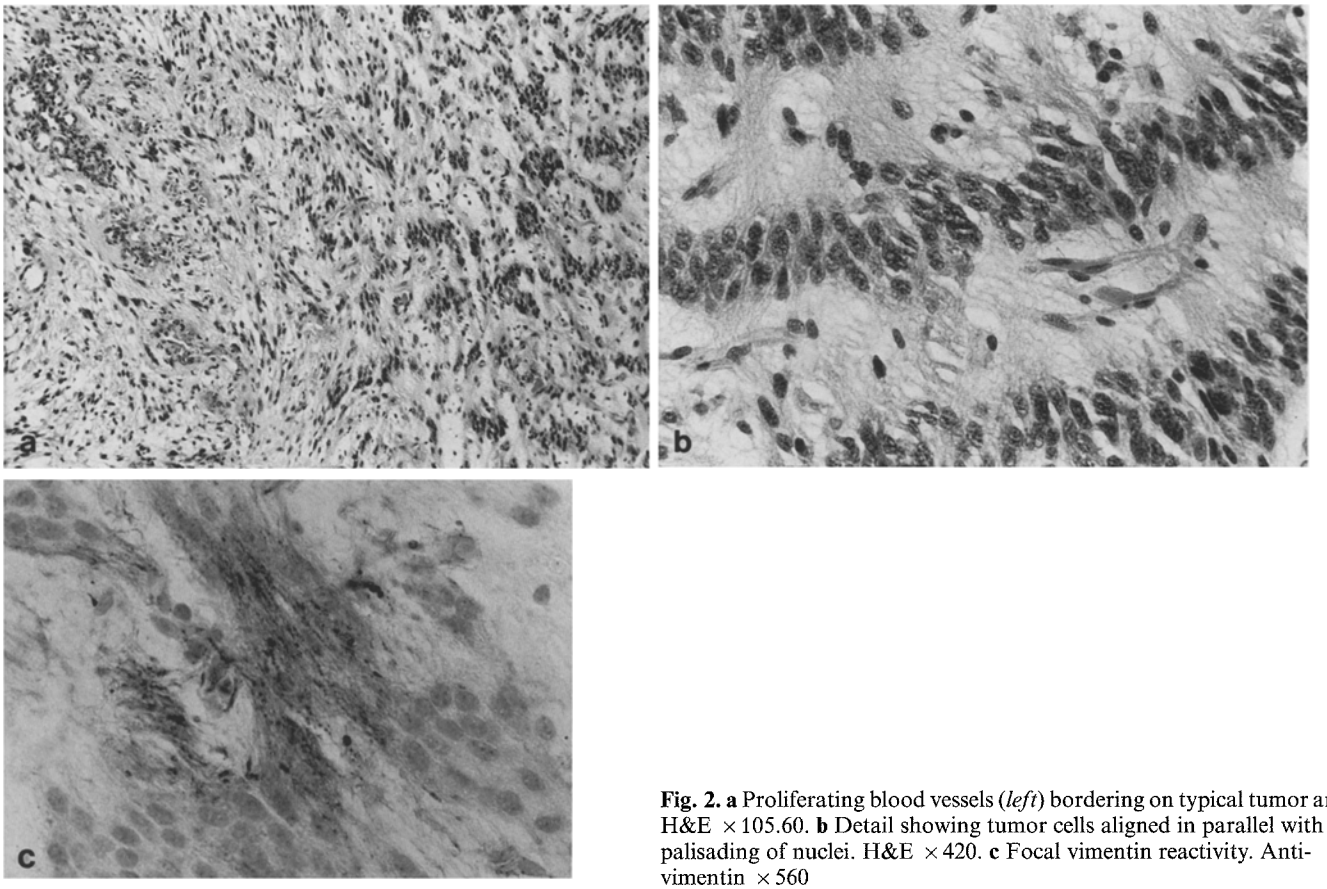

Fig. 2. a Proliferating blood vessels (*left*) bordering on typical tumor area. H&E ×105.60. **b** Detail showing tumor cells aligned in parallel with palisading of nuclei. H&E ×420. **c** Focal vimentin reactivity. Anti-vimentin ×560

cellular processes of most cells microtubuli, probably neurotubuli, are a prominent finding. In the processes of a few tumor cells densely packed filaments, resembling glial fibers, are present. Secretory granules are not found at the vascular endings of the processes, which occasionally insinuate between layers of vascular basement membrane (Fig. 3a–e).

Discussion

The polar spongioblastoma is morphologically a distinct neoplasm of the central nervous system. Eleven cases have been presented until now, two with limited clinical data [6]. Disregarding a case with extremely late presentation [11], the mean age of onset of the presenting symptoms was 7.8 years and there was an equal female-male ratio (Table 2). In seven out of ten cases the tumor was localized in the brain stem or cerebellum and in two cases in the spinal cord. Localization in the cerebral hemispheres was until now unreported. CSF metastasis was found in three out of six fatal cases [2, 5, 8]. Histologically the present case shows a tumor part resembling the juvenile-type pilocytic astrocytoma, a benign entity. Also an area with vascular proliferation is present, suggestive of a more malignant nature. Survival time seems to be remarkably increased compared to the first described cases, but this is based on too few cases to draw definite conclusions.

