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Microcystic Meningioma

- A Case Report -

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Microcystic meningioma is a distinct morphological variant of meningioma, characterized by loose texture and microcysts with formation of large extracellular spaces containing edematous fluid. The tumor cells have stellate and vacuolated cytoplasm with long cytoplasmic processes. We experienced a case of typical microcystic meningioma occurring in the falx cerebri adjacent to the left superior parasagittal sinus in a 41-year-old man. The tumor showed the typical histologic features with a few nests of meningothelial whorls. The tumor cells showed diffuse immunoreactivity for vimentin, epithelial membrane antigen (EMA), and S-100 protein. Ultrastructural study confirmed the meningiomatous nature of the tumor.

Key Words : *Microcystic meningioma, Immunohistochemistry, Ultrastructure*

INTRODUCTION

Microcystic meningiomas have recently been described as a distinct variant of meningiomas (Michaud and Gagne, 1983). They are characterized by the formation of numerous large extracellular microcystic spaces containing edematous fluid, and by the stellate and vacuolated appearance of the tumor cells, with occasional large hyperchromatic and pleomorphic nuclei. Their unusual histological appearances may lead to confusion with schwannomas, chordomas, astrocytomas, hemangioblastomas, and angioblastic meningiomas. However, clinical features and prognosis of the tumor do not differ from those of usual benign meningiomas (Ng et al., 1989). We report the light microscopy, immunohistochemistry, and ultrastructure of a microcystic meningioma with review of the literature.

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CASE REPORT

A 41-year-old man suffered from brief episodes of seizure on two occasions over a period of four months. Neurologic examination was unremarkable at the time of admission. Laboratory examinations showed normal findings except for type III hyperlipoproteinemia. A magnetic resonance image (MRI) scan showed a well demarcated peripheral enhancing mass over the left superior parasagittal sinus (Fig. 1). At surgery, the tumor arose from falx cerebri and was a well circumscribed noninvading mass on the anterior portion of Rolandic vein of the left frontal lobe. The tumor was completely removed. The resected tumor was well encapsulated by the thin fibrous capsule with focal area of fibrous adhesion. It was soft, spongy, and measured $3 \times 2 \times 1.5$ cm in dimensions. The cut surface was yellow-light brown and slimy. Hemorrhage and necrosis were absent (Fig. 2).

Microscopically, the tumor showed a loosely reticular or lace-like appearance with numerous large extracellular cystic spaces. Cytoplasmic vacuoles were distinctive (Fig. 3). These spaces were delimited by cytopla-

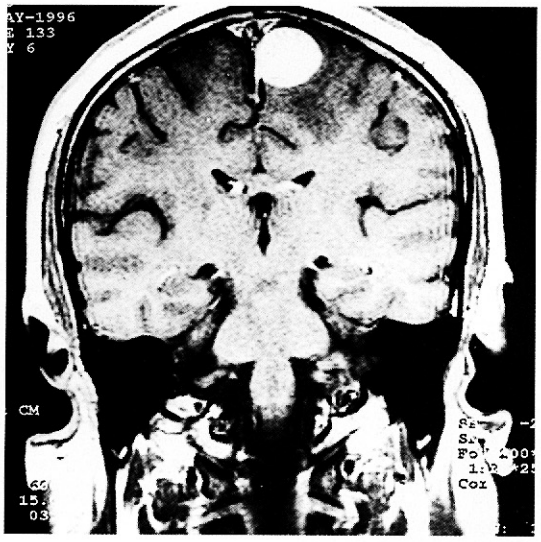

Fig. 1. A magnetic resonance image showing a well demarcated peripheral enhancing mass over the left superior parasagittal sinus.

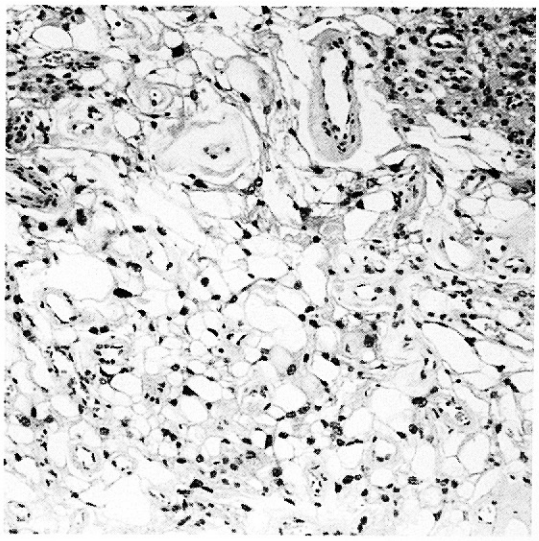

Fig. 3. The tumor showing a loosely reticular or lace-like appearance with numerous extracellular cystic spaces. Note solid meningothelial nests (H & E, ×100).

Fig. 2. The cut surface of the tumor showing yellow-light brown, soft, and spongy appearance.

