A novel ADOA-associated *OPA1* mutation alters the mitochondrial function, membrane potential, ROS production and apoptosis

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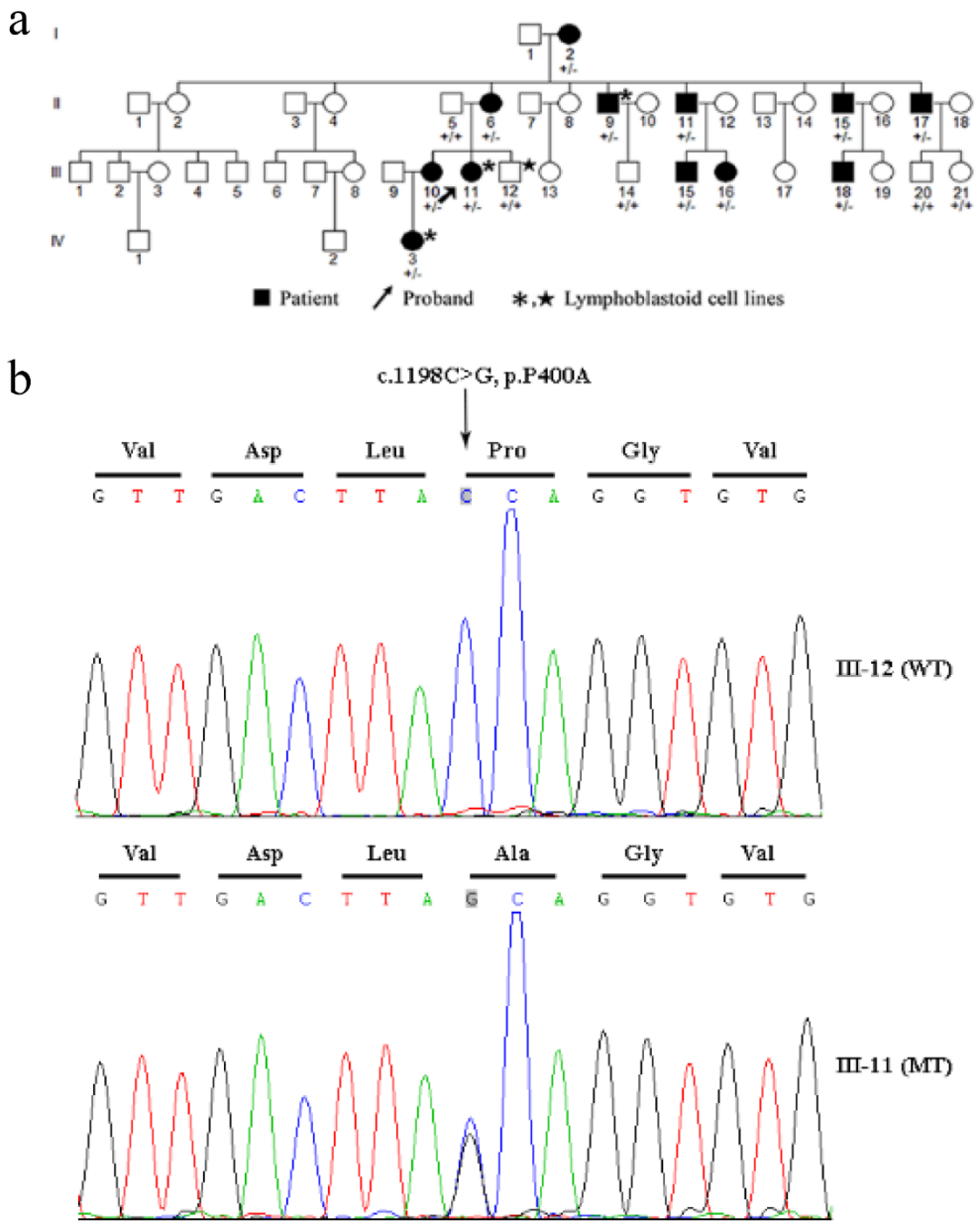
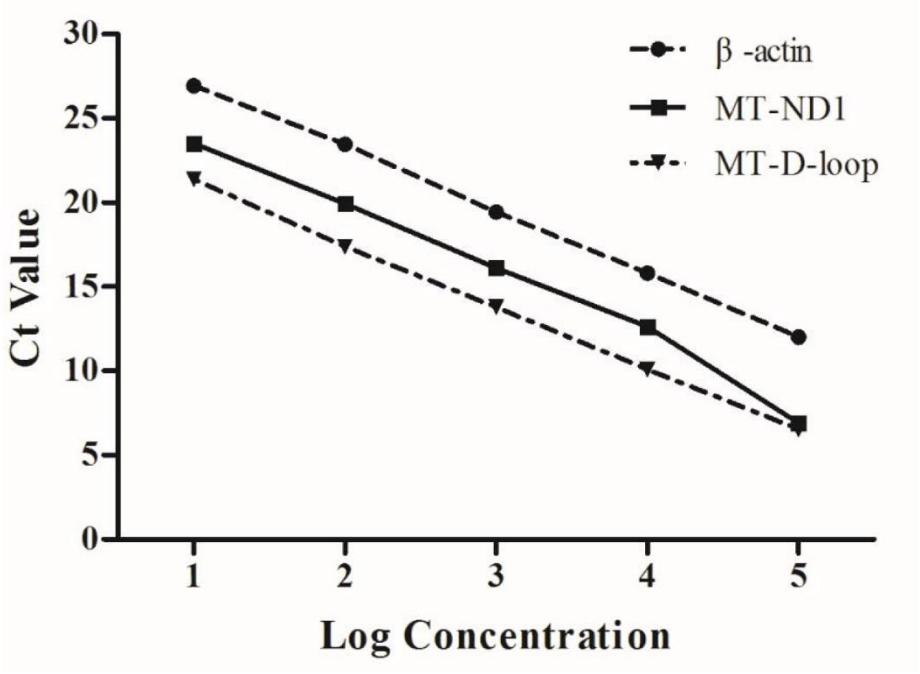
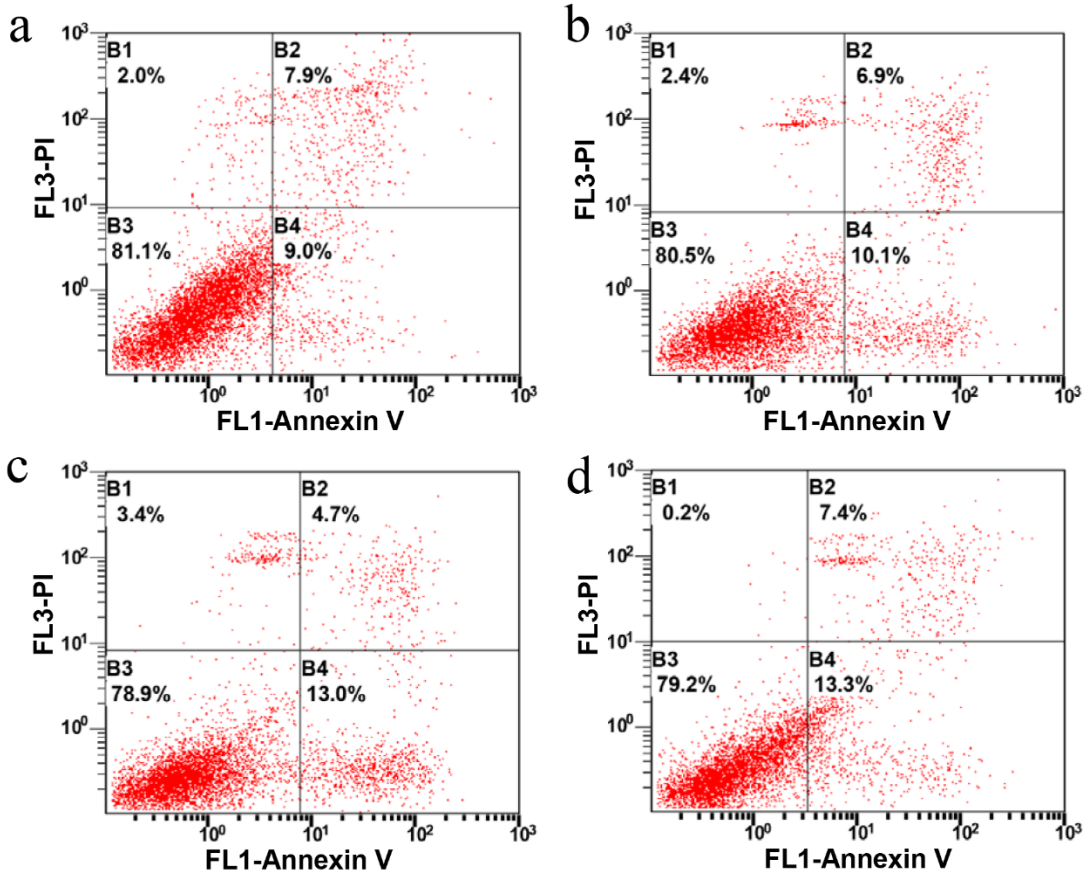


Supplementary Figure 1. Construction of lymphoblastoid cell lines and identification of cell lines. (a) Genotype analysis of the Chinese pedigree. Filled symbols: individuals with visual impairment, arrow indicated the proband; +/+: homozygote; +/-: heterozygote; *, Construction of lymphoblastoid cell lines. (b) Partial sequence chromatograms of the *OPA1* gene from lymphoblastoid cell lines of affected individual III-11 and the married-in control subject III-11. Arrow: location of the base changes at position 400.



Supplementary Figure 2. Standard curves of primers in QPCR. The standard curves were drawn with a ten-fold dilution series (10^0-10^4) of reference DNA.



Supplementary Figure 3. The ratio of apoptotic cells from the third to sixth generation. Flow cytometric analysis of the third to sixth generation cells stained with both PI and annexin V-FITC to demonstrate the ratio of later stage apoptosis and early necrosis.