Mitochondria dynamically change their shape through frequent fusion and fission to continuously perform their function in the cell. Although a change in mitochondrial morphology was reported in amyotrophic lateral sclerosis (ALS), detailed changes of mitochondrial fusion and fission proteins have not been reported in ALS model mice. In transgenic (Tg) mice with the G93A human SOD1 mutation (G93ASOD1), both mitochondrial fusion proteins (Mfn1 and Opal) and fission proteins (Drp1 and Fis1) showed a significant increase in the anterior half of the lumbar spinal cord. Such changes in Tg mice were already noticeable at presymptomatic 10 week (W) compared with wildtype (WT) mice, detected through immunohistochemical as well as Western blot analyses. Furthermore, fusion protein levels of Mfn1 and Opal showed a progressive decrease from 10 to 18 W in Tg mice while fission protein levels of P-Drp1 and Fis1 maintained a high level of expression in Tg mice from 10 to 18 W. These data suggest that abnormal changes in mitochondrial morphology began before the onset of ALS and that the balanced mitochondrial morphology becomes altered by fissions in motor neurons (MNs) in this ALS model.