The polar spongioblastoma has been claimed to be cytogenetically derived from the primitive radial glia [7]. In astrocytomas and oligodendrogliomas growth patterns similar to polar spongioblastomas occur [9]. At the immunohistochemical and electron microscopical level, the present case focally shows signs of astrocytic differentiation. However, in most of the tumor NSE reactivity, microtubuli and prominent endoplasmatic reticulum are present. Although developing glioblasts need not contain glial fibers, the presence of many microtubuli and well-developed endoplasmatic reticulum is more characteristic of neuronal elements [13]. A possible neuronal cytogenesis of the polar spongioblastoma is also supported by the presence of neurosecretory granules and some microtubuli in a similar case [3]. Besides, neuroblastomas are capable of forming structures closely resembling the typical palisading polar spongioblastoma structures [4].

The coexpression of neuronal and glial elements in this tumor is intriguing and may cast doubt on a cytogenesis strictly from radial glia. Ultrastructural and immunohistochemical data of more cases are needed to establish the exact nature and lineage of this tumor.

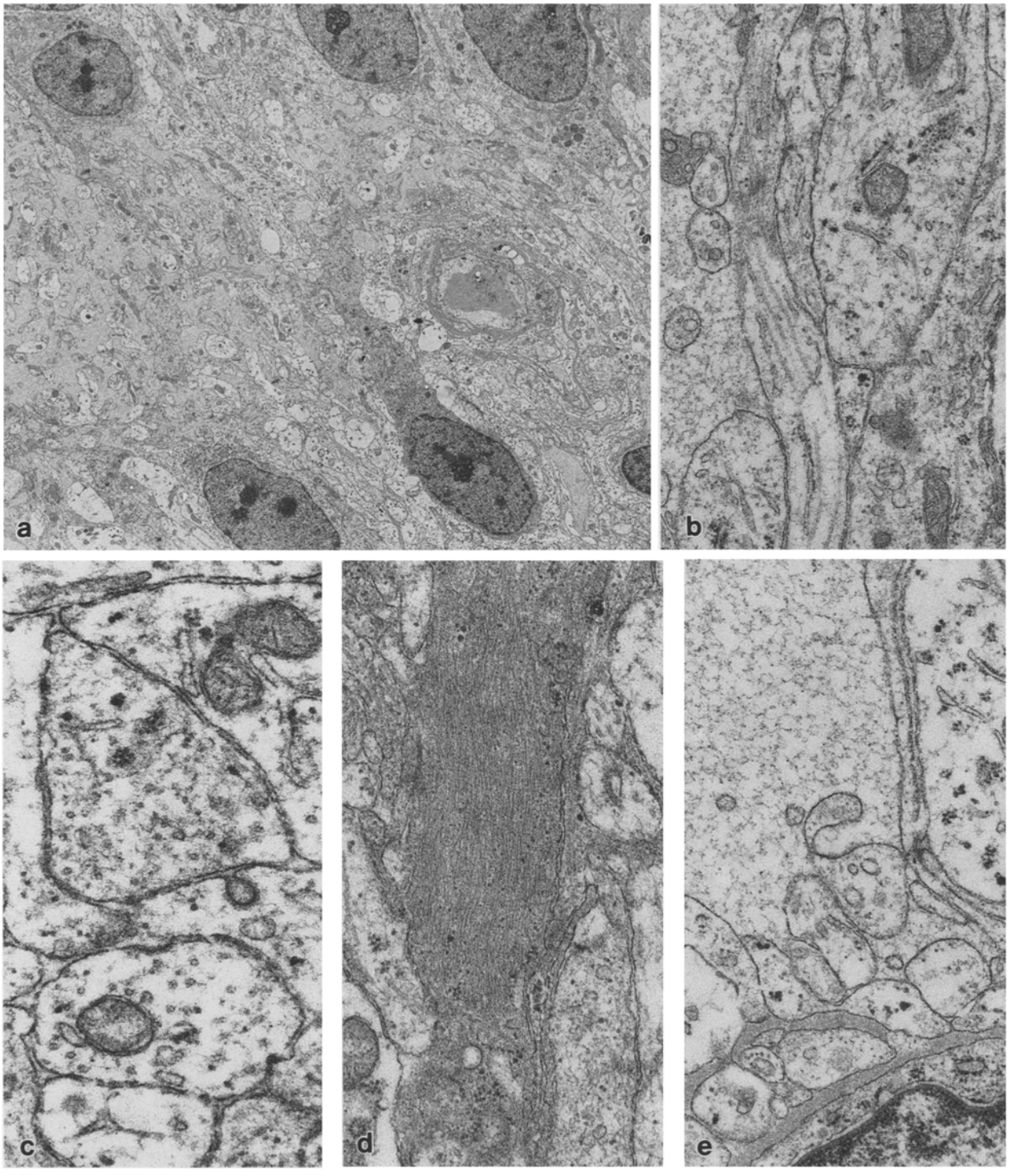

Fig. 3. a Tumor cells with palisading of nuclei. ×2200. **b** Longitudinally sectioned thin tumor cell processes filled with microtubuli. ×23700. **c** Cross-sectioned cell processes showing microtubuli. ×35000. **d** Cell processes containing glial fibers. ×2300. **e** Perivascular part of processes lacking secretory granules. At lower left processes insinuate between vascular basement membrane. ×25500

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