Variant Creutzfeldt-Jakob Disease (vCJD) has been linked to Bovine Spongiform Encephalopathy (BSE) - a prion disorder in cows. The emergence of vCJD has been linked to an earlier epidemic of BSE in the United Kingdom through the consumption of contaminated beef (1). Since the first report in 1996, a total of 229 vCJD cases have been recorded worldwide: 177 in the United Kingdom; 27 in France; and 25 cases distributed in 10 countries, including the United States (1).

Methods

Using publicly available data from the Centers for Disease Control and Prevention (CDC), the United States Department of Agriculture (USDA), and case reports including the newest confirmed diagnosed vCJD case (May 2014), we reviewed the cases of BSE and vCJD that have been found in the United States. For the vCJD patients, we compared age at symptom onset, symptom duration, clinical presentation, year of diagnosis and type of tissue confirmation for diagnosis.

BSE Cases in the US

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>8.5 year old dairy cow, Washington State, Canadian import 2001</td>
<td>12 year old cow, Texas, born, 1st epidemic case</td>
<td>10 year old cow from Alabama, unknown origin</td>
</tr>
<tr>
<td>Strain</td>
<td>unknown</td>
<td>Atypical</td>
<td>H-type</td>
</tr>
</tbody>
</table>

Conclusions

- The 4 US cases of vCJD exhibited a similar presentation to vCJD cases worldwide. One notable finding in the most recent case is the presence of the cortical ribbon sign on MRI brain.
- There are 4 neuropathologically confirmed vCJD cases in the US, most recently in May 2014, and 4 confirmed BSE cases (1,2,3,5).
- There is no evidence for indigenous transmission of vCJD in the United States and all 4 affected individuals have evidence supporting contraction of the disease outside of the United States (1,2).
- The most recent confirmed case underlies the need for continued surveillance and the continued need for physicians to identify possible cases of vCJD.
- Increasing evidence points to different strains of BSE: the typical BSE strain responsible for the outbreak in the United Kingdom and two atypical strains (H and L strains) for 3 of the US BSE cases (5).
- There is still a vital role of worldwide surveillance of vCJD since the incubation period of vCJD can last for decades and the presentation at onset can be non-specific.

References:

1. CDC website. [www.cdc.gov]
5. United States Department of Agriculture, USDA website. [www.aphis.usda.gov]