Cerebellar liponeurocytoma (LPN) is a rare central nervous system tumour recently recognized as a distinct clinicopathological entity. It was included in the previous 2000 edition of the World Health Organization (WHO) classification as a grade I neoplasm under the heading of glioneuronal tumours. With increasing recognition of this entity and longer clinical follow-up periods in its investigation, it has become clear that this tumour has a rate of recurrence that is higher than previously thought and not compatible with a grade I designation. In light of this, the current 2007 WHO classification assigns the cerebellar liponeurocytoma to WHO grade II tumour. In this paper, we report a new case of a recurrent cerebellar LPN with supratentorial extension despite consistently benign histological features. To the best of our knowledge, recurrent cerebellar LPN with supratentorial extension has never been reported before, as the nine cases of recurrent cerebellar LPN published to date were confined to the posterior fossa.

CLINICAL HISTORY

A 42-year-old previously healthy man, presented with a five-month history of headaches, vertigo, visual disturbance and progressive unsteadiness of gait. On admission, neurological examination showed signs of cerebellar dysfunction, including ataxia, wide-based gait, right dysmetria on finger-to-nose testing and a positive Romberg sign. The fundus examination revealed unilateral papilloedema. Computed tomogram scan (Figure 1) demonstrated a hypodense lesion within the vermis and right cerebellar hemisphere compressing the 4th ventricle and enhancing after contrast injection. Magnetic resonance imaging (MRI) scan showed a heterogeneous mass measuring 5 × 4 cm, characterized by discrete hyperintensity on T1-weighted images and high signal intensity on T2-weighted images. Several central parts of the lesion were very hypointense on both T1 and T2-weighted sequences suggesting fat content. An intra-operative frozen section was reported as being consistent with a diagnosis of cerebellar liponeurocytoma. A post-operative MRI performed three days after the surgery, demonstrated no residual tumour. No adjuvant radiation therapy was administered post-operatively. At the last routine follow-up visit (ten years post-operatively), the patient complained of recurrent headaches and vertigo. Magnetic resonance imaging revealed a vermal mass with bilateral paramedian and supratentorial extension (Figure 3). The patient underwent further tumour resection without adjuvant radiotherapy. The histological examination of the surgical specimen showed no features suggesting that the tumour had undergone progression or contained additional elements. Furthermore, microvascular proliferation and necrosis were absent and the proliferative fraction remained small (<3%). The patient experienced an uneventful post-operative recovery with no evidence of recurrence one year after the second surgical intervention.

DISCUSSION

There is scant knowledge regarding the natural history and the optimum treatment strategy of cerebellar LPN. Twenty-nine cases of cerebellar LPN cited on indexed articles so far, cannot represent a significant number of patients on which to base treatment. The rarity of this tumour and paucity of pertinent information regarding its biological potential and natural history have resulted in the application of various treatment modalities. The questions remaining are how aggressive removal should be...
and whether to expose the patient to postoperative treatment such as chemo- or radiotherapy or not. Furthermore, the role of irradiation is still a matter of debate: does it prevent recurrences or stabilize putative tumour residues? According to Jackson et al, resection and adjuvant radiotherapy to the posterior fossa seem to be the best approach to reduce the local recurrence rate of cerebellar LPN. On the other hand, Cacciola et al stated that complete macroscopic resection with long-term follow-up seems to be the most appropriate treatment. If tumour recurrence does occur, then additional surgery, followed by fractionated radiotherapy, may be necessary. In our patient, gross total resection of the primary tumour was performed without adjuvant radiation therapy and he experienced tumour recurrence ten years post-operatively. The basic question in this case is whether radiotherapy would have prevented supratentorial extension if it had been initially administered. In an attempt to put forward a hypothesis on the recurrence pattern of cerebellar LPN, we undertook a literature review of recurrent cerebellar LPN. Nine cases have been published in the literature to date (Table) which gives us a recurrence rate of approximately 31%. Patients

Figure 1: A) Axial MRI (at the initial presentation), showing a right hemispheric and vermian cerebellar lesion with mass effect on the fourth ventricle. B) Sagittal MRI (at the initial presentation) demonstrating a heterogeneous right hemispheric cerebellar lesion with hyperintense streaks due to the adipose content.

