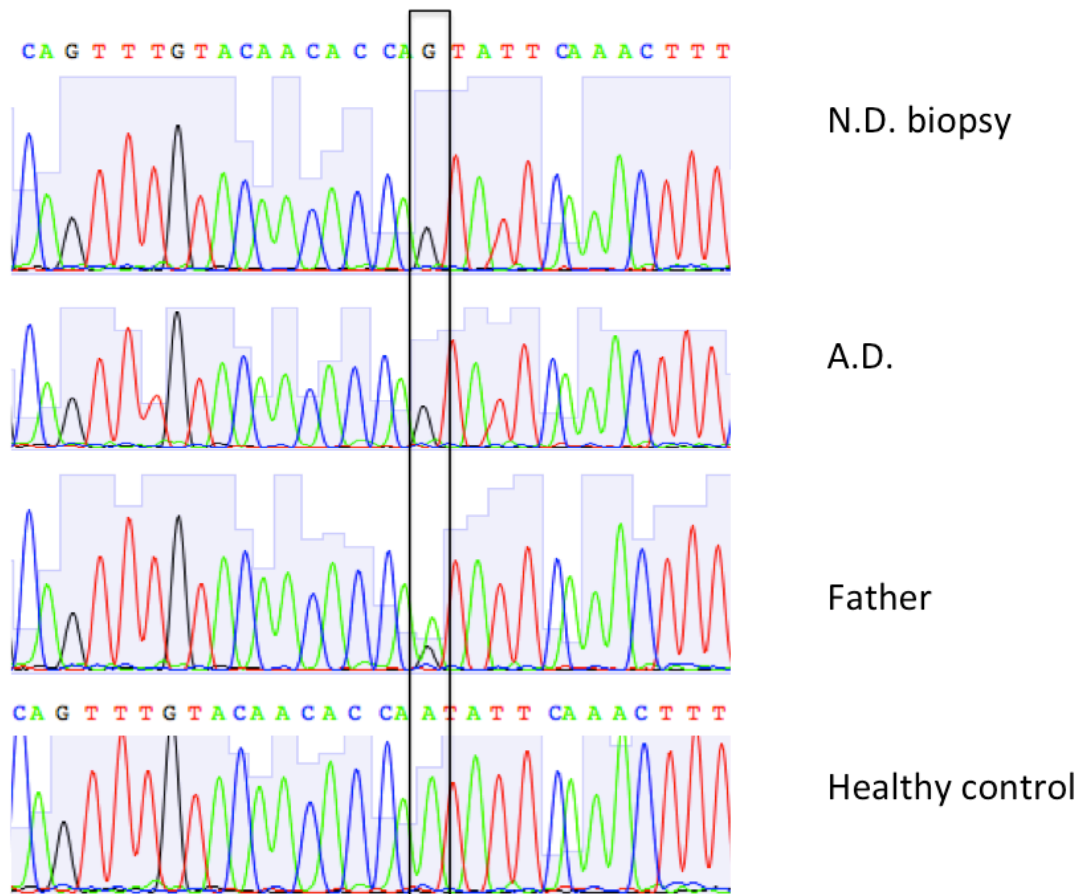


**Figure S1.**

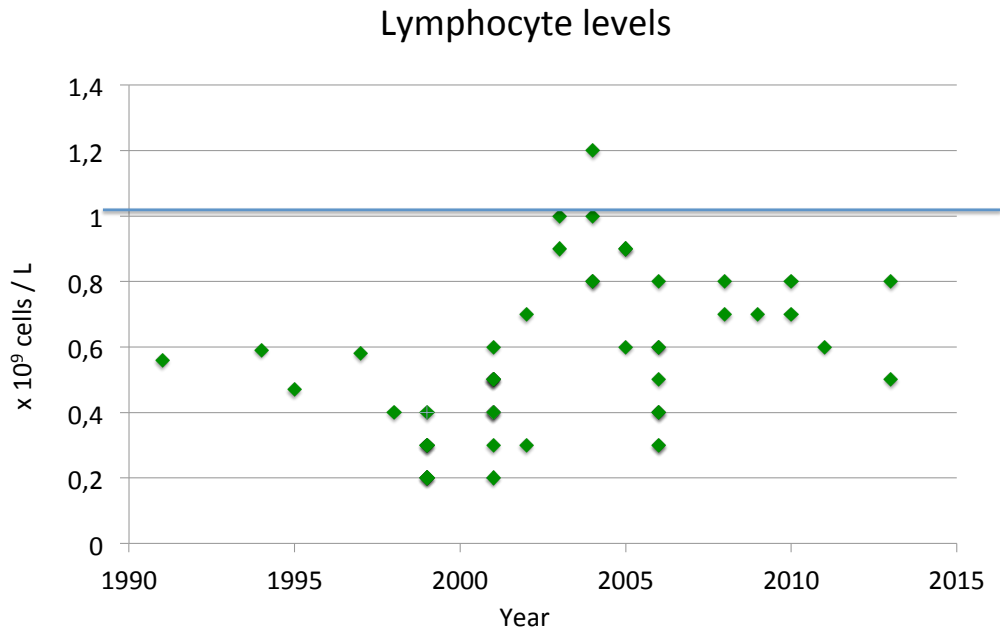


**Figure S1. Sequencing data including material from the biopsy**

DNA was extracted from a 40 years old skin biopsy from the deceased N.D. PCR-amplified and sequenced using the BigDye Terminator cycle sequencing kit. For optimal sequences this read was performed in reverse orientation. As controls DNA from Patient A.D., the father and a healthy control were included. The mutated base (A>G) is indicated by the square.

