Cerebellar liponeurocytoma

Liponeurocytomas are rare and slow-growing tumours located predominantly in the cerebellum.

In 2000, the World Health Organization (WHO) Pathology and genetics of tumours of the nervous system described cerebellar liponeurocytoma as a distinct entity from medulloblastoma in terms of prognostic, epidemiological and clinical aspects. This rare tumour is WHO grade I–II, generally with an accordingly indolent behaviour. Since 1978, when liponeurocytoma was first described, there have been more than 40 reported cases.

Epidemiology

Patel et al., in 2009 found 42 cases of liponeurocytoma reported in the literature. The patients' ages ranged from 4 to 69 years, with a median of 49 years. Ten (24%) of the lesions were found in patients aged 30 years and younger, and 32 (76%) of the lesions were found in patients older than 30 years. There were 22 female (52%) and 20 male (48%) patients. Thirty-six lesions (86%) were found within the cerebellum, and 6 (14%) were found in a supratentorial location. The average follow-up for patients undergoing surgery for liponeurocytoma was 48 (range 0–192) months. There have been a number of recurrences, with a mean time from diagnosis to first local recurrence presentation of 10.6 (range 10–12) years.

Pathology

They are characterized by many lipidized cells found in clusters or scattered between small neoplastic cells. Microscopically, liponeurocytoma has small, round to ovoid cells with scanty eosinophilic cytoplasm. Lipidized cells resemble mature adipocytes and are found interspersed throughout the lesion.

Immunohistochemical staining has demonstrated that both neuronal and glial differentiation is present. Mitotic activity is generally low in these lesions.

Cytogenetic Findings

see Cerebellar Liponeurocytoma: Relevant Clinical Cytogenetic Findings [1].

Treatment

Surgical resection is recommended as the initial treatment modality for patients with liponeurocytoma to establish a diagnosis. The rare occurrence and diagnosis of this type of tumour as well as its variable appearance on imaging can make radiological diagnosis challenging.

There is no consensus regarding the treatment of liponeurocytoma, specifically whether chemo- or radiotherapy is a necessary part of the postoperative treatment regimen [2].

Patel et al., recommend that longer follow-up periods are required. Most follow-ups in the literature are shorter than 5 years, a period during which the tumour may still be in a silent phase, starting to regrow. However, rather than exposing the patients to the risks and side-effects of radiotherapy, without any evidence to support its usefulness in preventing recurrence, they recommend reoperating on a recurrent tumour, which has only a slightly more aggressive histology than the primary presentation, with the option of using adjuvant radiotherapy at this time. Further studies regarding the natural history of this lesion are warranted [3].

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Tucker A, Boon-Unge K, McLaughlin N, Ibrahim H, Rao N, Martin N, Everson R, Khanlou N. Cerebellar Liponeurocytoma: Relevant Clinical Cytogenetic Findings. J Pathol Transl Med. 2016 Oct 16. doi: 10.4132/jptm.2016.07.24. PubMed PMID: 27750407.
2)
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2724827/
3)
Patel N, Fallah A, Provias J, Jha NK. Cerebellar liponeurocytoma. Can J Surg. 2009 Aug;52(4):E117-E119. PubMed PMID: 19680499; PubMed Central PMCID: PMC2724827.