CASE NOTE

Cerebellar liponeurocytoma

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Liponeurocytomas are rare and slow-growing tumours located predominantly in the cerebellum.[1] In 2000, the World Health Organization (WHO) Pathology and genetics of tumours of the nervous system[2] 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.[2]

Liponeurocytomas occur generally in the cerebellum and are characterized by many lipidized cells found in clusters or scattered between small neoplastic cells. Immunohistochemical staining has demonstrated that both neuronal and glial differentiation is present. Mitotic activity is generally low in these lesions.[3]

As liponeurocytoma is a newly defined clinical entity with relatively short follow-up data, there is a lack of consensus in the literature with respect to its epidemiology and prognosis as well as management strategies. In this report, we present the case of a woman with a liponeurocytoma, review the literature and make suggestions for management.

Case report

A 40-year-old woman presented with a 1-year history of worsening, nonspecific headaches. Although the results of her neurologic exam were unremarkable, computed tomography (CT) and magnetic resonance imaging (MRI) scans showed a 3.7 cm × 3.6 cm relatively ill-defined, poorly enhancing, round lesion in the right cerebellum that extended into the cerebellopontine angle region, causing moderate mass effect (Fig. 1). After complete microscopic tumour resection, pathologic examination revealed a cellular neoplasm composed of small cells that were round to polygonal in shape with some lightly eosinophilic cytoplasm, oval to round nuclei and a generally open nuclear chromatin pattern. The tumour cells, in areas, were mixed with lipomatous cells (Fig. 2). Immunohistochemical analysis was diffusely positive for neuronal markers synaptophysin, neuron-specific enolase and tau. The lipocytes were also positive for S100. The tumour cells were largely negative for glial fibrillary acidic protein stains for astrocytes. These pathology features were consistent with liponeurocytoma. In 3 years of postoperative follow-up at the regional cancer centre, the patient has received no adjuvant treatment and there has been no evidence of tumour recurrence.

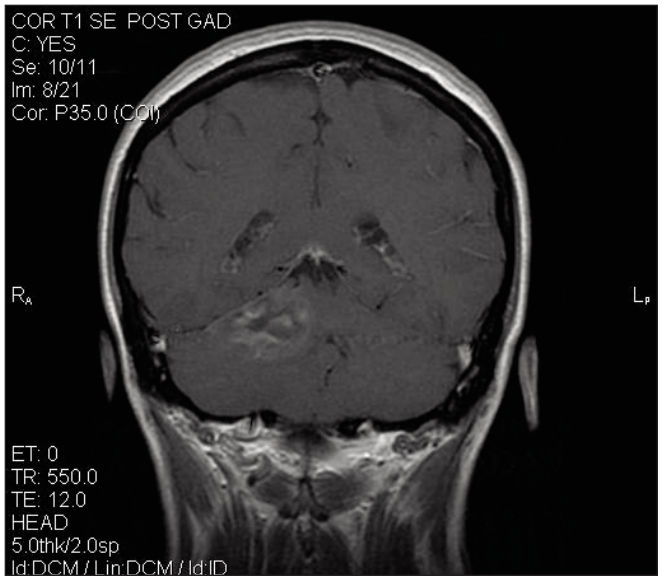

Fig. 1. T1-weighted coronal magnetic resonance imaging scan with gadolinium shows a fairly large well demarcated right cerebellopontine angle mass with heterogenous contrast enhancement.

Discussion

We 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.[4]

Microscopically, liponeurocytoma has small, round to ovoid cells with scanty eosinophilic cytoplasm. Lipidized cells resemble mature adipocytes and are found interspersed throughout the lesion.[4]

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.[5]

