Primary fibroblasts derived from sporadic amyotrophic lateral sclerosis patients do not show ALS cytological lesions

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Résumé en anglais	OBJECTIVE: Sporadic amyotrophic lateral sclerosis (sALS) is a fatal neurodegenerative disorder affecting upper and lower motor neurons. In view of the heterogeneous presentation of the disease, one of the current challenges is to identify diagnostic and prognostic markers in order to diagnose sALS at early stage and to stratify patients in trials. In this study, we sought to identify cytological hallmarks of sALS in patient-derived fibroblasts with the aim of finding new clinical-related markers of the disease. METHODS: Primary fibroblasts were prospectively collected from patients affected with classical, rapid, and slow forms of sALS. TDP-43 localization, cytoskeleton distribution, mitochondrial network architecture, and stress granules formation were analyzed using 3D fluorescence microscopy and new super-resolution imaging. Intracellular reactive oxygen species (ROS) production was assessed using live imaging techniques. RESULTS: Six sALS patients (two classical, two rapid, and two slow) and four age-matched controls were included. No difference in fibroblasts cell growth, morphology, and distribution was noticed. The analysis of TDP-43 did not reveal any mislocalization nor aggregation of the protein. The cytoskeleton was harmoniously distributed among the cells, without any inclusion noticed, and no difference was observed regarding the mitochondrial network architecture. Basal ROS production and response to induced stress were similar among patient and control fibroblasts. CONCLUSIONS: ALS cytological lesions are absent in patient-derived fibroblasts and thus cannot contribute as diagnostic nor prognostic markers of the disease.

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