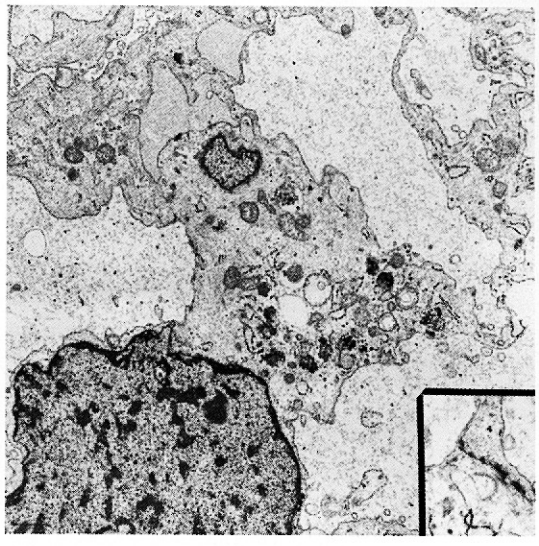

Fig. 4. Long and irregular cytoplasmic processes delineate large extracellular spaces. Processes are joined by desmosomes (inset) (Original magnification, ×4,000 ; inset, ×8,000).

smic processes of stellate-shaped tumor cells with cytoplasmic borders. The cystic spaces contained granular, faintly eosinophilic, edematous fluid which did not stain with either mucicarmine, PAS or alcian blue. More solid area showed some fibrillary appearance, and occasional small typical meningothelial whorls. Markedly hyalinized blood vessels were abundant, which simulated angiomatous meningioma. Although most tumor cells had small vesicular nuclei, some tumor cells had bizarre, pleomorphic nuclei. The immunohistochemical

study showed diffuse positivity for vimentin, EMA, and S-100 protein, but negativity for cytokeratin, GFAP, and Factor VIII related antigen. Electron microscopy showed stellate or slightly elongated tumor cells with irregular long processes bordering large extracellular spaces which were empty or filled with a fine granular substance (Fig. 4). The cytoplasmic processes were occasionally interdigitated and joined by desmosomes. The nuclei were round, ovoid, or slightly irregular in contour. The heterochromatin was marginated and finely stippled. The cytoplasm contained Golgi apparatus, mitochondria, rough endoplasmic reticulum, ribosomes, polysomes, and filaments. Basement membrane-like substance was not found.

DISCUSSION

Microcystic meningiomas have recently been recognized as an unusual morphological variant of meningiomas. They were originally described by Masson who labeled it "humid" because of its gross appearance (Michaud and Gagne, 1983). Similar lesions have been subsequently reported under various designations such as humid and myxomatous (Dahmen, 1979), vacuolated (Eimoto and Hashimoto, 1977), and microcystic (Kleinman et al., 1980). They have been classified as a distinct subgroup of meningiomas in the WHO classification of central nervous system tumors (Kleihues et al., 1992). To our knowledge, about 23 cases so far have been reported in English literature.

The pathogenesis of these tumors is unclear but transudation of low-protein fluid was suggested (Michaud and Gagne, 1983). Histochemical stains in our case indicate extracellular fluid is neither mucin nor glycogen. The age of affected patients ranges from 3 to 74 years of age. There is a slight female predominance. Most cases present as tumors of the cerebral convexities or parasagittal (Ng et al., 1989 ; Nishio et al., 1994a). Grossly, microcystic meningiomas are well delimited, with a smooth bosselated or lobulated external surface. The texture is usually soft and spongy. The cut surface discloses a yellowish-light brown homogeneous and a glistening appearance. Hemorrhage and necrosis are never found (Michaud and Gagne, 1983 ; Sobel and Michaud, 1985 ; Nishio et al., 1994b).

Microscopically, these tumors show numerous cystic spaces filled with edematous fluid and are lined by stellate-shaped meningothelial cells. Some areas show the conglomerations of smaller cystic spaces, which create a vacuolated, myxoid and loosely reticular appearance (Michaud and Gagne, 1983 ; Ng et al., 1989 ; Kuchna et al., 1994). Foci of nuclear pleomorphism are occassionally noted, but this finding does not indicate aggressive behavior (Ng et al., 1989 ; Nishio et al., 1994b). The tumor shows occasionally typical meningiomatous whorls (Michaud and Gagne, 1983). Meningiomas demonstrate concurrent mesenchymal and epithelial differentiation as shown by their consistent and reliable immunohistochemical positivity for both EMA and vimentin (Schnitt and Vogel, 1986). S-100 protein can be found focally in many meningiomas (Taylor and Cote, 1994). The immunohistochemistry of microcystic meningiomas is not different from other types of meningiomas. The tumor cells of reported cases showed usually positive reactivity for vimentin and EMA, but negative for cytokeratin, S-100 protein, GFAP, and Factor VIII related antigen (Michaud and Gagne, 1983 ; Ng et al., 1989). The present case showed same immunohistochemical results except for diffuse positivity for S-100 protein. Ultrastructural findings are similar to those of usual meningioma except for extracellular spaces. The microcysts are in extracellular location and contain granular and floccular low electron dense material (Michaud and Gagne, 1983 ; Sobel and Michaud, 1985 ; Rutherfoord and Marus, 1987).

The importance of recognizing this lesion lies in differential diagnosis from other central nervous system tumors with a myxomatous appearance (Ng et al., 1989). Myxomatous schwannomas can be distinguished by positive staining for S-100 protein and negativity for EMA. Loose fibrillary cytoplasms simulate pilocytic astrocytomas, which can be distinguished by immunostaining with anti-GFAP. Chordomas would show positive staining for cytokeratin and S-100 protein. Careful distinction should be given to metastatic carcinomas in view of the nuclear pleomorphism. The absence of mitotic figures and occasional meningothelial whorls are important features distinguishing from a metastatic carcinoma. Hemangioblastomas show focal positive for S-100 protein and GFAP, but negative for EMA. Abundant blood vessels with marked hyalinization mimick angiomatous or angioblastic meningiomas, which show positive staining for vimentin, but negative for EMA.

In summary, microcystic meningiomas are an unusual, but distinct morphological variant of meningioma, which are clinically and immunohistochemically similar to conventional meningiomas. They have to be distinguished from other lesions of similar histology.

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