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LETTER TO THE EDITOR

Reply: Very early-onset frontotemporal dementia with no family history predicts underlying fused in sarcoma pathology

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Sir, cases previously described as atypical frontotemporal lobar degeneration with ubiquitinated inclusions (FTLD-U) were recently found to be characterized by FUS pathology (Neumann *et al.*, 2009). As described in this article and in the initial papers introducing the term atypical FTLD-U (Mackenzie *et al.*, 2008; Roeber *et al.*, 2008) as well as in more recent papers (Seelaar *et al.*, 2010; Urwin *et al.*, 2010), the clinical phenotype of these cases is characterized by a very early age at onset with absence of family history for dementias.

Loy *et al*. (2010) used the criterion of early age at onset (<40 years), in the absence of family history, to screen their 64 Brain Bank cases with FTLD and identified one case meeting this criterion and, indeed, this case showed FUS pathology.

This report consolidates that there is a characteristic clinical phenotype in FTLD-FUS (atypical FTLD-U) and that the already published clinical features are highly suggestive for FTLD-FUS (atypical FTLD-U).

However, to prove the usefulness (sensitivity/specificity) of these clinical features as predictors for FTLD-FUS (atypical FTLD-U) it would have been interesting to know whether FUS pathology was also present in any of the other 63 cases in their cohort.

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