Primary leptomeningeal gliomatosis

Primary leptomeningeal gliomatosis (PLG) is a poorly recognized tumor of the central nervous system.

Leptomeningeal gliomatosis is a known, yet uncommon, complication of malignant gliomas. In rare instances it can present with non-specific symptoms prior to the development of detectable intraparenchymal lesions, posing a diagnostic challenge. Gliomatosis cerebri is also a rare disease, characterized by extensive diffuse infiltration of neoplastic glial cells. For both entities, limited data exist to guide treatment and prognosis is poor.

They often presents with symptoms and physical findings of chronic inflammatory meningitis and raised intracranial pressure, and lacks specific clinical, radiologic, and diagnostic criteria.

Case series

2016

Results of a multicentric retrospective study of 6 PLG cases (3 pediatric and 3 adult) were compared with literature data.

The mean age was 54.7 years for adults and 8.7 years for children, with 3 males and 3 females. Clinical symptoms were nonspecific. Cerebrospinal fluid analyses showed a high protein level often associated with pleocytosis but without neoplastic cells. On neuroimaging, diffuse leptomeningeal enhancement and hydrocephalus were observed, except in 1 case. PLG was mostly misinterpreted as infectious or tumoral meningitis. The first biopsy was negative in 50% of cases. Histopathologically, PLG cases corresponded to 1 oligodendroglioma without 1p19q codeletion and 5 astrocytomas without expression of p53. No immunostaining for IDH1R132H and no mutations of IDH1/2 and H3F3A genes were found. Overall survival was highly variable (2-82 months) but seems to be increased in children treated with chemotherapy.

This study shows the difficulties of PLG diagnosis. The challenge is to achieve an early biopsy to establish a diagnosis and to begin a treatment, but the prognosis remains poor. PLG seems to have a different molecular and immunohistochemical pattern compared with intraparenchymal malignant gliomas [1)].

Case reports

2010

A patient who presented with symptoms of increased intracranial pressure and diffuse leptomeningeal enhancement in the brain and spinal cord on MRI. After a period of surveillance, intraparenchymal lesions developed in association with widespread diffuse infiltration. The diagnosis of gliomatosis cerebri with diffuse leptomeningeal gliomatosis was established in hindsight. Initial treatment consisted of six cycles of temozolomide chemotherapy. Following radiological progression, the patient received craniospinal radiotherapy. Four months later the patient's symptoms had resolved and MRI demonstrated near complete response of leptomeningeal enhancement and intraparenchymal

lesions. Six months after radiotherapy, the patient remains clinically well without radiographic recurrence [2].

2009

A case diagnosed post-mortem, highlighting the diagnostic difficulty in identifying PDLG as the cause of chronic meningitis, even when a neoplastic etiology is suspected. Because multiple cytologies and even a leptomeningeal biopsy did not reveal the diagnosis ante-mortem, we emphasize the consideration of multi-site or repeat leptomeningeal biopsy when a persistent inflammatory infiltrate is found and neurological symptoms are progressive [3].

1)
Tauziede-Espariat A, Maues de Paula A, Pages M, Laquerriere A, Caietta E, Delpont B, Viennet G, Medeiros de Bustos E, Moulin T, Barnerias C, Vauleon E, Grill J, Chiforeanu D, Vasiljevic A, Varlet P. Primary Leptomeningeal Gliomatosis in Children and Adults: A Morphological and Molecular Comparative Study With Literature Review. Neurosurgery. 2016 Mar;78(3):343-52. doi: 10.1227/NEU.0000000000001028. PubMed PMID: 26397750.
2)
Knox MK, Ménard C, Mason WP. Leptomeningeal gliomatosis as the initial presentation of gliomatosis cerebri. J Neurooncol. 2010 Oct;100(1):145-9. doi: 10.1007/s11060-010-0138-4. Epub 2010 Feb 10. PubMed PMID: 20146082.
3)
Ko MW, Turkeltaub PE, Lee EB, Gonatas NK, Volpe NJ, Moster ML, Galetta SL. Primary diffuse leptomeningeal gliomatosis mimicking a chronic inflammatory meningitis. J Neurol Sci. 2009 Mar 15;278(1-2):127-31. doi: 10.1016/j.jns.2008.11.026. Epub 2009 Jan 8. PubMed PMID: 19135216.