Acta Neuropathol (2005) 110: 196–198
DOI 10.1007/s00401-005-1037-1

LETTER TO THE EDITOR

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Rhabdoid cystic papillary meningioma with diffuse subarachnoid dissemination

Received: 29 March 2005 / Revised: 21 April 2005 / Accepted: 21 April 2005 / Published online: 25 June 2005

Papillary meningioma is a rare variant of meningioma defined by the presence of a perivascular pseudopapillary pattern. Because papillary meningioma typically displays invasion of the brain, local recurrence, and distant metastasis, it has been graded as a WHO grade III tumor [8]. Rhabdoid meningioma is a relatively newly recognized, distinct WHO grade III tumor that frequently develops on a background of other meningioma subtypes [8]. Perry et al. [9] described the first case of rhabdoid meningioma with papillary architecture resembling ependymoma in a 13-year-old girl. Subsequently, three additional cases of rhabdoid papillary meningioma have been reported [1, 4, 10]. Here, we report a case of rhabdoid papillary meningioma with macroscopic and microscopic cysts, displaying extensive leptomeningeal dissemination after frequent local recurrence.

A 12-year-old girl was examined for focal motor seizures of the neck. On neurological examination, she showed double vision. Computed tomography (CT) scan showed a right frontal hypodense cystic lesion with a heterogeneously enhanced nodule and enhancement of the cyst wall (Fig. 1a). At operation, the dura mater showed no evidence of invasion. The solid tumor was located in the superficial part of the cortex and contained calcification. The border with the cerebral parenchyma was partly unclear. Gross total removal of the tumor was achieved. Xanthochromic fluid was obtained from the cyst. The cyst wall was not removed. After a histological diagnosis of anaplastic ependymoma had been established at an outside institution, the patient received a course of radiotherapy totaling 50 Gy to the surgical area. She was discharged from our hospital, without neurological deficit. However, five local recurrences were noted over the following 11 years, and the patient died of diffuse subarachnoid dissemination at the age of 25 years.

Paraffin sections from the surgical and autopsy materials were stained with hematoxylin and eosin (HE), periodic acid-Schiff (PAS), and silver impregnation for reticulin. Other sections were immunostained using a polyclonal antibody against glial fibrillary acidic protein (GFAP; Dako, Glostrup, Denmark; 1:500), and monoclonal antibodies against vimentin (Dako Cytomation, Carpinteria, CA; 1:50), cytokeratin (AE1/AE3; Dako Cytomation; 1:50), epithelial membrane antigen (EMA; Dako Cytomation; 1:50), desmin (Dako; 1:50), α-smooth muscle actin (Dako; 1:100), synaptophysin (Boehringer, Mannheim, Germany; 1:500), neurofilament (Sanbio, Uden, The Netherlands; 1:100), BAF47/SNF5 (BD Transduction Labs, San Diego, CA; 1:250) [6] and Ki-67 (MIB-1; Dako; 1:50).

Microscopic examination of the original tumor showed a sheet-like structure throughout most of the specimen. Mitotic figures and small necrotic foci were occasionally seen. In some areas, however, rhabdoid morphology was defined as sheets of loosely cohesive cells with eccentric nuclei and hyaline, paranuclear inclusions (Fig. 1b). In addition, loss of cellular cohesion led to the focal emergence of papillary architecture (Fig. 1c), accompanied by a dense network of perivascular reticulin fibers (Fig. 1d). Moreover, microcysts of various sizes or ependymal canal-like structures were found. Areas showing transition from the sheet-like structure to the microcystic areas were also evident (Fig. 1e). The wall of the microcysts showed an epithe-

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lial lining of tumor cells, whose cytoplasm was negative for PAS staining. No whorl formation was noted. Immunostaining for vimentin (Fig. 1f) and EMA (Fig. 1g) was extensively positive, whereas cytokeratin, GFAP, desmin, α-smooth muscle actin, synaptophysin and neurofilament were all negative. BAF47/SNF5 expression was preserved in the tumor. In the microcysts, EMA immunoreactivity was present in the apical surface of the tumor cell lining. MIB-1 stain for Ki-67 antigen was positive in 7% of the tumor cell nuclei. The histology of the recurrent tumors was fundamentally similar to that of the original tumor, although rhabdoid components became increasingly prominent with subsequent resections.

Postmortem examination revealed many subdural tumor deposits in the anterior and middle cranial fossa (Fig. 1h). Microscopically, the anaplastic character of the tumor was more evident at autopsy. The tumor was composed of round or polygonal cells with abundant eosinophilic cytoplasm and clearly defined borders (Fig. 1i). The tumor in the subarachnoid space showed brain invasion along the Virchow-Robin spaces. The MIB-1 labeling index of the final tumor was 8%. An electron microscopic study revealed desmosomes between the tumor cells (Fig. 1j).

Rhabdoid meningiomas are defined by the presence of variably abundant eosinophilic cytoplasm, eccentric nuclei, hyaline paranuclear inclusions, and meningothelial differentiation at the light microscopic, immunohistochemical, or ultrastructural level. Papillary meningiomas are characterized by a perivascular pseudopapillary pattern and a rich peripapillary reticulin network. The present case showed both rhabdoid and papillary morphologies from the onset. These histological features were also evident in the recurrent tumors, although the tumor obtained at autopsy was composed entirely of sheets of rhabdoid cells. Therefore, we considered the present tumor to be an example of a rhabdoid papillary meningioma. Four cases of rhabdoid papillary meningioma have been described previously [1, 4, 9, 10]; focal recurrence was seen in two cases [9, 10] and diffuse leptomeningeal spread in one [1]. This is in line with the clinical aggressiveness of both rhabdoid and papillary meningiomas.

