Chordoid meningioma in a child

Case report

HITOSHI KOBATA, M.D., AKINORI KONDO, M.D., KOICHI IWASAKI, M.D., HIROFUMI KUSAKA, M.D., HIROFUMI ITO, M.D., AND SHINJI SAWADA, M.D.

Departments of Neurosurgery, Neurology, and Clinical Pathology, Kitano Medical Research Institute and Hospital, Osaka, Japan

A case of chordoid meningioma occurring in a 15-year-old girl is presented. The patient manifested seizures as the initial symptom and subsequently exhibited subclinical microcytic hypochromic anemia. The tumor, located in the falcotentorial region and associated with diffuse edema, was totally resected. On histological examination of the surgical specimen, the clustering pattern of partly vacuolated cells in the mucoid stroma mimicked chordoma; however, positive staining of individual cells for vimentin and epithelial membrane antigen led to a diagnosis of meningioma. Interestingly, the tumor cells were surrounded by a periodic acid–Schiff– and type IV collagen–positive substance. Electron microscopy demonstrated a strikingly dense and thick basal lamina. The patient’s microcytic hypochromic anemia disappeared after the tumor was removed. Both the clinical and pathological features of this case resemble those of chordoid meningioma, a rare meningioma variant.

KEY WORDS • brain lesion • meningioma • chordoid meningioma • basal lamina • children

Unusual variants of meningioma containing abundant lymphoplasmacellular infiltrates are reported to be frequently associated with hematological abnormalities. The most recent World Health Organization classification includes two subtypes of this entity: lymphoplasmacyte-rich (LPR) and chordoid meningioma. Both are rare variants, and the latter in particular is characterized by its resemblance to chordoma. However, the clinical and pathological features of these tumors are not well characterized because few case reports are available. To the best of our knowledge, only nine cases of chordoid meningioma have been reported since Kepes, et al., first described this entity. We present a recent case of chordoid meningioma in a young girl.

Case Report

History. On November 16, 1993, this 15-year-old girl suddenly complained of blurred vision and visual field constriction; she subsequently lost consciousness and developed generalized tonic–clonic seizures. On admission to our hospital 3 days later, she was alert and showed no abnormal neurological signs or symptoms. Beginning 10 months prior to admission, she had been suffering seizures that caused visual field constriction several times per month.

Examination. Physical examination revealed no somatic or genital retardation and the patient’s menstruation cycle was regular. Hepatosplenomegaly was not detected. At admission her height was 158 cm and her weight 46 kg.

Laboratory findings included a hematocrit level of 37.2%, hemoglobin count of 11.1 g/dl, mean corpuscular volume of 84.5 fl, mean corpuscular hemoglobin of 25.2 pg, and mean corpuscular hemoglobin concentration of 29.8 g/dl. The patient’s serum immunoglobulin level was normal, with an immunoglobulin G value of 1840 mg/dl (normal range 1000–2000 mg/dl). Results of extensive serological studies for autoimmune disease or antibodies to human T-cell leukemia virus were all negative. An electroencephalogram revealed sporadic sharp waves in the parietooccipital region and a computerized tomography scan showed a round brain tumor 3 cm in diameter attached to the right falcotentorium. On magnetic resonance (MR) imaging, the tumor displayed an iso- to slightly hypointense signal in relation to the cerebral cortex and was homogeneously enhanced with intravenous administration of gadolinium on T1-weighted studies. A T2-weighted MR image revealed a tumor of high signal intensity with a rim of low intensity surrounded by profound brain edema (Fig. 1). Digital subtraction angiography showed the tumor to be fed mainly by the posterior meningeal artery.

J. Neurosurg. / Volume 88 / February, 1998

319
Chordoid meningioma is a rare variant of meningioma so named by Kepes, et al., in 1988. The first known case was reported by Connors in 1980 and later reviewed by Dimand, et al. This case involved a 15.5-year-old boy who harbored a posterior fossa tumor attached to the tentorium and who showed retarded somatic and sexual development, hepatosplenomegaly, iron-refractory hypochromic microcytic anemia, and bone marrow plasmacytosis with dysgammaglobulinemia (Castleman syndrome). After tumor resection he resumed normal growth and sexual development.

