Focal brain enhancing lesions of evanescent type in patients with Neurofibromatosis-1

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Authors: S. Vinhais¹, S. Nunes¹, D. Salgado²; ¹Lisboa/PT, ²Lisbon/PT
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Purpose

Serial MR examinations in patients with neurofibromatosis type-1 (NF-1) presenting optic nerve gliomas or other brain abnormalities, such as low grade-gliomas and "unidentified bright objects" (UBOs) [1], may disclose focal brain enhancing lesions (FBEL), worrisome at first glance, but thereafter assigned as "benign", as they show a pattern of evanescent type.

The rarity of these lesions, seldom described in the literature [2], and heterogeneity in their presentation [3], are obstacles to an easy understanding of their nature, thus representing a pathological category that might alarm clinicians and be an incognita for neuroradiologists.

We report a short series of these lesions, herein denominated FBEL.

Methods and Materials

Review of 3 cases, all NF-1 children with chiasmatic-hypothalamic gliomas and lesions of the described type - FBEL, evaluated regularly by brain MR at our institution.

The clinical data were collected and compared, and the imaging studies available analyzed from our PACS.

Results

Patients were 2 female and 1 male, all having NF-1 manifested by chiasmatic-hypothalamic gliomas, among other criteria of disease (mean age at NF-1 diagnosis 4.7y).

The girls presented new focal brain enhancement lesions classified as FBEL after being submitted to chemotherapy - protocol for low-gliomas (carboplatin and vincristine), both in location at the supratentorial white matter (mean time after QT 2.8y; 4.8y after NF-1 diagnosis). One lesion was left temporal, the other left frontal.

The boy, who did not received QT, had a similar abnormality in the cerebellum vermis (4.3y after NF-1 diagnosis).

Mean age of FBEL presentation in this small group of patients was 10y and maximum size of the lesions ranged from 7 to 11 mm on T1-WI with gadolinium.
Only one lesion was symptomatic, associated to seizures, corresponding to a left temporal cortico-subcortical abnormality, which the electroencephalographic studies proved to be in relation to the epileptogenic focus. Patient was treated medically with more than one anti-epileptic drug. The other two FBEL were asymptomatic.

In terms of evolution, the referred temporal FBEL reduced the size in 55% spontaneously, and remained represented by a rounded enhancement focus on T1-WI with gadolinium, also with expression on FLAIR/T2-WI. When characterized by diffusion imaging there was no signal restriction. The several attempts to obtain it´s spectral profiles for monitoring were all invalid though, because of artifacts related to the lesion’s location. The others FBEL evanesced by losing the enhancement completely, although one still recognizable on FLAIR/T2-WI, 80% smaller in comparison to it´s maximum size measured on T1-WI with gadolinium in previous studies.

Mean follow-up time of FBEL was about 4 years.

The results are summarized in the tables below:

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age at NF-1 diagnosis (years)</th>
<th>Main CNS disease</th>
<th>QT Age at FBEL diagnosis (years)</th>
<th>Time from QT (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>8</td>
<td>hypothalamiço chiasmatic glioma</td>
<td>13</td>
<td>26</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>3</td>
<td>hypothalamiço chiasmatic glioma</td>
<td>8</td>
<td>40</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>3</td>
<td>hypothalamiço chiasmatic glioma</td>
<td>8</td>
<td>#</td>
</tr>
</tbody>
</table>

Table 1 - Patients´ Features

<table>
<thead>
<tr>
<th>Case</th>
<th>FBEL´s size (on T1-WI with Gd)</th>
<th>Lesion Location</th>
<th>Presenting Symptoms</th>
<th>Treatment</th>
<th>FBEL Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11 mm</td>
<td>left temporal (cortico-subcortical)</td>
<td>seizures</td>
<td>antiepileptic drugs</td>
<td>65</td>
</tr>
</tbody>
</table>
Table 2 - FBEL’s Features

<table>
<thead>
<tr>
<th>Case</th>
<th>T1-WI with gadolinium</th>
<th>FLAIR/T2-WI</th>
<th>ADC/DWI (b=1000)</th>
<th>Spectroscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>focal enhancement lesion 55% smaller than initially</td>
<td>focal hyperintense lesion 70% smaller than initially</td>
<td>hyperintense signal on ADC, no restriction on DWI</td>
<td>uncertain</td>
</tr>
<tr>
<td>2</td>
<td>no expression</td>
<td>focal hyperintense lesion 80% smaller than initially</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>3</td>
<td>no expression</td>
<td>practically imperceptible</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>

Table 3 - Late Imaging Findings

Brief Discussion:

The reported lesions probably represent low-grade gliomas, different from those arising from the optic tract characteristically found in NF-1 patients. Unlike other NF-1 brain abnormalities called UBOs, FBEL are recognizable on T1-WI as hypointense focal lesions and enhance after gadolinium administration. In alternative, they may correspond to inflammatory/demyelinating lesions.

Curiously, all FBEL were detected 5 years after the initial diagnosis of NF-1, during the follow-up of patients in their late childhood/adolescence.

Not always asymptomatic, FBEL should be kept under imaging surveillance, with a special attention in the first 2-3 years (time window when they are expected to vanished), but we also advise a longer period till the adult age, when the grow changes cease.
Conclusion

These lesions under scope are intriguing. At first, some embarrassment comes from the enhancement feature, which separates FBEL from other differential entities recognized as of indulgent course, namely the UBOs.

Motoring by imaging is crucial to ensure that the detected abnormality is "benign", as in a simple way demonstrated in face of a spontaneous regression (even if not complete). In addition, it allows obtaining a precise imaging pattern, valuable for general comparison. It is hoppe that some of the recent MR techniques could help in the process of FBEL characterization, better separating them from other lesions more often observed in NF-1 patients. Some efforts were already done in the large field involved [4, 5, 6], but certainly it’s necessary to gather more cases and compare them properly to take more detailed conclusions.

References


Personal Information