

A



B

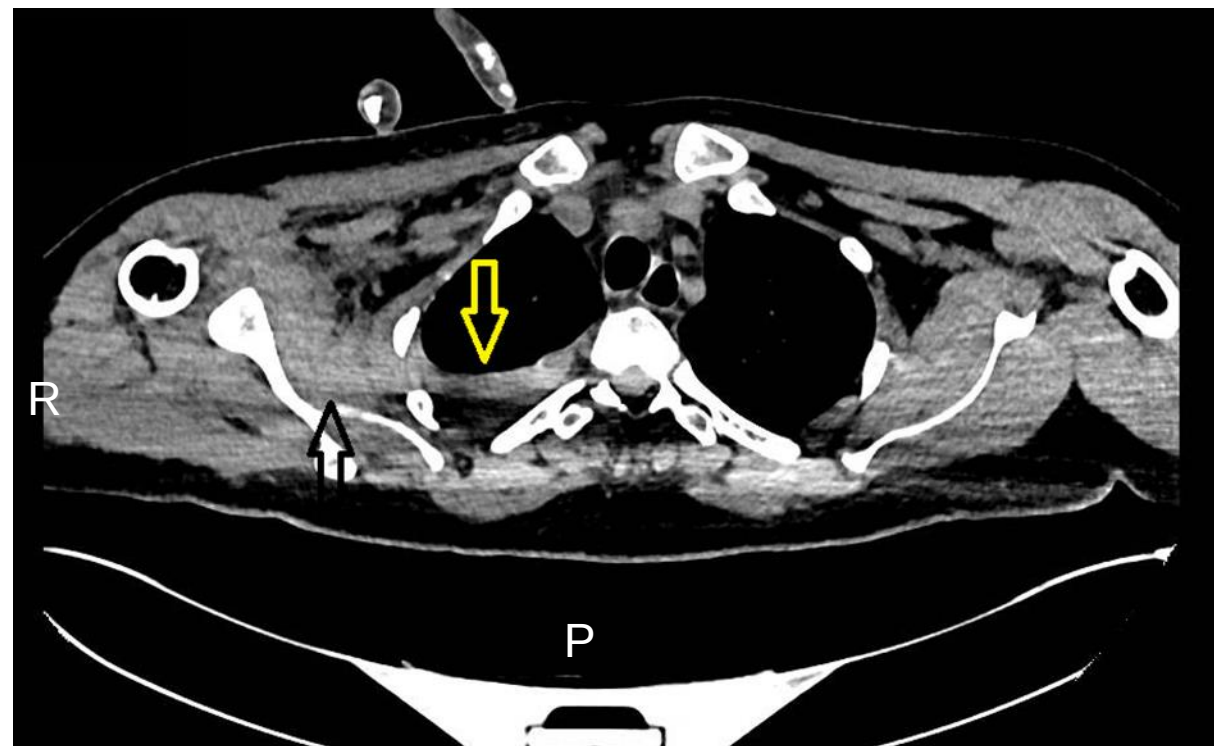
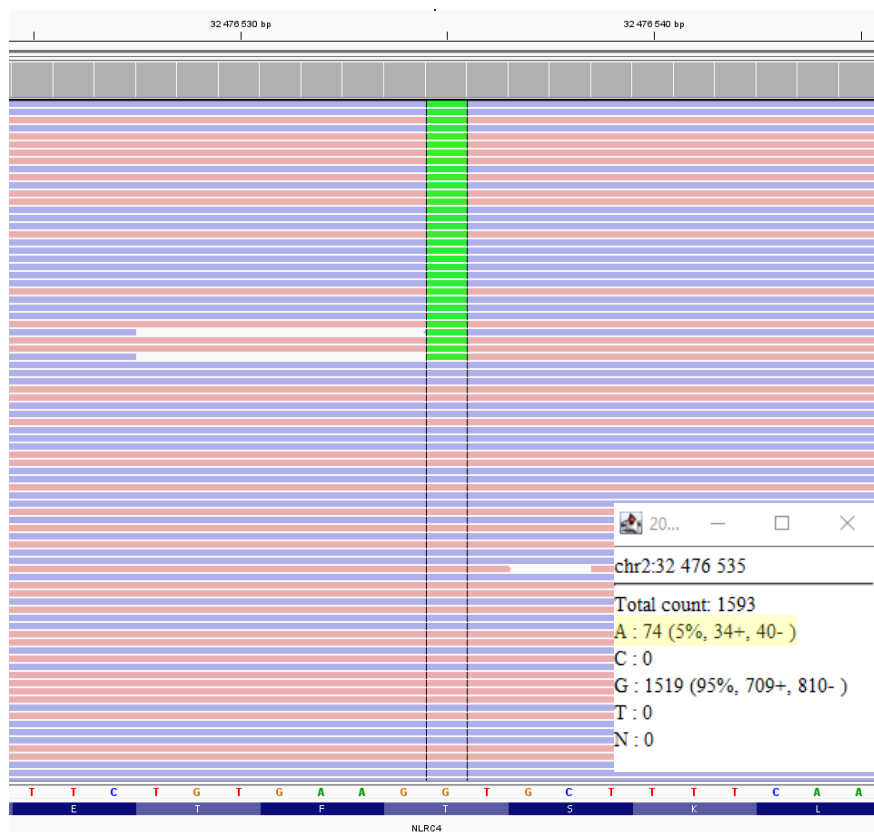
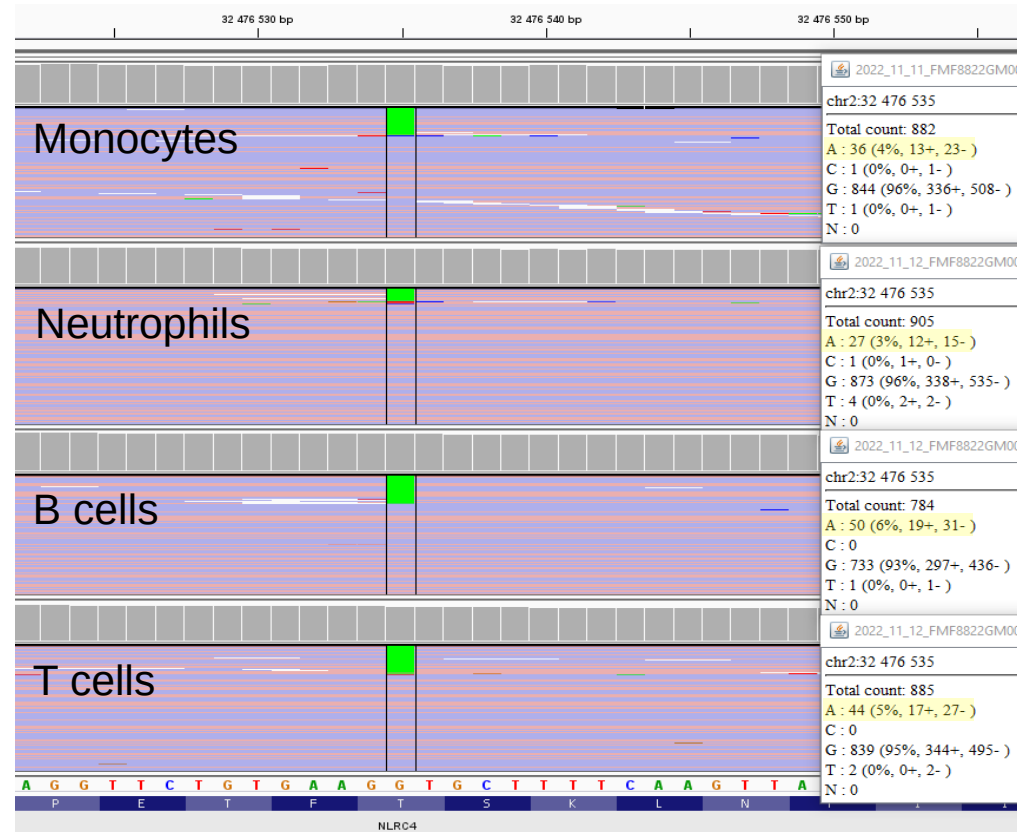


