Bilateral cerebellopontine angle lipomas in an infant with encephalocele: illustrative case

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BACKGROUND Bilateral cerebellopontine angle (CPA) lipomas are extremely rare. Herein the authors present a case of bilateral CPA lipomas in an infant along with a literature review of bilateral CPA lipomas.

OBSERVATIONS A newborn girl was incidentally found to have bilateral CPA lipomas during the workup for an occipital encephalocele. The encephalocele was repaired primarily on day 2 after birth. The patient demonstrated no symptoms associated with the bilateral CPA lipomas. Eight cases of bilateral CPA lipomas were identified in the literature review and are summarized. Conservative management is the consensus strategy, given minimum growth of the tumor and the high risk of surgical intervention.

LESSONS This is the first reported case of bilateral CPA lipomas in an infant as well as the first with a coexisting intracranial malformation. Intracranial lipomas share an extremely low growth rate and typically do not cause severe symptoms. The management of asymptomatic or mildly symptomatic bilateral CPA lipomas is usually conservative.

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KEYWORDS case report; cerebellopontine angle; encephalocele; lipoma; pediatric neurosurgery

Bilateral cerebellopontine angle (CPA) lipomas are an extremely rare condition; only 8 cases have been reported in the literature.1–7 Here we report a case of asymptomatic bilateral CPA lipomas in a newborn, which were diagnosed incidentally during the workup for an occipital encephalocele. This is the first case of bilateral CPA lipomas in an infant as well as the first case with a coexisting intracranial anomaly. A review of the literature in PubMed was also conducted using the keywords “bilateral” and “cerebellopontine lipoma” or “CPA lipoma.” All articles describing cases of bilateral CPA lipomas were identified and summarized.

Illustrative Case

A newborn girl was found to have an occipital encephalocele at birth (Fig. 1A). An ultrasound revealed bilateral CPA hyperechoic lesions (Fig. 1B) in addition to the encephalocele. Subsequent magnetic resonance imaging (MRI) of the brain without contrast demonstrated bilateral CPA lipomas on both T2-weighted (Fig. 1C) and T1-weighted (Fig. 1D) imaging, measuring 0.8 × 2 cm (left) and 0.6 × 1.3 cm (right) in size. The bilateral facial-vestibulocochlear complexes were found encased within the lipomas (Fig. 1D). The encephalocele was also well visualized on sagittal T1-weighted (Fig. 1E) and axial T2-weighted MRI (Fig. 1C). The patient passed the newborn hearing screening and demonstrated no symptoms related to the bilateral CPA lipomas; therefore, no intervention was offered for the lipomas. Surgical repair of the encephalocele with primary closure was successfully performed on day 2 of life (Fig. 1F), as the lesion was not covered by skin. Chromosomal fluorescence in situ hybridization (FISH) test results were normal. She was safely discharged home on day 7 after birth with scheduled neurosurgery outpatient follow-up.

Literature Review

A total of 9 cases are summarized in Table 1, including 8 collected from the literature review and the case presented herein.1–7 The cohort includes 6 male (67%) and 3 female (33%) patients. With regard to symptoms, 5 cases were found incidentally (56%), 2 patients experienced tinnitus (22%), 1 patient reported frontal headaches (11%), and 1 case was identified during autopsy (11%). No surgical intervention was performed in any case.
Discussion

Observations

Bilateral CPA lipomas are an exceedingly rare condition. Intracranial lipomas are found in only 0.08% of cases in autopsy series and comprise 0.1%–1.5% of all intracranial tumors. The majority (45%) of all intracranial lipomas have been located at the corpus collosum area, followed by the quadrigeminal cistern (25%), suprasellar area (15%), and CPA area.

Among all intracranial lipomas, only 10% are located in the CPA region, furthermore, a systematic review of 219 CPA lipomas found that bilateral lipomas accounted for only 4% of all reported CPA lipomas with laterality available. Only 8 cases of bilateral CPA lipomas have been reported in the literature to date. The