In the present tumor, macroscopic and microscopic cyst formation was another feature of interest. Pathologically, macroscopic cyst formation tends to occur most often in meningothelial meningiomas of adulthood [12], whereas the fibroblastic form predominates in infancy [7]. However, cystic components have also been demonstrated in some cases of papillary meningioma by CT or magnetic resonance imaging scans [11]. On the other hand, cyst formation is not a feature of rhabdoid meningiomas. Although previous studies have reported

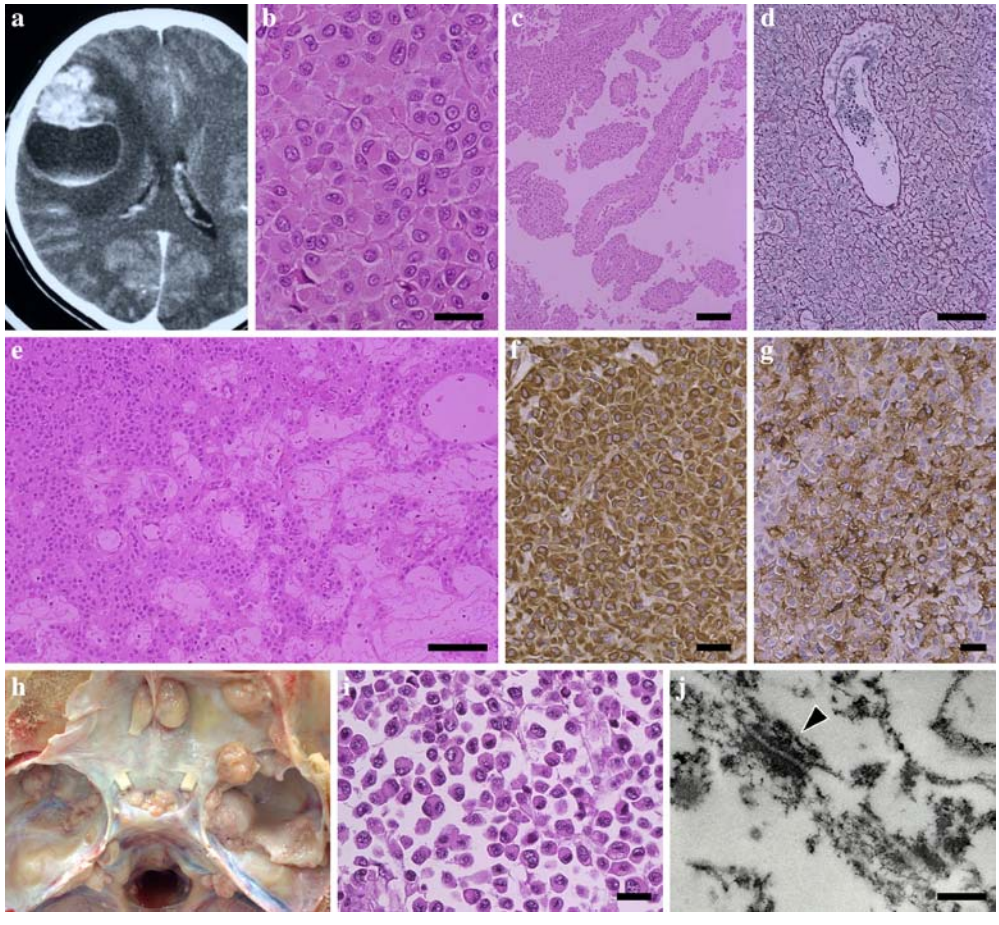

Fig. 1 **a** CT scan with contrast material showing a solid and cystic lesion (5×6 cm) in the right frontal region. The solid mass shows non-homogeneous enhancement, and the cyst wall is also enhanced. **b**–**g** Histology of the solid component of the original tumor. **b** An area showing rhabdoid morphology of the tumor cells; HE staining. **c** An area showing papillary structures; HE staining. **d** Rich reticulin network arising from the blood vessels and surrounding individual tumor cells; Gitter stain. **e** A zone showing transition from a sheet-like (*left upper*) to a microcystic (*right lower*) structure; HE staining. **f**, **g** Strong immunoreactivity for vimentin (**f**) and EMA (**g**). **h**–**j** Gross and histological findings at autopsy. **h** Skull base showing many tumor deposits in the subdural surface and cranial nerve roots. **i** Sheets of loose rhabdoid cells in the subarachnoid space; HE staining. **j** Electron micrograph showing desmosomes between the tumor cells (*arrowhead*) (*HE* hematoxylin and eosin, *EMA* epithelial membrane antigen). *Bars* **b, i** 25 μm; **c**–**g** 100 μm; **j** 200 nm

that cystic meningiomas are usually benign, it is noteworthy that they often recur [3, 5]. Several authors have suggested that tumor cells can be present in the cyst wall, and therefore that incomplete extirpation of the tumor capsule is the main reason for recurrence of cystic meningiomas [3]. Recently, Chen et al. [2] reported that among 15 cases of cystic meningioma, 6 were atypical and 2 were malignant. Considering that cyst formation is not uncommon in papillary meningiomas, and that these tumors are aggressive, it is also possible that recurrent cystic meningiomas may include a significant number of papillary meningiomas.

In conclusion, we consider that the present case was a rare example of rhabdoid cystic papillary meningioma with anaplastic features, suggesting a histological link between rhabdoid, cystic, and papillary meningiomas. Cyst formation may be one of the features of papillary meningiomas.

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