Figure S1. Clinical images reveal erythematous edema of the patient's left foot **(A)**. The cervico-cerebral CT scan revealed right cervical cutaneous cellulitis and a pleural effusion **(B)**.

A



B



C

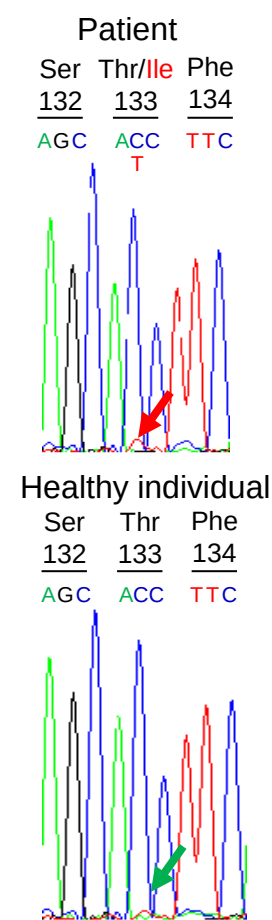


Figure S2. NGS sequencing reads covering the somatic mosaic p.Thr133Ile variant (NM_021209.4: c.398C>T) in the patient's whole blood DNA (A) and in the DNA from sorted patient's leukocytes (B). Sanger sequencing chromatograms showing the NLRC4 genotype at codon 133. The top panel displays the chromatogram from urine-derived DNA of the patient carrying the heterozygous c.398C>T (p.Thr133Ile) variant in a mosaic state. The bottom panel shows the chromatogram from a healthy individual at the same position.(C).

