Ventricular Liponeurocytoma

Epidemiology

Al-Umran et al. performed a systematic literature review using the Pubmed, Scopus, and Cochrane Library database for all cases of lateral ventricular liponeurocytoma.

The described cases from 1997 onward include 14 cases that have been published in full papers in the English literature. Six additional cases are reported in short English abstracts in full non-English papers, and one case was described in a central neurocytoma report. There is a definite male predominance of 70% (14 male) and a mean age of 37 years (range 24-62)¹⁾.

Diagnosis

Heterogenous enhancement and signals in magnetic resonant images (MRI) are the radiological characteristics.

In all reported cases, the presence of lipocytes and fat vacuoles is considered the paramount histopathological feature $^{2)}$.

Treatment

Only two cases (including ours) received radiation therapy. Recurrence was seen in two patients during follow-up that was treated by radiation therapy in one and surgery in the other. The proliferation index is mostly below 5% in all cases, with the Ki-67 range between < 1% to 10%.

Lateral ventricular liponeurocytoma has been treated effectively by surgical resection in a limited number of cases. The decision for radiation therapy is based on a high proliferation index and tumor recurrence.

Total surgical resection was achieved in 80% (12 out of 15) of the cases $^{3)}$.

Systematic reviews

2021

Al-Umran MR, Al-Umran SR, Arab AF, Anas Dababo M, Alotaibi FA. Lateral Ventricular Liponeurocytoma: Review of Literature and Case Illustration. Neurochirurgie. 2021 Mar 22:S0028-3770(21)00074-6. doi: 10.1016/j.neuchi.2021.03.004. Epub ahead of print. PMID: 33766564 ⁴⁾.

2018

https://www.sciencedirect.com/science/article/pii/S2214751916301402

Case reports

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Two patients with intraventricular liponeurocytomas in Beijing Tiantan Hospital between July 2000 and July 2016. The main clinical manifestations of the two patients were headache. The supratentorial intraventricular liponeurocytoma appeared as isodense to slight hyperdense on CT scan and heterogeneous intensity on T1-weighted imaging (T1WI) and T2-weighted imaging (T2 weighted image). The plaque-like hypodense on CT images and hyperintensity on T1WI resembling fat could be seen inside the tumor. The liponeurocytoma located in the fourth ventricle showed isointensity on T1 and T2 weighted image as well as slight enhancement on contrast. Two patients accepted gross total resection of tumors. Two intraventricular tumors demonstrated similarly histopathological features, such as isomorphic small tumor cells with clear cytoplasm, sheets of monomorphic round cells and focal lipomatous differentiation. In addition, expression of synaptophysin, neuron specific enolase, microtubule-associated protein 2 and S-100 were found. No radiological or clinical evidence of recurrence of the tumors was observed in their follow-up surveys. In conclusion, intraventricular liponeurocytoma has a favorable clinical course, radiological features may be useful in the diagnosis of this rare tumor before surgery ⁵.

Kuchelmeister et al. in 2006 reported a liponeurocytoma of the left lateral ventricle in a 35-year-old man represents the fifth recorded case of a supratentorial intraventricular liponeurocytoma. In this location, liponeurocytomas are very exceptional, whereas it is the typical site for classic central neurocytomas.

Conversely, neurocytomas of the cerebellum are predominantly liponeurocytomas with until now more than 25 reported cases. Thus, cerebellar liponeurocytoma is the most frequent neuroepithelial CNS tumor with adipose-like cells followed by ependymomas with a lipid component and supratentorial intraventricular liponeurocytoma. Adipose-like cells in neurocytomas may originate by lipidization of tumor cells, metaplastic transformation of neuroectodermal cells into fat cells or by true adipocytic differentiation. The case showed also focal glial differentiation with GFAP-positivity of some tumor cells as often seen in cerebellar liponeurocytomas but much rarer in central neurocytomas. Pathogenetic and nosologic implications of supratentorial intraventricular liponeurocytomas are discussed. Future WHO tumor classification should consider that liponeurocytomas are not restricted to the cerebellum. Reports on cerebellar liponeurocytomas with a less favorable clinical course suggest a WHO grade II for liponeurocytomas ⁶.

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Xu N, Cai J, Du J, Yang R, Zhu H, Gao P, Zhou J, Li X. Clinical features and prognosis for intraventricular liponeurocytoma. Oncotarget. 2017 Mar 8;8(37):62641-62647. doi: 10.18632/oncotarget.16024. PMID: 28977976; PMCID: PMC5617536.

Kuchelmeister K, Nestler U, Siekmann R, Schachenmayr W. Liponeurocytoma of the left lateral ventricle-case report and review of the literature. Clin Neuropathol. 2006 Mar-Apr;25(2):86-94. PMID: 16550742.

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