An unusual cystic appearance of disseminated low-grade gliomas

Abstract We report five cases of pediatric disseminated low-grade gliomas of the brainstem or spinal cord that exhibited an unusual, cystic pattern. Leptomeningeal disease was present in three of these at diagnosis, and was detected shortly afterwards in the other two. Four patients are alive up to 5 years later, following minimal to no intervention, while one is dead.

Keywords Gliomas · Low-grade · Disseminated · Leptomeningeal metastases · Cysts

Introduction Classically, leptomeningeal spread (LMS) of CNS tumors is described as nodularity, thickening, or coating of subarachnoid or ependymal surfaces. Although this pattern was present in the following cases of disseminated low-grade gliomas, an unusual, diffuse, cystic pattern was also noted. These cystic lesions seemed to preferentially affect the cerebellar hemispheres and other areas adjacent to the basal cisterns – including the brainstem and medial and inferior temporal and frontal lobes, and to actually extend into the underlying parenchyma.

Leptomeningeal disease was present at diagnosis in three of these pediatric patients and was detected shortly after in the other two. Each patient presented with symptoms suggesting LMS, and with hydrocephalus. Also, in each patient, although CSF protein was elevated, cytology findings were negative. Four of five patients are alive up to 5 years later, following minimal to no intervention.

Case reports

Case 1 A previously healthy 3.5-year-old girl presented with signs and symptoms of increased intracranial pressure. Enhanced head CT scan demonstrated hydrocephalus but no obstructing mass. She was shunted.
Fig. 1A–D  Case 1: a 3.5-year-old girl with primary fibrillary astrocytoma of the spinal cord. A Sagittal proton density-weighted MR image demonstrates bilobed cystic mass extending from C6 to T3. B Three months later, axial proton density- and T2-weighted MR images reveal cystic lesions of the cerebellum, pons, and medial temporal lobes. C Ten months later, sagittal T1-weighted MR image with contrast enhancement shows non-enhancing cystic lesions primarily of the cerebellum, but also of the brainstem and inferior frontal lobes. Leptomeningeal enhancement of the basal cisterns and spinal canal is also demonstrated. D Four years later, axial T2-weighted MR image demonstrates progression of diffuse cysts of the inferior and medial frontal and temporal lobes, cerebellum, and brainstem.

Shortly after, acute spinal tenderness and truncal rigidity led to a spinal MRI being taken, which demonstrated a bilobed cystic mass extending from C6 to T3 (Fig. 1A). Subtotal resection demonstrated fibrillar astrocytoma. Post-surgical neurological examination demonstrated only mild right hemiparesis. No further therapy was administered and the first post-operative MRI showed almost complete resolution.

Three months later, follow-up examination revealed a spongy-like appearance of the cerebellum and of the mesencephalic region (Fig. 1B), along with possible growth of the primary cord lesion. Extensive infectious and autoimmune disease work-ups were negative. CSF cytology findings were repeatedly negative. CSF protein progressively increased (from 90 mg/dl to 988 mg/dl in 9 months). However, as there was no diagnosis, the decision was made to follow the patient, who remained stable with repeated shunt revisions.

MRI 10 months later demonstrated progression of the non-enhancing cyst-like lesions, which surrounded and extended into the cerebellum, the brainstem, the medial temporal lobes, and the frontal lobes. There was also linear and nodular enhancement of the basal cisterns (Fig. 1C), the supratentorial leptomeninges, and of the spine. A cerebellar biopsy was alternatively interpreted as “possible neoplastic infiltration by glial cells,” or “non-specific cerebellar degeneration with lymphocytes.” Findings from CSF cytology remained persistently negative. However, since enlargement of the spinal lesion was also noted, diagnosis of disseminated spinal glioma was made, and systemic chemotherapy was begun. This resulted in clinical deterioration accompanied by progression of brain and spine findings, and therapy was withheld after 3 months. Radiation therapy was not performed.

Over the next 4 years, the patient’s MRI studies demonstrated continued progression in size and extent of the primary cord tumor, of cyst-like lesions (Fig. 1D), and of diffuse leptomeningeal