Case Report

Bilateral Rasmussen's encephalitis associated with type II focal cortical dysplasia: Dormant 'second' epileptogenic zone in contralateral disease

Thomas Frigeri a, Marta Hemb a, Eliseu Paglioli a, João Rubião Hoefel a, Vinicius Silva a, Harry Vinters b,⁎, Andre Palmini a

a Porto Alegre Epilepsy Surgery Program, Hospital São Lucas da Pontifícia Universidade Católica do Rio Grande do Sul (PUCRS), Porto Alegre, Brazil
b Department of Pathology, UCLA, Los Angeles, USA

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A B S T R A C T
Rasmussen's encephalitis (RE) is an inflammatory, probably autoimmune disorder manifested by refractory seizures and progressive deterioration of one cerebral hemisphere [1]. Clinical suspicion arises when a patient with partial seizures or epilepsy partialis continua (EPC) also has (even mild) unilateral atrophy and no other lesion on MRI. The unilaterality of the pathological and clinical involvement has been a puzzle for epileptologists since the description of this disorder, and several theories have been proposed to accommodate this fact [2,3].

Increased awareness about RE led to reports of a few patients in whom this classically unilateral disease affected both hemispheres [5]. These bilateral cases are even more puzzling because they, in a sense, challenge any simplistic explanation for a unilateral pathogenesis [4]. One aspect which has not been specifically examined is whether bilateral RE may be more prone to be associated with another pathology (dual pathology).

Here, we describe the history of an unfortunate girl with a progressive disorder which, upon clinical, neuroimaging, and histopathological evaluation, proved to be bilateral RE associated with type II focal cortical dysplasia. Whether the second pathology is relevant for the extent of the disease is discussed.

1. Introduction

Rasmussen's encephalitis (RE) is an inflammatory, probably autoimmune disorder manifested by refractory seizures and progressive deterioration of one cerebral hemisphere [1]. Clinical suspicion arises when a patient with partial seizures or epilepsy partialis continua (EPC) also has (even mild) unilateral atrophy and no other lesion on MRI. The unilaterality of the pathological and clinical involvement has been a puzzle for epileptologists since the description of this disorder, and several theories have been proposed to accommodate this fact [2,3].

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Here, we describe the history of an unfortunate girl with a progressive disorder which, upon clinical, neuroimaging, and histopathological evaluation, proved to be bilateral RE associated with type II focal cortical dysplasia. Whether the second pathology is relevant for the extent of the disease is discussed.

2. Case report

This girl had an uneventful history until starting with partial motor seizures at age 4.5 years. The attacks involved the entire right hemibody, which accounted for the EPC on the right hemibody at presentation. In addition, there was unequivocal progressive cortical and subcortical atrophy of the right hemisphere, which accounted for the EPC on the left hemibody. This is highly compatible with RE (+/− FCD) in the right hemisphere as well. Although the association of FCD and RE – as well as the occasional occurrence of bilateral RE – has already been reported [3–5], this is the first such case in which bilateral RE and FCD co-occur.

⁎ Corresponding author. Tel.: +1 352 870 7660.
E-mail address: tfrigeri@terra.com.br (H. Vinters).
Fig. 1. Preoperative MRI showing the alterations described above. Note that there is no hint of pathology on the contralateral hemisphere.

Fig. 2. Intraoperative ECoG mapping the epileptogenic areas.

Fig. 3. A and B: Pathological specimen showing dysmorphic neurons. C: Pathological evidence of neuronal loss, gliosis, and perivascular cuffing.
was also cortical and subcortical atrophy of the right hemisphere, which was unequivocal upon comparison with the MRI performed a few years earlier (Fig. 4).

In extremis, she underwent functional hemispherectomy of the left hemisphere, complementing the initial resection, in the hope that this might interfere with disease progression. Nonetheless, left-sided EPC and cognitive deterioration persisted. She is currently tetraparetic, bedridden, and does not communicate. Pathology again confirmed RE (Fig. 5).

3. Conclusion

We demonstrated histopathological evidence of RE and type II FCD in the left hemisphere, which led to EPC on the right hemibody at presentation. In addition, there was unequivocal progressive cortical and subcortical atrophy of the right hemisphere, which accounted for the EPC on the left hemibody. This is highly compatible with RE (+/− FCD) in the right hemisphere as well. Although the association of FCD and RE − as well as the occasional occurrence of bilateral RE − has already been reported, this is the first such case in which bilateral RE and FCD co-occur.

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