TABLE 1. Summary of bilateral CPA lipomas in the literature

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Presentation</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wolbach and Millet, 1913</td>
<td>1</td>
<td>M</td>
<td>Autopsy</td>
<td>NA</td>
</tr>
<tr>
<td>Amonkar et al., 1997</td>
<td>2</td>
<td>M</td>
<td>Incidentally found</td>
<td>Conservative</td>
</tr>
<tr>
<td></td>
<td>15</td>
<td>M</td>
<td>Incidentally found</td>
<td>Conservative</td>
</tr>
<tr>
<td>Bigelow et al., 1998</td>
<td>14</td>
<td>F</td>
<td>Headaches</td>
<td>Conservative</td>
</tr>
<tr>
<td>Ergün et al., 2009</td>
<td>23</td>
<td>M</td>
<td>Incidentally found</td>
<td>Conservative</td>
</tr>
<tr>
<td>Gobel et al., 2010</td>
<td>70</td>
<td>F</td>
<td>Incidentally found</td>
<td>Conservative</td>
</tr>
<tr>
<td>Ventura et al., 2012</td>
<td>36</td>
<td>M</td>
<td>Tinnitus</td>
<td>Conservative</td>
</tr>
<tr>
<td>Bacciu et al., 2014</td>
<td>43</td>
<td>M</td>
<td>Tinnitus</td>
<td>Conservative</td>
</tr>
<tr>
<td>Present case</td>
<td>1*</td>
<td>F</td>
<td>Incidentally found</td>
<td>Conservative</td>
</tr>
</tbody>
</table>

CPA = cerebellopontine angle; NA = not applicable.
* Age listed in days.
present report is the first case of bilateral CPA lipomas in an infant with imaging, as well as the first reported case with an additional intracranial anomaly, which was an occipital encephalocele. A literature review of CPA lipomas published in 2009 revealed a male dominance pattern, with a male-to-female ratio of 2:1, which is similar to the bilateral CPA lipoma cohort in the present study (67% male). However, a recent literature review including more comprehensive data on CPA lipomas reported a nearly equal gender distribution (53% male).

There has been ongoing discussion of the neural embryological association between intracranial lipomas and developmental disorders, and the pathogenesis of intracranial lipomas remains debatable. The theory of tumorgenesis is no longer being considered, given that neurovascular structures are typically incorporated into intracranial lipomas rather than being displaced by the lesion, suggesting that intracranial lipomas are likely a congenital anomaly. Furthermore, no histopathologic evidence of cellular atypia has been reported. A theory similar to the development of intraspinal tentorial lipomas has been proposed. This theory potentially explains the huge co-occurrence of intracranial lipomas with other intracranial malformations. Finally, a theory of “maldifferentiation of meninx primitive” (primitive meninges) has emerged as the most widely accepted thus far. This theory, intracranial lipomas are thought to result from failure of the pathologically persistent meninx primitiva, a mesenchymal derivative of the neural crest that gives rise to the subarachnoid cisterns and is subsequently reabsorbed. The pathologically persistent meninx primitiva maldifferentiates into adipose tissue, giving rise to intracranial lipomas. This theory accounts for the frequent cisternal location of intracranial lipomas and the lipomas’ association with other midline malformations, such as agenesis/dysgenesis of the corpus callosum or the absence of the septum pellucidum. Nevertheless, there is no definitive conclusion on the mechanism of intracranial lipoma formation and its embryological association. Although 55% of the supratentorial lipomas were associated with other brain malformations, no bilateral CPA lipoma case has been reported along with another malformation. This is the first report of the coexistence of bilateral CPA lipomas and an intracranial malformation, which was an occipital encephalocele.

All cases in the cohort were managed conservatively (Table 1). Rarely, growth was observed during surveillance (1.4% of all cases with only minimum growth). It is recommended that an audiogram be obtained annually for 5 and 10 years in adults and pediatric patients, respectively; brain MRI should be performed every 2–3 years or in cases of audiogram changes. Surgical intervention can be technically challenging and carries a high risk of neurological deficits, as the facial-vestibulocochlear complex is usually encased by the CPA lipomas. Additionally, surgical intervention has not been reported to provide improvement in symptoms. Thus, we recommend conservative management in such cases.

Lessons

We presented an extremely rare case of incidental bilateral CPA lipomas in a 1-day-old girl, who also had an occipital encephalocele. This is the first case of bilateral CPA lipomas in an infant and the ninth bilateral CPA lipomas case overall in the literature. Observation should be the first line of management in these cases, given the extremely low rate of growth and the relatively high risk of neurological deficits associated with surgical intervention.

References


Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Zhao. Acquisition of data: Gernsback, Zhao, Prather. Analysis and interpretation of data: Tavakol. Drafting of the article: Zhao, Prather, Tavakol. Critical revising the article: Gernsback, Prather, Tavakol. Reviewed submitted version of the manuscript: Gernsback, Prather, Tavakol. Approved the final version of the manuscript on behalf of all authors: Gernsback. Administrative/technical/material support: Tavakol. Study supervision: Gernsback.

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