There was considerable disagreement on the distinguishing histological features of this disease until Kepes, et al., in addition to reviewing Connors’ case, reported six additional cases with similar histological and clinical features and characterized all of them as chordoid meningiomas. The tumors in these cases displayed characteristics of primary meningeal tumors that had undergone a peculiar mucoid degeneration resulting in histological characteristics closely imitating those of chordoma; in addition, they were all surrounded by a heavy mantle of reactive lymphoplasmacellular infiltrate often showing follicles and germinal centers. Clinically they were associated with varying degrees of microcytic hypochromic anemia; some also exhibited features of Castleman syndrome. In all cases surgical resection of the tumor produced improvement in systemic disease.

**Discussion**

**Operation.** On December 3, 1993, the pinkish and elastic hard tumor was totally removed via an occipital interhemispheric approach. The dural attachment to the tumor adjacent to the inferior sagittal sinus was coagulated.

**Postoperative Course.** The patient’s postoperative course was uneventful. Her postoperative electroencephalographic and hematological findings returned to normal. An MR image showed no tumor recurrence 2 years after surgery.

**Pathological Findings.** On histological examination, the tumor was found to be composed of various elements. Although a few areas showed a meningothelial pattern (Fig. 2 upper left), the larger portion of the tumor tissue was characterized by its resemblance to chordoma. Clusters of tumor cells arranged in chainlike fashion (some of them containing vacuolations) were recognized against an intercellular substance that displayed myxoid changes (Fig. 2 upper right). Some tumor cells mimicked so-called “physaliferous cells.” Few whorl formations were seen. There were no cellular atypia, mitotic features, or necrotic foci indicative of malignancy. Profuse lymphoplasmacellular infiltrates surrounded the island of tumor cells (Fig. 2 center left), with perivascular cuffing adjacent to the brain parenchyma. Most tumor cells were enclosed by a layer of periodic acid–Schiff (PAS)–positive substance and the myxoid matrix stained faintly by alcian blue (Fig. 2 center right).

The vast majority of the tumor cells were strongly positive for vimentin and epithelial membrane antigen (EMA) (Fig. 2 lower left) but negative for glial fibrillary acidic protein, cytokeratin, carcinoembryonic antigen, and S-100 protein. The PAS-positive substance surrounding the tumor cells was also positive for type IV collagen (Fig. 2 lower right). Most cells were positive for L26 in a heavy mantle of lymphoplasmacellular infiltrates, and cells positive for UCHL 1 were primarily observed around vessels.

Electron microscopy revealed tumor cells containing irregular nuclei, scattered mitochondria, well-developed Golgi apparatus, rough endoplasmic reticula, vesicles, and granules (Fig. 3 upper). Intermediate filaments were observed in the cytoplasm in varying numbers and patterns of distribution from cell to cell. Some cells had intracytoplasmic vacuolar spaces. The cell surface displayed pseudopodia coated with thick basal laminae, which in some areas extended discontinuously into the intercellular space (Fig. 3 center). Tumor cell processes were surrounded by an electron-dense substance in some areas (Fig. 3 lower). Sparsely distributed tumor cells were connected by desmosomal attachments; however, interdigitations of cell processes were rare. Characteristically for this type of tumor, a considerable amount of basal lamina–like material was seen in the extracellular space.

Based on these histological findings, it was concluded that the tumor was a chordoid meningioma. The associated microcytic hypochromic anemia, which improved after tumor resection, was compatible with this type of meningioma, as was its occurrence in a young person.
It is interesting to note that although the incidence of meningioma in the general pediatric population is extremely low, all the cases of chordoid meningioma reported to date, including ours, have occurred in the first two decades of the patient’s life. The present case displayed both the clinical and histopathological characteristics of chordoid meningioma.

