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Total Cerebrospinal Fluid Enhancement Following Intravenous Gd-DTPA Administration in a Case of Meningiomatosis

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Summary: We report a case of severe meningiomatosis in a young girl, and describe the clinical course and MR appearance.

Index terms: Meninges, neoplasms; Cerebrospinal fluid, magnetic resonance; Contrast media, paramagnetic

Gd-DTPA has proved to be an invaluable agent in the evaluation of leptomeningeal disease. Focal or diffuse enhancement may be seen following intravenous contrast administration in various pathologic conditions including neoplastic and inflammatory entities. The enhancement, however, is usually restricted to the dural coverings. We present a case of diffuse cerebrospinal fluid (CSF) enhancement in a young child with meningiomatosis.

Case Report

The patient first presented at age 5 with a transient episode of speech impairment. Her neurologic evaluation at this time was unremarkable and there was no family history of neurofibromatosis. At age 9, the patient developed recurrent low back pain in association with episodes of headache and vomiting. Her physical examination was remarkable for hyporeflexia and nonenhanced spinal magnetic resonance (MR) imaging yielded a normal study. The patient's condition failed to improve and she required hospitalization. Physical examination revealed a lethargic child with bilateral papilledema, hypactive reflexes, lower extremity weakness, and a positive Kernig maneuver. A computed tomography (CT) scan of the head demonstrated hydrocephalus. Due to the findings of lower extremity paresis and weakness involving the girdle muscles, and the lack of a mass lesion on the CT scan to explain the hydrocephalus, a CT myelogram was performed. CT myelography was negative at this time. A ventriculoperitoneal shunt resulted in clinical improvement. In addition to revealing a normal ventricular size, contrast-enhanced MR showed prominent suprasellar, tentorial, and diffuse dural enhancement (Figs. 1A and 1B). The CSF analysis during this admission demonstrated an elevated protein value (170 mg/%) and negative cytology. Observation was continued without specific treatment. The patient subsequently developed episodes of aphasia and, 10 months later, a lumbar puncture was again remarkable for an elevated CSF protein and negative cytology. Contrast-enhanced cranial MR revealed increased meningeal enhancement with new nodular lesions involving the suprasellar cistern, tentorium, and convexities (Fig. 2). The suprasellar mass appeared to encroach upon the optic chiasm. The patient underwent meningeal biopsy from the region of the suprasellar cistern and right sylvian fissure that demonstrated meningiomatosis. Histology showed menigioma with mixed features, including meningothelial, fibroblastic, fibrous-histiocytic, lipoblastic, and poorly differentiated highly cellular areas.

Fig. 1. Initial enhanced MR (TR 700, TE 32).
A and B, Enhanced T1-weighted axial images demonstrate thick meningeal enhancement within the suprasellar cistern, perimesencephalic cisterns, tentorium and frontal, parietal, and occipital dura, as well as the leptomeninges in the Sylvian fissures. Intraventricular enhancement is not present (B).
Although there was an absence of necrosis and mitotic activity, the large area of high cellularity was indicative of an aggressive tumor. Soon after the biopsy, the child developed decreased visual acuity bilaterally that responded to a combination of steroid therapy and surgical decompression of a perichiasmal meningeal lesion. Myelography identified the presence of diffuse dural thickening involving the entire spinal neuraxis. CSF analysis now showed a significantly elevated protein content (812 mg/\%\) with negative CSF cytology. Radiation therapy was
administered to the craniospinal neuraxis for a total dose of 4500 rads. One year after radiation therapy and tapering of the corticosteroids, both visual and neurologic function returned to normal. Though meningeal enhancement was still present on a contrast MR 7 months later, the nodular lesions had resolved. The patient remained asymptomatic, and another contrast MR examination 1 year later (27 months after onset) demonstrated not only persistent meningeal enhancement involving the dura adjacent to the cerebral convexities and the falx cerebri, but also the meninges lining the cervical spinal cord. Moreover, striking diffuse CSF enhancement was noted throughout the entire neuraxis, including the ventricles, basal cisterns, and thoracic and lumbar subarachnoid spaces (Figs. 3A and 3E). This cerebrospinal fluid enhancement was not present on the earlier examinations. The intraventricular enhancement was probably due to the flow of gadolinium-enhanced CSF from the basal cisterns toward the patient's ventricular shunt tube.

