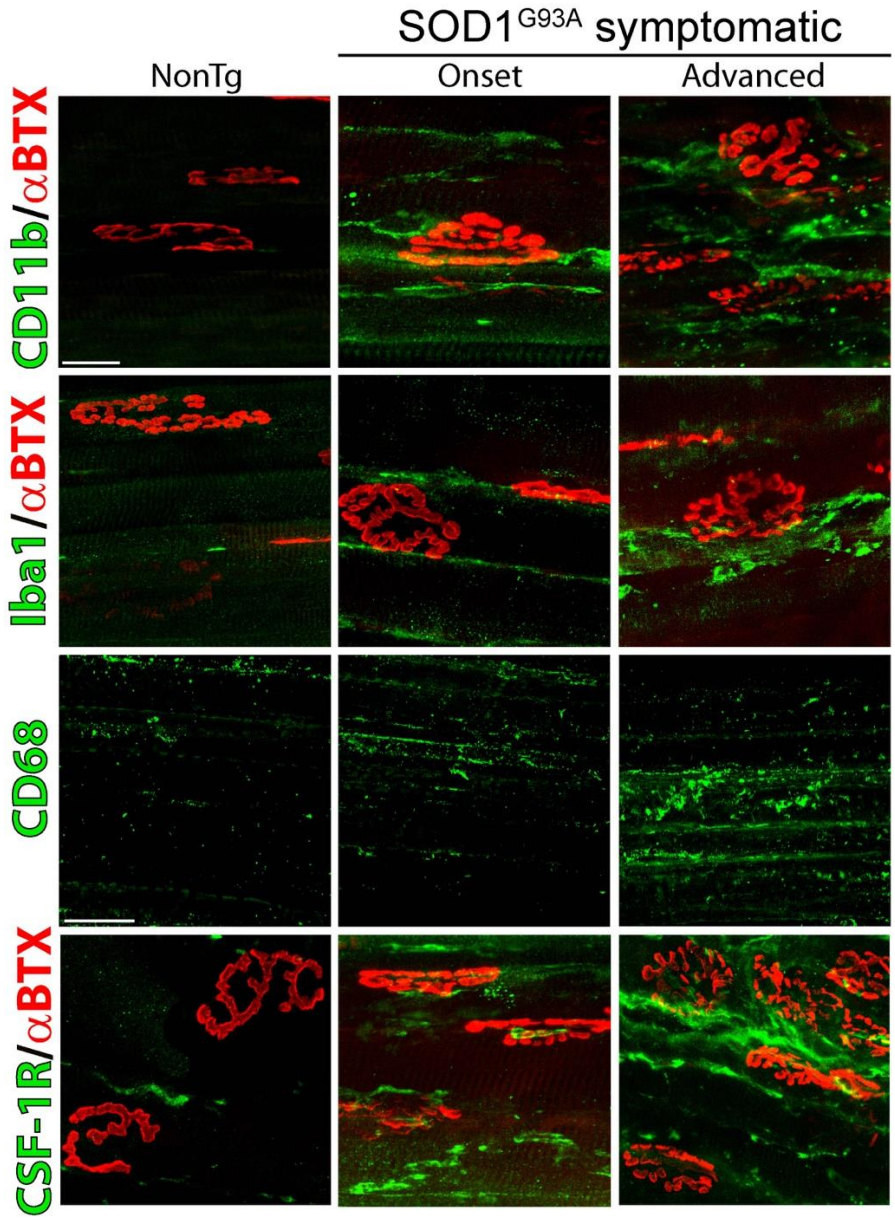
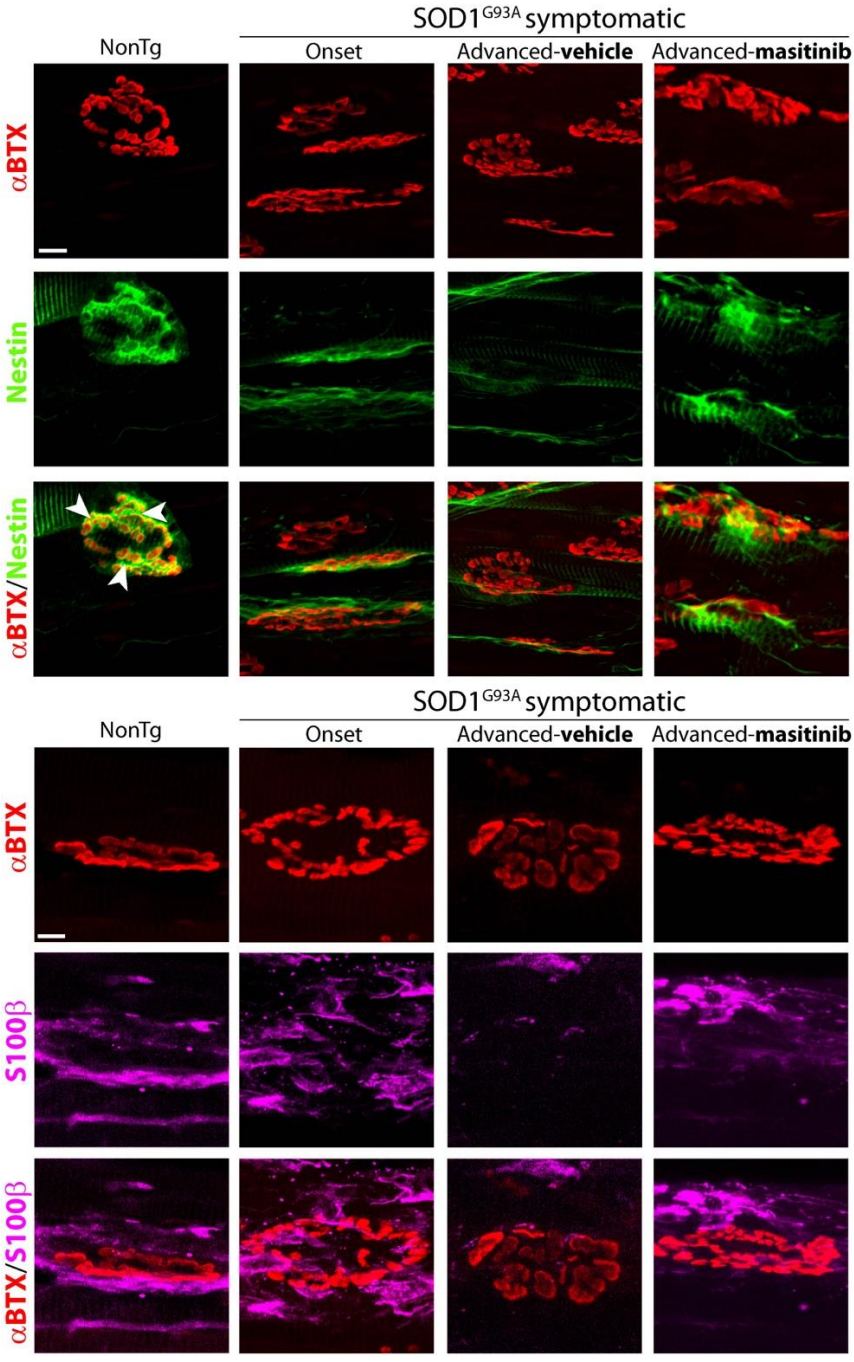
Supplemental Data