The ultrastructure of this type of tumor has been de-
scribed in three cases. In two of these cases, interdigitations with numerous desmosomal attachments were observed. Although Zuppan, et al., stated that the extracellular matrix of the chordoid meningioma they examined included regions of amorphous “fuzzy” material with no evidence of an external lamina, Kepes, et al., reported that in one case numerous cells displayed microvilli and some portions of their surface were covered by stretches of basal lamina, whereas in another case the tumor cells were surrounded by a fluffy granular intercellular substance, with occasional condensation of basal lamina–like material. Our case was characterized by pseudopodia and dense basal lamina surrounding the tumor cells to varying degrees.

Type IV collagen is found incorporated into a netlike structure in the lamina densa of basement membranes. The ability of meningioma cells to produce extracellular matrix protein has been demonstrated in tissue culture studies as well as in surgical specimens. Basement membrane proteins such as type IV collagen and laminin have also been observed on the surface of notochord and chordoma cells. The dense pericellular distribution of the extracellular matrix in our case suggests production of the matrix by tumor cells, which may have contributed to the peculiar myxoid appearance of the stroma. The dual differentiation into epithelial and mesenchymal cells poses diagnostic difficulties.

The majority of infiltrative lymphocytes in meningioma consist of T cells. It is curious that chordoid and LPR meningiomas are strongly associated with predominantly type B lymphocytes and plasma cells. Immunostaining for κ and λ chains confirms the polyclonality of the plasma cells. Castleman syndrome may affect the central nervous system. Chordoid and LPR meningiomas may accompany this uncommon syndrome, as may dysgammaglobulinemia and microcytic hypochromic anemia. These systemic symptoms, often occurring in young patients, can be explained as effects of the associated lymphoplasmacellular infiltrates.

Diagnosis of chordoid meningioma should be based on the chordoma-like histological appearance of the tumor as well as on the meningothelial cell–like pattern that should be recognizable in at least some areas. Differential diagnosis should be established from chordoma, other variants of meningioma, and certain tumorlike conditions associated with a mass and lymphoplasmacellular infiltration. Our immunohistochemical study showed positive staining for vimentin and EMA but not for cytokeratin or S-100 protein; the latter two are strongly expressed by chordoma. Myxoid and microcystic variants show considerable overlap with some features of chordoid meningioma in that all may display some degree of myxoid stromal changes and cytoplasmic vacuolation. However, these variants lack inflammatory cell infiltrates. Lymphoplasmacyte-rich meningiomas do not exhibit chordoid features.

There is little information on the neuroradiological findings associated with this tumor. Peritumoral edema has been identified on computerized tomography scans in two of three patients examined by means of this imaging modality. Magnetic resonance findings have been reported in only one case thus far: these included a hypointense mass detected beneath the left tentorium with intense enhancement, and associated bilateral cerebellar white matter edema. Diffuse edema has also been seen on MR imaging in association with LPR meningioma. This edema may reflect deep lymphoplasmacellular infiltration. Finally, it is noteworthy that two of seven patients in the series reported by Kepes, et al., experienced recurrence, and that in another case chromosomal analysis revealed deletions involving chromosome 22.

Acknowledgments

The authors thank Drs. Asao Hirano, Division of Neuropathology, Montefiore Medical Center, and Akira Tsutsumi, Division of Neurology, Montefiore Medical Center.
Chordoid meningioma in a child

Department of Central Laboratory, Osaka Medical College, for helpful comments on the histological findings. We also thank Mr. Satoshi Fukui for excellent technical assistance.

References


Manuscript received March 31, 1997. Accepted in final form September 8, 1997

This case was presented at the 14th Annual Brain Tumor Pathology Meeting, April 26, 1996, Tokyo, Japan.

Address reprint requests to: Hitoshi Kobata, M.D., Department of Neurosurgery, Osaka Medical College, 2-7 Daigakumachi, Takatsuki, Osaka, 569, Japan.