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Diffuse Leptomeningeal Glioneural Tumor

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Case Summary

A child with a history of hydrocephalus and seizures presented with fever and headaches of several weeks' duration. Computed tomography (CT) without and with contrast, and with contrast-enhanced magnetic resonance imaging (MRI) of the head were performed.

Imaging Findings

Noncontrast CT demonstrated a shunt catheter; there was no intracranial mass nor was there any evidence of hemorrhage. Subsequent MRI without and with contrast demonstrated multiple nodular hyperintense lesions within the posterior fossa involving the subpial space of the cerebellum and basal cisterns on the T2 sequence. The T1 postcontrast sequence revealed diffuse leptomeningeal enhancement within the cerebral hemispheres and greater enhancement within the suprasellar cistern, prepontine cistern, the basal cisterns, and cavernous sinuses. Sagittal T2 of the spine demonstrated a hyperintense lesion involving the intramedullary region of the thoracic cord. There was also diffuse leptomeningeal enhancement throughout the spine.

Diagnosis

Diffuse leptomeningeal glioneural tumor (DLGNT)

Discussion

Diffuse leptomeningeal glioneural tumor is a rare entity that has been described using numerous terms, including primary diffuse leptomeningeal oligodendroglioma and diffuse leptomeningeal oliodendroglimatosis. This has resulted in confusion, and the current WHO 2021 preferred term is diffuse leptomeningeal glioneuronal tumor.[1,2] CT may show only the secondary findings of the disseminated tumor, such as hydrocephalus. Hydrocephalus is secondary to tumor infiltration that obstructs the foramens of Luschka and Magendie, which are the outlets for cerebrospinal fluid flow from the fourth ventricle.

The major finding of leptomeningeal enhancement on MRI represents an obstruction caused by a tumor and requiring insertion of a shunt. Enhancement within the spine can vary from thin enhancement along the spinal cord and nerve roots, as well as thick, plaque-like, or nodular deposits and discrete masses. Nodular T2 hyperintense lesions within the subpial surface of the brain and spine are common. Intracranial enhancement is most conspicuous in the posterior fossa and basal region, with a sheet-like coating of the cerebellum and brainstem.

One study has described a dominant intraparenchymal lesion in 81% of patients and which was most common in the spinal cord (57%), followed by the cerebral cortex and brainstem.[3] In addition to postcontrast imaging of the brain, postcontrast imaging with gadolinium-based contrast agents of the spine is important to demonstrate the enhancing intramedullary lesion within the spinal cord and plaque-like enhancement throughout the cord.

These findings combined are most consistent with DLGNT, which requires biopsy for confirmation.[4] In the 2021 World Health Organization Classification, DLGNT is a low-grade tumor.[5] However, a subset of the DLGNT has anaplastic features and molecular alterations correlating with shorter survival.

Conclusion

Imaging of DLGNT typically reveals leptomeningeal enhancement within the brain and spine, as well as nodular T2 hyperintense lesions within the subpial surface of the brain. Enhancement is most conspicuous in the posterior fossa and basal region, with sheet-like coating of the cerebellar folia and brainstem. In cases with diffuse leptomeningeal enhancement at the basal cisterns with T2 hyperintense lesions within the posterior fossa and the subpial space of cerebellum, postcontrast T1 sequences of the spine should be performed. If the dominant lesion is within the spinal cord, this supports the diagnosis of DLGNT, since that is the most common location of the dominant lesion in DLGNT.

References

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