Supplemental Figure 1



Supplemental Figure 1. Interaction of macrophage-like cells and neuromuscular junctions (NMJs) in the *extensor digitorum longus* (EDL) muscle of SOD1^{G93A} rats. Representative confocal microscopic fields obtained from dissociated EDL muscle preparations from NonTg and SOD1^{G93A} rat (onset and advanced paralysis) EDL muscles. Motor endplates were labelled with α-bungarotoxin (red). Macrophages were labelled with Iba1 (green), CD68 (green) and CD11b (green), the latter also recognizing myeloid cells. Colony stimulating factor 1-receptor (CSF-1R) immunoreactivity (green) was also observed to interact with NMJs, largely co-localizing with macrophage markers. Note, the number of Iba1+/CD11b+ cells as well as CSF-1R immunoreactivity increase as the symptoms progress in advanced paralysis. n=4 animals/condition. Scale bars 20 μm.

Supplemental Figure 2



Supplemental Figure 2. Remodeling of perisynaptic Schwann cells during advanced paralysis is prevented my masitinib. Longitudinal cryostat sections of NonTg and SOD1^{G93A} rat (onset and advanced paralysis) *extensor digitorum longus* (EDL) muscles were processed for immunohistochemistry to visualize neuromuscular junctions (NMJs) and perisynaptic Schwann cells, as described in Fig 5. Confocal spatial interactions between NMJs (red) and perisynaptic S100β+ (magenta) and nestin+ (green) Schwann cells. Note Schwann cells surrounding and covering the endplates (yellow staining in nestin+ Schwann cells), and filling the spaces between adjacent gutters (arrowheads) of the NonTg rat muscle. This

interaction is lost in advanced paralysis and prevented by masitinib treatment. n=4 animals/condition. Scale bars 10 μm.