Discussion

Meningiomatosis is the rarest form of neuraxis involvement by meningiomas. These can be divided into single meningiomas, multiple meningiomas, meningiomas associated with neurofibromatosis, and diffuse meningiomatosis. Meningiomatosis affects the meninges diffusely, as well as the underlying cerebral cortex. Diffuse meningiomatosis has remained difficult to manage. A combination of chemotherapeutic regimens and radiation therapy is usually employed, with surgical resection reserved for more focal symptomatic nodules, as in this case, where resection of a perichiasmal nodule was required to preserve vision. In this case report, a consequence of this infiltrative meningeal process is discussed, namely, the presence of Gd-DTPA throughout the subarachnoid space and ventricular system.

Unlike the brain and the leptomeninges, there is no blood-tissue barrier in the dura mater. Meningeal enhancement with Gd-DTPA is commonly seen in normal patients and is increased after craniotomy, particularly in children. Nodular and/or thick meningeal enhancement, however, is always associated with a pathologic process such as tumor or infection (1). A recent study revealed significant overlap between patients with meningitis and patients with meningeal carcinomatosis, in terms of meningeal enhancement (2). Attempts at classifying the patterns of enhancement into dural and leptomeningeal have not demonstrated an increase in specificity (3).

CSF, following intravenous Gd-DTPA administration, does not enhance under normal conditions. In fact, CSF is often used as an internal standard in the evaluation of pre- and postcontrast images (4). An unusual pronounced enhancement of the CSF in the subarachnoid spaces and ventricles occurred after the intravenous administration of 10 mL of Gd-DTPA in this 11-year-old child with histologically proven diffuse meningiomatosis. Diffuse enhancement of the meninges is well known and has been observed in several meningeal pathologic processes. CSF enhancement, however, is rare and was observed in the lumbar spinal canal in a few patients after surgery for primary intracranial tumors (5). In these cases, the enhanced CSF was difficult to differentiate from true meningeal neoplasia. In these postoperative patients, the CSF enhancement was transient, disappearing on repeat scans. The mechanism of the transient CSF enhancement was attributed to meningeal irritation by blood and/or blood products, or to methemoglobin effect (5). The cause of the CSF enhancement in the patient reported here is believed to be due to major leakage of Gd-DTPA into CSF as a consequence of a gross alteration of the blood-tissue barrier over a large area in direct contact with the subarachnoid space.

MR is very sensitive to small concentrations of Gd-DTPA, thus depicting CSF enhancement with greater ease than is possible by CT. In this patient’s rare disease, the entire meningeal barrier has been infiltrated by predominantly “en plaque” lesions, resulting in breakdown of the blood-tissue barrier and a vicarious outpouring of contrast molecules into the CSF, and a concomitant pronounced CSF protein elevation. The excessive outpouring of gadolinium was further promoted by the high vascularity of the meningiomatous tumors. In the setting of overwhelming meningeal disease, the blood-tissue barrier was rendered ineffective, becoming permeable to large molecules, as evidenced by the markedly elevated CSF protein (812 mg/dL) without the presence of other causes for this rise, such as a CSF spinal block (6, 7). Additionally, the corresponding CSF analyses failed to show subarachnoid hemorrhage, meningitis, or tumor cells.

A comparative model to this phenomenon has been demonstrated in experimental rabbits whose meninges have been infused with tumor cell suspensions, resulting in meningeal carcinomatosis. Subsequent study by contrast-enhanced MR and pathologic correlation showed extensive meningeal enhancement as a direct result of plaque-like tumor lesions infiltrating the fibrous stroma of the meninges, leading to a breach of the blood-
brain barrier. In those cases where the meningeal carcinomatosis was allowed to progress, contrast enhancement of the CSF was identified (8). Thus, a positive correlation exists between disease extent and degree of enhancement; the experimental animal model observations parallel the observations in this patient. That CSF enhancement in contrast MR is such a rare phenomenon initially seems paradoxical given the relative higher frequency of meningeal carcinomatosis. Meningeal carcinomatosis may be seen in up to 28% of patients with solid tumors (8). Typically, abnormal meningeal enhancement is usually restricted to the basal and perisylvian cisterns, cortical sulci, tentorium, or nodules in the CSF. The findings in the patient reported here and in the rabbit model, however, support the notion that CSF enhancement is the consequence of a severe tumor burden with exceptionally “leaky” vessels allowing the extravasation of large quantities of gadolinium as well as protein.

References