Chordoid glioma of the third ventricle

Chordoid glioma of the third ventricle located exclusively in the mid-anterior portion of the third ventricle along the anterior aspect of the hypothalamic wall near the hypothalamus and optic nerves, with the histological features of a chordoma and immuno-labeling for glial fibrillary acidic protein [1].

The term “chordoid glioma” was introduced to denote a circumscribed, apparently, low-grade neoplasm arising in or preferentially involving the third ventricle of middle-aged women [2].

Epidemiology

A rare (only ≈ 80 case reports). Primarily a tumor of adulthood. Female:male ratio=3:1. Solid, enhancing mass on imaging. Mitotic activity is absent in most. GFAP immunostaining is common, S100 reactivity is variable. Histologically similar appearing to chordoid meningioma, which lacks GFAP staining. Attachment to the wall of third ventricle (hypothalamus) may prevent total removal.

Diagnosis

The MR appearances possess unique features which are very helpful in diagnosis of chordoid glioma, but to make a definite diagnosis is based on clinical and histopathological features and immunohistochemical staining [3].

Characterized by clusters and chords of epithelial cells expressing GFAP, within a mucinous stroma often containing a lymphoplasmacytic infiltrate.

Treatment

Low dose gamma knife radiosurgery after less invasive microsurgery is both safe and effective for the control of chordoid glioma of the third ventricle over a very long follow-up period [4].

Outcome

Slow-growing, noninvasive, benign tumor (WHO grade II)

1)
Al-Zubidi N, McGlynn MM, Chévez-Barrios P, Yalamanchili S, Lee AG. Neuro-Ophthalmologic Features of Chordoid Glioma. J Neuroophthalmol. 2013 Oct 9. [Epub ahead of print] PubMed PMID: 24113763.
2)
Vajtai I, Varga Z, Scheithauer BW, Bodosi M. Chordoid glioma of the third ventricle: confirmatory report of a new entity. Hum Pathol. 1999 Jun;30(6):723-6. PubMed PMID: 10374785.

3)

Shi HY, Tian Y, Shi YZ, Wang JD, Lu GM. [The MRI findings in chordoid glioma of the third ventricle]. Zhonghua Yi Xue Za Zhi. 2013 Jan 29;93(5):373-5. Chinese. PubMed PMID: 23660212.

4)

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