CASE REPORT

Rhabdoid and papillary meningioma with leptomeningeal dissemination

Meningiome rhabdoïde et papillaire avec dissémination lepto-méningée

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KEYWORDS
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Summary Rhabdoid meningioma is a rare variant of meningioma classified as grade III under the new World Health Organization (WHO) classification of brain tumors. Although this tumor is known for its aggressive behavior, dissemination into cerebral spinal fluid (CSF) is extremely rare. We report here a case of rhabdoid meningioma in a young man, operated on twice previously, who presented with multiple CSF areas of seeding in the brain and spinal cord. The imaging findings for this tumor, including diffusion and perfusion MR sequences, are highlighted. This particular histological subtype of meningioma has a poor prognosis and must be treated aggressively.

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INTRODUCTION

Meningiomas comprise approximately 15% of all primary central nervous system (CNS) neoplasms. Meningiomas

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arise from the arachnoid cells lining the meninges or choroid plexus. Several histological subtypes of meningioma have been described, but have no prognostic significance. However, malignant meningiomas, including anaplastic, papillary and rhabdoid variants, are considered grade III, according to the World Health Organization (WHO) classification. Rhabdoid meningioma was first described by Kepes et al. [5] and later by Perry et al. [13]. According to the currently available literature, rhabdoid meningiomas are highly aggressive tumors known for their recurrence and often carry a poor prognosis. We report an unusual case of rhabdoid meningioma and describe its radiological findings which, despite aggressive surgery and radiation therapy, recurred with dissemination along the cerebral spinal fluid (CSF) spaces.

Case report

A 21-year-old man presented with headache and double vision for one month in February 2006. Examination revealed early papilledema and bilateral lateral rectus palsy. The rest of his neurological and general examination was unremarkable. MRI revealed a large extra-axial mass lesion in the right temporal region, with a poor interface with the adjacent brain parenchyma. The mass appeared isointense on T1-weighted sequences, isointense to mildly hyperintense on FLAIR sequences and heterogeneously hyperintense, with interspersed hypointensities, on T2-weighted sequences. Postcontrast study showed heterogeneous intense contrast enhancement. Small cystic degenerations were observed within the mass lesion (Fig. 1a–c).

The patient underwent surgery and the tumor was completely excised. Microscopic examination revealed islands of neoplastic cells showing rhabdoid characteristics in the form of abundant eosinophilic and granular cytoplasm, with eccentrically placed nuclei that showed varying degrees of nuclear atypia. Cells were arranged in and around the central core of fibrovascular tissue, giving rise to a papillary arrangement. Lymphocytic infiltration was observed within the neoplastic cells as well as in the connective tissue septa. Focal neuronal invasion and occasional mitotic figures were noted (Fig. 1d).

Later, in October, the patient again presented with complaints of persistent holocranial headache with bilateral lateral rectus palsy and papilledema. MRI revealed a recurrence of mass lesion that appeared hyperintense on T2 and FLAIR sequences and hypointense on T1 sequences. The tumor appeared to be heterogeneous with thin hypointense septations, giving a honeycomb appearance on the T2 images. The tumor showed intense heterogeneous contrast enhancement with two small satellite nodules lying adjacent to the tumor. In addition, there was thin ependymal lining enhancement of the right lateral ventricle, which had been missed on the initial reading (Fig. 2a–e). Gradient images revealed microbleeds within the mass. Diffusion-weighted images showed areas of restricted diffusion within the tumor. The mean rCBV ratio of the tumor compared with apparently normal white matter was 7.08. Mean curve analysis demonstrated an increase in blood volume within the tumor with an incremental decrease in signal intensity after the first pass, postenhancement, to below the baseline, with widening of the curve (Fig. 2f–i, k).

Surgery revealed a highly vascular tumor in the right middle cranial fossa with a poor plane of cleavage from the adjacent brain parenchyma. Histopathological studies showed a highly cellular and diffusely infiltrating neoplasm with cells showing rhabdoid features. Mitotic figures and areas of neuronal invasion were visualized in several areas of the tumor (Fig. 2j). Following tumor excision, the patient underwent 28 cycles of radiotherapy and was kept on follow-up.