Figure 2: A) Isomorphic neurocytic elements with focal lipomatous differentiation (Hematoxylin & eosin, original magnification x 100) B) Tumour cells and lipidized cells demonstrating synaptophysin immunopositivity (Immunohistochemistry, original magnification x 400)
were aged between 36 and 64 years with a male predominance (seven males, two females). All of the cases of recurrent cerebellar LPN underwent surgical resection of the primary tumour. Gross total resection was performed in eight cases and subtotal resection in only one case. The latter was the unique patient to receive post-operative radiotherapy; however, he developed tumour recurrence 12 months post-operatively. Except for this case, cerebellar LPN generally recurs after a long period of time ranging from 1 year to 12 years (mean = 8.5 years). A second recurrence, however, seems to occur following a shorter lapse of time ranging from 3 months to 5 years (mean = 2.5 years). All the reported cases of recurrent cerebellar LPN were confined to the posterior fossa without supratentorial extension. Our patient was the first to develop tumour recurrence 12 months post-operatively. Except for this case, cerebellar LPN generally recurs after a long period of time ranging from 1 year to 12 years (mean = 8.5 years). A second recurrence, however, seems to occur following a shorter lapse of time ranging from 3 months to 5 years (mean = 2.5 years). All the reported cases of recurrent cerebellar LPN were confined to the posterior fossa without supratentorial extension. Our patient was the first to develop tumour recurrence 12 months post-operatively. This is in contrast to the case reported by Buccoliero et al that showed histopathological aggressive features (mitoses pleomorphism, moderate endothelial proliferation and a high proliferation index as evaluated by MIB-1) in the primary lesion that the biological behaviour of cerebellar liponeurocytomas is much more benign than that of the highly aggressive medulloblastomas.

In summary, we reported a new case of a recurrent cerebellar LPN with supratentorial extension despite persistently benign histological features. We conclude that there is not always a correlation between histological features and biological behaviour of this neoplasm. The number of cases of cerebellar

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**Table: Cases of recurrent cerebellar liponeurocytoma published in the literature**

<table>
<thead>
<tr>
<th>Author / year</th>
<th>Age / gender</th>
<th>Tumour location</th>
<th>Treatment</th>
<th>Follow-up - Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Giangaspero (1996)[8]</td>
<td>36 / M</td>
<td>Lt hemis</td>
<td>GTR</td>
<td>*Recurred 10 yrs post-op- Reop *Recurrence 5 yrs after 2nd resection – Reop - ANR</td>
</tr>
<tr>
<td>Buccoliero (2005) [11]</td>
<td>64 / M</td>
<td>Rt hemis</td>
<td>GTR</td>
<td>* Recurrence 3&amp;1/2 years post-op Reop + intra-op RT NED 5 months post-op</td>
</tr>
<tr>
<td>Present case (2008)</td>
<td>42 / M</td>
<td>Vermis &amp; Rt hemis</td>
<td>GTR</td>
<td>** Recurrence 10 years post-op NED 1 yr after 2nd resection.</td>
</tr>
</tbody>
</table>

ANR: alive and no recurrence; CPA: cerebellopontine angle; F: female; GTR: gross total removal; Hemis: hemisphere; Intra-op: intra-operative; LPN: liponeurocytoma; Lt: left; M: male; NED: no evidence of disease; Post-op: post-operatively; Rt: right; Reop: reoperation; RT: radiotherapy; STR: subtotal removal; Yrs: years; *: Recurrence limited to the posterior fossa; **: Recurrence with supratentorial extension.

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In summary, we reported a new case of a recurrent cerebellar LPN with supratentorial extension despite persistently benign histological features. We conclude that there is not always a correlation between histological features and biological behaviour of this neoplasm. The number of cases of cerebellar

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In summary, we reported a new case of a recurrent cerebellar LPN with supratentorial extension despite persistently benign histological features. We conclude that there is not always a correlation between histological features and biological behaviour of this neoplasm. The number of cases of cerebellar
LPN reported to date is small and data are unfortunately incomplete; that’s why it is important to continue recording cases with longer follow-up periods so as to better understand the natural history of this neoplasm and determine its optimal treatment strategy.

REFERENCES