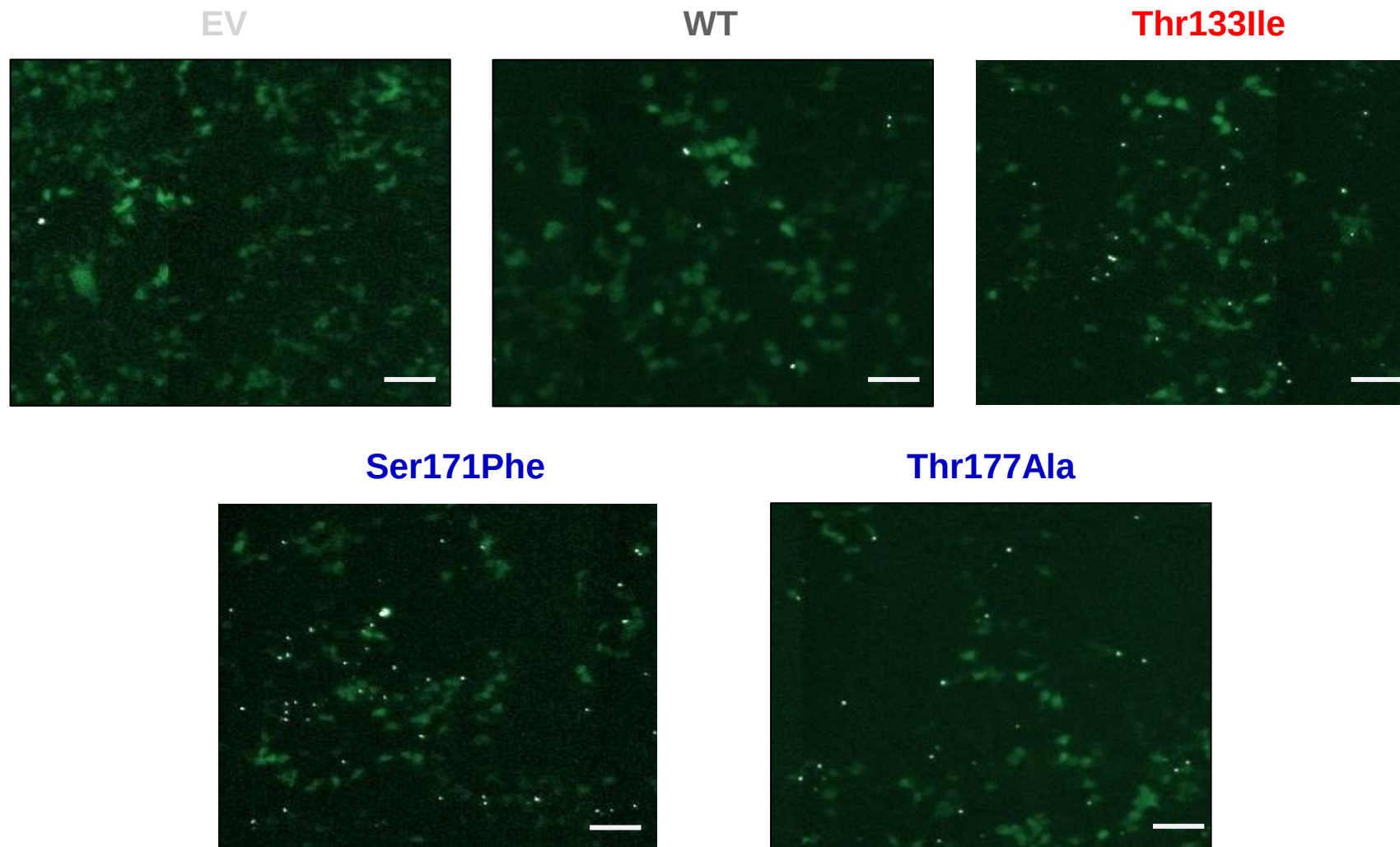


Figure S3. Representative images of HEK293T ASC GFP_C1-FLAG cells transiently transfected with pEV, pNLRC4_WT and each of the mutated plasmids. ASC specks are represented by white dots to differentiate them from the ASC GFP-expressing cells. Scale bar 40 μ m.