In July 2007, he again presented to our hospital with sudden-onset weakness in both upper and lower limbs and urinary incontinence. Examination revealed a complete loss of power in his lower limbs and exaggerated extensor plantar reflexes on both sides. MRI showed multiple well-defined nodules in the wall of the left lateral ventricle, lateral wall of the third ventricle and near the cervicomedullary junction and thoracic spinal canal, suggestive of leptomeningeal dissemination (Fig. 3a–c). The patient refused surgery and radiotherapy and was discharged against medical advice.
Figure 2  October 2006. Patient presented with headache and double vision. Axial a: T2-weighted and b: FLAIR images show a mildly hyperintense and heterogeneous tumor; c: T1-weighted image shows a hypointense lesion; d, e: contrast-enhanced T1-weighted images show intense and heterogeneous contrast enhancement of the tumor; note the small satellite lesions and ependymal enhancement; f, g: tumor shows areas of minimal restricted diffusion; the ADC value is $0.90 \pm 0.1 \times 10^{-3}$ mm$^2$/s; h: microhemorrhages are evident in the gradient image; i: perfusion imaging with rCBV and color-overlay map shows increased areas of perfusion within the tumor; j: photomicrograph (hematoxylin & eosin stain) shows typical cells with rhabdoid features and mitoses (original magnification $\times 200$); k: signal intensity of tumor on dynamic susceptibility-contrast MRI versus time shows the increase in blood volume within the tumor.

Discussion

Rhabdoid meningioma, a recently described entity, is included as a grade III tumor in the 2000 WHO classification of CNS tumors [9]. The mean age of presentation for these tumors is 51 years and it is predominantly seen in women (the female-to-male ratio is 4:3). A review of the published literature on rhabdoid meningioma showed that recurrence occurs in more than half of the patients [10].

The histology of rhabdoid meningioma resembles that of a primary atypical teratoid/rhabdoid tumor of the brain. The latter is typically an intra-axial tumor in young children and
often associated with leptomeningeal dissemination and a poor prognosis [11]. Rhabdoid morphology resembles that of rhabdomyoblasts without differentiation into skeletal muscles. The cells show abundant cytoplasm with eosinophilic nuclei, hyaline paranuclear inclusions and meningotheelial differentiation [5,13,15]. These cells stain positive with vimentin, epithelial membrane antigen (EMA) and monoclonal antibody MIB-1 or proliferating cell nuclear antigen (PCNA) [5,13]. In most of the reported cases, the rhabdoid morphology was observed only after the first recurrence [2,5,13]. However, the cases reported by Kepes et al. [5] as well as others [7,10,12] described rhabdoid features at the time of diagnosis. In our case, rhabdoid features were observed at the time of the onset and there were no features to suggest rhabdoid transformation of a benign meningioma. This supports the view that this tumor might be of de novo origin.

In our case, the initial MRI showed a large extra-axial tumor with no clear-cut plane between the adjacent parenchyma, suggesting brain parenchymal invasion, which was confirmed at surgery. Subsequent MRI showed recurrence of the mass lesion, with small satellite nodules in the adjacent parenchyma with ependymal enhancement. Following resection and radiotherapy, our patient presented again with symptoms and MRI revealed a recurrent mass in the right temporal region and nodular metastatic deposits in the ependymal lining of the ventricles and spinal cord. A recent study of the imaging features of rhabdoid meningioma reported a strong tendency of these tumors towards cystic degeneration, bony involvement and prominent peritumoral edema [6]. However, apart from small cystic degenerations within the tumor, we could find no other features in our patient.

The diffusion and perfusion characteristics of this variant of meningioma have not been previously reported. Diffusion-weighted studies showed areas of restriction within the tumor, suggesting compact packing of the cells within the tumor. The apparent diffusion coefficient (ADC) values for this tumor were similar to those reported for such meningiomas [3]. The incidence of hemorrhage within the meningioma is reported to be very low [8]. Nevertheless, we found multiple microhemorrhages within the tumor in our case. Meningiomas with atypical MRI findings are often difficult to distinguish from intra-axial tumors and perfusion studies may be helpful in differentiating these tumors from intra-axial tumors, as reported by Hakymez et al. [4]. In our case, the tumor showed a high rCBV with a type-3 mean curve, which was similar to that described earlier [4].

Rhabdoid tumors are highly aggressive and prone to recur despite radical excision and radiotherapy. Nevertheless, intracranial dissemination of meningiomas into the subarachnoid space is exceedingly unusual [14]. Leptomeningeal spread of rhabdoid meningioma is rarely seen and only two cases are reported in the literature [1,15]. In one of those cases, leptomeningeal metastases appeared two months after surgery and radiation [1] and, in the other case, the patient developed CSF dissemination of tumor after 13 years [15]. Both of those cases presented with rhabdoid and papillary features.

Conclusion

This report is of a rare case of meningioma with rhabdoid and papillary features and CSF dissemination. The rhabdoid variant of meningioma can be extremely aggressive and recur, despite radical surgery and radiotherapy. There are no particular imaging findings to distinguish these tumors preoperatively. These tumors have the potential to spread along the leptomeninges and, hence, require close follow-up and aggressive management. However, the prognosis remains dismal and the optimal therapy for these tumors is yet to be determined.

References