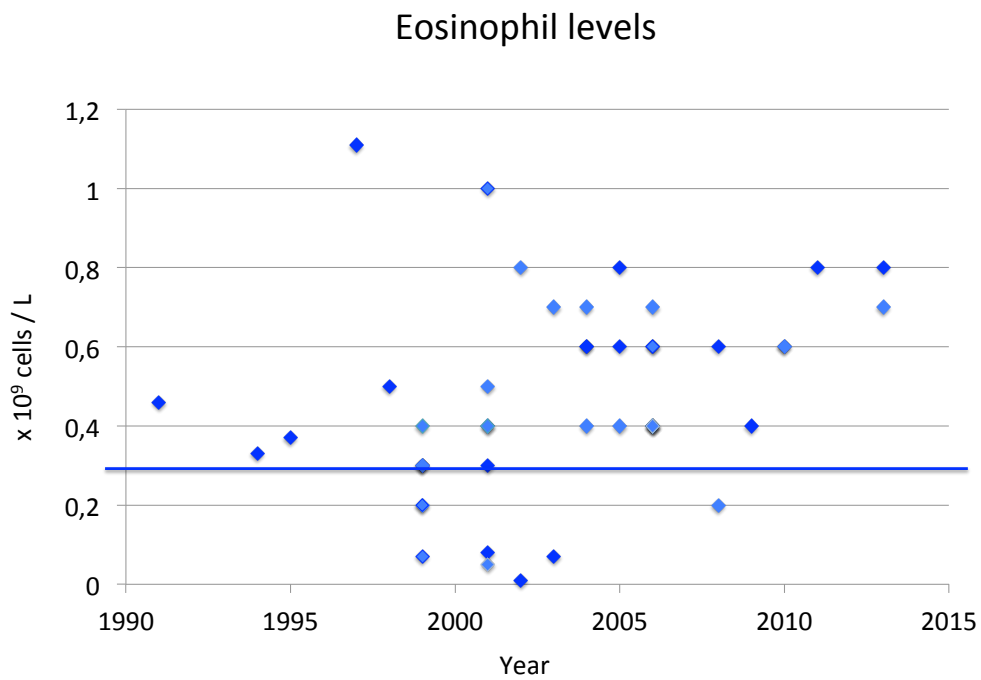
**Figure S2.**

**A**



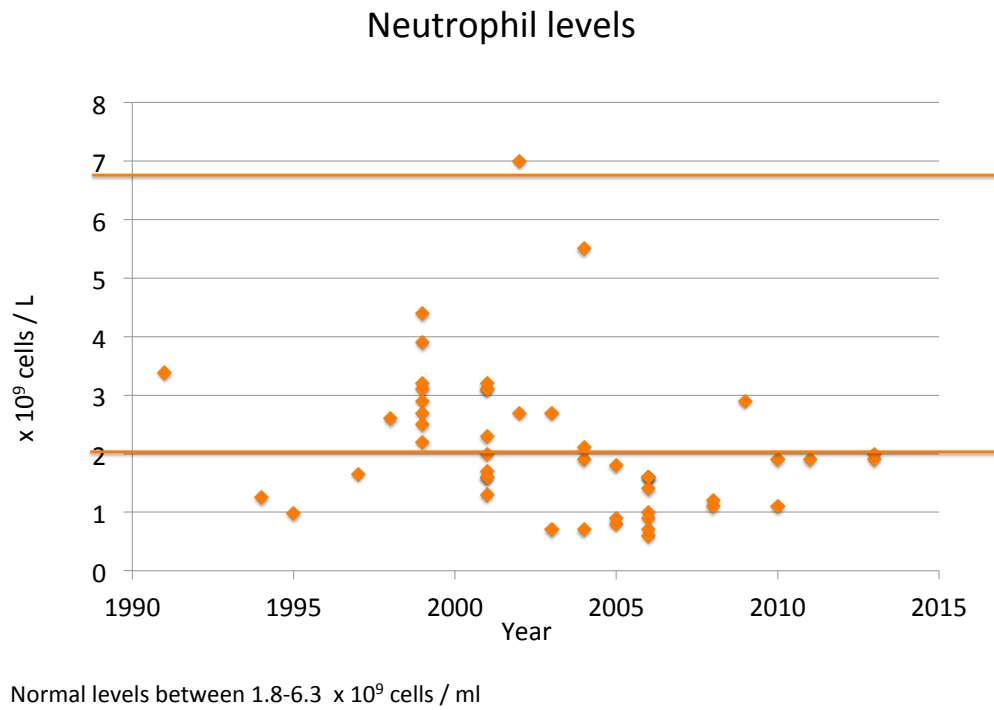
Normal levels between 1.0-3.5 x 10<sup>9</sup> cells / ml

**B**



Normal levels below 0.3 x 10<sup>9</sup> cells / ml

C



**Figure S2. Cell counts for patient A.D.**

Information regarding number of lymphocytes (panel A), eosinophils (panel B), and neutrophils (panel C) are shown on yearly basis since 1991. During infection the cell counts for eosinophils and neutrophils were temporarily normalized.