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

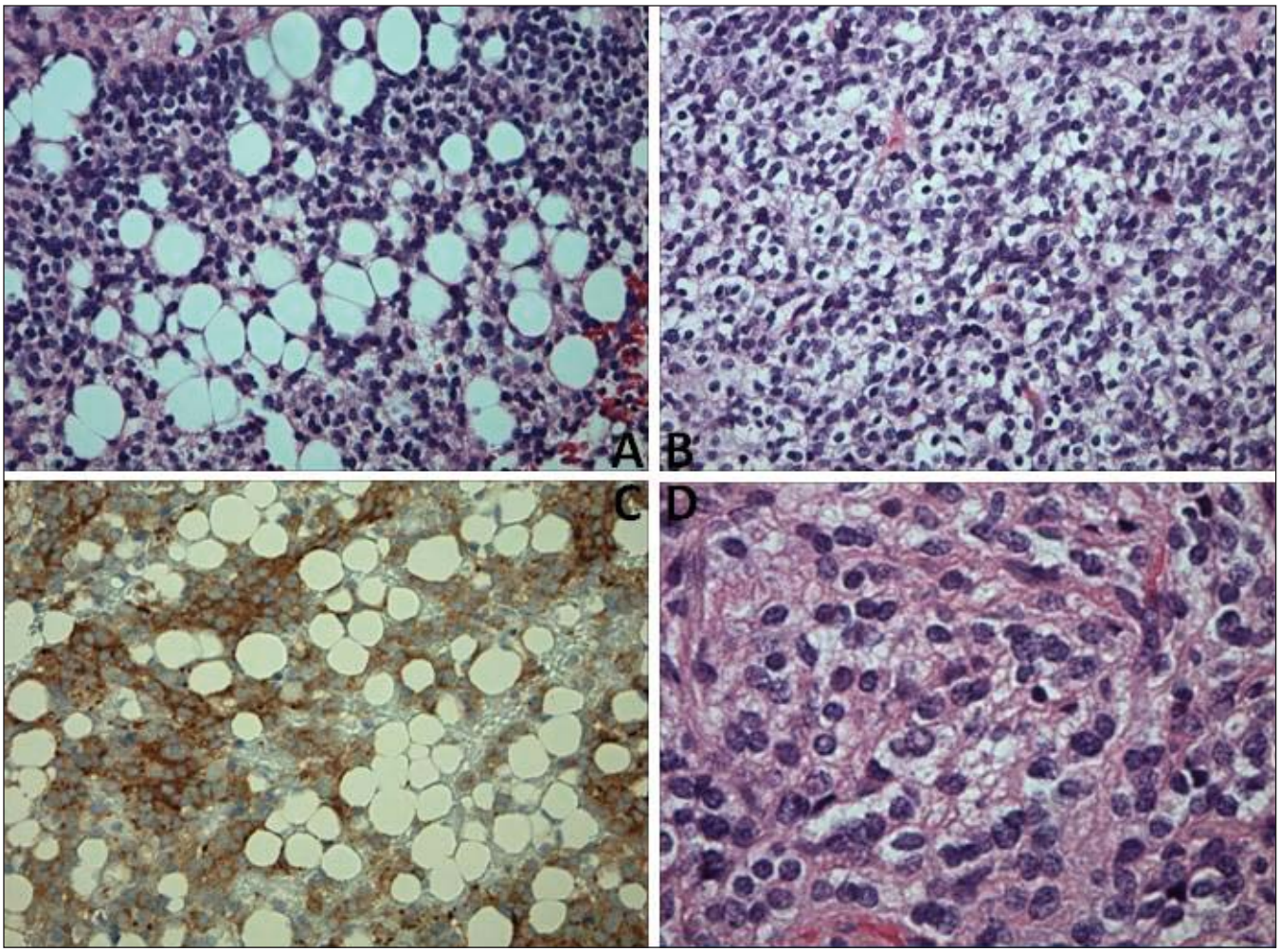

Fig. 2. Histology results showing (**A**) a typical field of liponeurocytoma (note the lipomatous cells admixed with neurocytes), (**B**) a neurocytic-predominant area of the tumour (hematoxylin and eosin stain, original magnification ×200), (**C**) strong positivity of the neurocytes, whereas the admixed lipomatous cells are immunonegative (synaptophysin immunohistochemistry, original magnification ×200) and (**D**) bland regular nuclei with open chromatin (hematoxylin and eosin stain, original magnification ×400).

References

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