ABSTRACT

Expansion of CTG trinucleotide repeats located in the 3' untranslated region (UTR) of myotonin protein kinase (*DMPK*) gene causes the most common adulthood neuromuscular disease, myotonic dystrophy 1 (DM1). Most classical DM1 patients did not start showing clinical phenotypes until young adulthood. To provide a model system for investigating the developmentally regulated pathogenic effects of DM1, we have examined the phenotypes of *Caenorhabditis elegans* expressing green fluorescent protein (*GFP*) gene with 0, 5, or 120 CTG and CAG repeats in the 3'-untranslated region driven by the *myo*3 promoter. Transgenic worms expressing GFP-CUG_{120} and GFP-CAG_{120} RNA, but not those expressing GFP-CUG_5 and GFP-CAG_5RNA, displayed an age-dependent impairment of GFP expression, locomotory behavior, moving rate, and muscle structure. In addition, RNA foci were found in the nuclei of the adult, but not L2, *myo*3::GFP-CTG_{120} animals, suggesting that the RNA foci formation may correlate with the decrease of GFP expression and the muscular dysfunction. These results first demonstrate that both the expanded CUG and CAG repeats could cause developmentally regulated defects on gene expression and muscle structure in *C. elegans*.