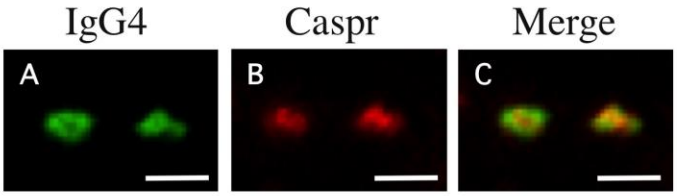
Supplementary data 2



Immunofluorescent studies. Longitudinal fresh-frozen sections from a patient with anti-neurofascin 155 antibodies (Patient 7) and a patient with anti-contactin 1 antibodies (Patient 10) were used to evaluate the deposition of autoantibodies and complements as described previously.[27,28] Sections were post-fixed in −20°C acetone for 10 min, blocked at room temperature for 1 hour and incubated overnight with primary antibodies at 4°C. We used sheep polyclonal antibody to IgG4 (The Binding Site Group Ltd., Birmingham, UK) and rabbit polyclonal antibody to Caspr (Abcam, Cambridge, UK) as primary antibodies to detect the deposition of autoantibodies. Rabbit monoclonal antibody to C3d (Abcam, Cambridge, UK) and sheep monoclonal antibody to IgG4 were used as primary antibodies to evaluate the deposition of complements. After washing, sections were incubated for 1 hour at room temperature in Alexa Fluor donkey anti-sheep IgG (H+L; 1:1000) or Alexa Fluor donkey anti-rabbit IgG (H+L; 1:1000). The stained sections were examined and photographed with a confocal laser scanning microscope (LSM5 Pascal, Carl Zeiss). We found co-localization of IgG4 and Caspr, suggesting the deposition of IgG4 at paranodes. Representative photographs taken from Patient 10 were shown in A to C. Scale bars = 2 μm. On the contrary, the deposition of C3d was not detected in either case.