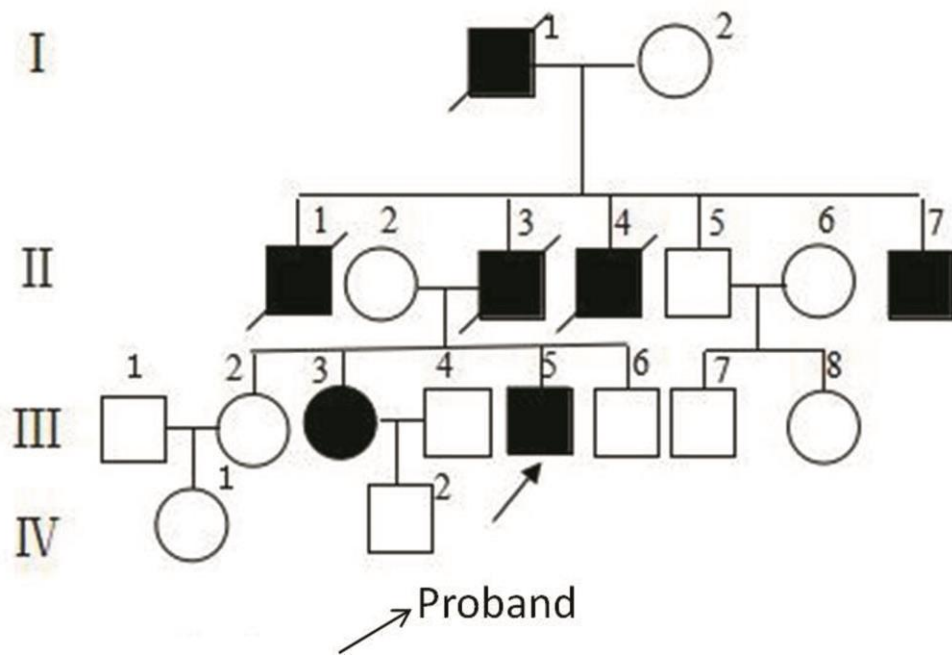


## Supplementary data:

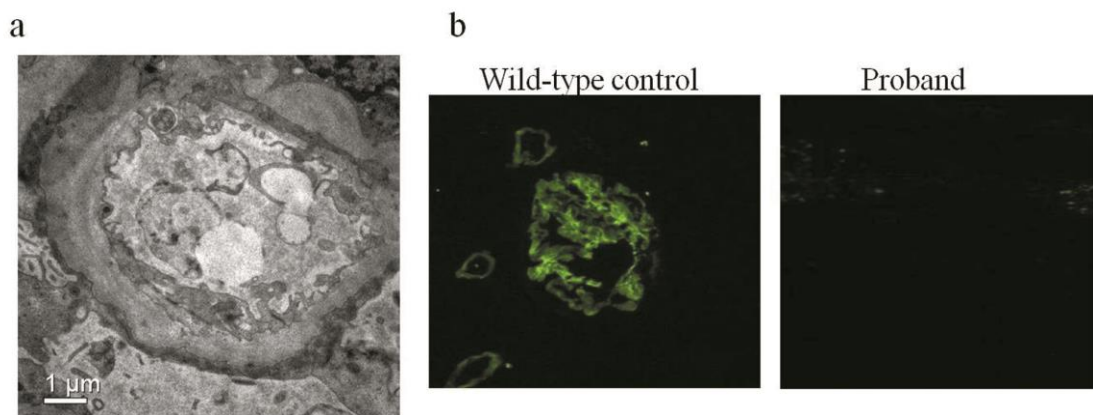
**Table 1.** Clinical data of this ADAS family.

Family members	Gender	age (yrs)	Creatinine	Urine	Urine
			clearance rate (mL/min)	protein	erythrocyte
I 2	Female	65	96	-	-
II 2	Female	47	102	-	-
II 5	Male	46	110	-	-
II 7	Male	45	76	2+	+
III 1	Male	27	118	-	-
III 2	Female	25	110	-	-
III 3	Female	26	73	+	2+
III 4	Male	24	112	-	-
III 5	Male	21	16	3+	2+
III 6	Male	23	115	-	-
IV 1	Female	2	110	-	-
IV 2	Male	5	108	-	-

Normal range of creatinine clearance rate: 80-120 mL/min for adult. -, Urine protein < 0.1 g / 24 h; +, urine protein 0.2-1.0 g / 24; 2+, urine protein 1.0-2.0 g / 24 h; 3+, urine protein 2.0-4.0 g / 24 h; +, urine erythrocyte>10; 2+, urine erythrocyte>20.



**Figure 1.** Pedigree structure of the studied family. Proband is denoted by an arrow. Squares represent males, circles represent females. Affected individuals are shaded.



**Figure 2.** Renal biopsy EM image and collagen IV immunofluorescence analysis of the proband. (a) Renal biopsy EM image from the proband showing thinning of the glomerular basement membrane. (b) Immunohistochemistry of collagen 3(IV) and 4(IV) in glomerular basement membrane. The expression of the collagen 3(IV) and 4(IV) exhibited normal in the Bowman's capsular basement membrane and tubular basement membrane of the wild-type control, and showed absent staining in the proband of the ADAS family.

a

gi 116256356 ref NP_000083.3	PGLPGA PGMRGPEGAMGLPGM	RGPPSGPGCKGEPGLDGRRGVDGV PGSPGP
gi 1034152188 ref XP_009442746	PGLPGA PGMRGPEGAMGLSGM	RGPPPGGCKGEPGLDGRRGVDGV PGSPGP
gi 795610428 ref XP_011918058.	PGLPGV PG L RGPEGAMGLPGM	RGPPPGGCKGEPGLDGRRGMDG I PGSPGP
gi 795599450 ref XP_011726579.	PGLPGV PG L RGPEGAMGLPGM	RGPPPGGCKGEPGLDGRRGMDG I PGSPGP
gi 594045436 ref XP_006047195.	PGLPGV PG P RGPEGTMGLPGM	RGPPPGGCKGEPGLDGRRGE DGLPGSPGP
gi 803310194 ref XP_012013465.	PGLPGV PG P RGPEGTMGLPGM	RGPLPGGCKGEPGLDGRRGE DGLPGSPGP
gi 32816561 gb AAP88582.1	PGLPGV PG P RGPEGAMGVPRM	RGPPPGGCKGEPGLEGRRE AG LPGPPGP
gi 1040227440 ref XP_017198661	PGLPGV PG P RGPEGAMGFPGQ	RGPPQGCKGEPGLDGKRGR DGVPGAPGP
gi 34328045 ref NP_031761.1	PGLPGV PG P RGPEGAMGEPGR	RGPLPGGCKGEPGPDGRRGQDG I PGSPGP
gi 209364566 ref NP_001129231.	PGAPGQPGV KGDGP L GPPG I	RGPCG P—R GQPGKDGKPGA PG PPGVKGS

\*\* \*\* \*\*; \* \* \*..\* \* \* \* \* : \*; \*\* .; \* .; \* \* \* \*

b

**Predictions**

Substitution at pos 237 from M to L is predicted to **AFFECT PROTEIN FUNCTION** with a score of 0.00.  
 Median sequence conservation: 3.05  
 Sequences represented at this position:137

**Figure 3.** Pathogenicity analysis of the exonic variant c.4195 A>T (p.Met1399Leu) *in silico*. (a) Conservation status of the exonic variant c.4195 A>T. Conservation status of this exonic variant was checked namely Clustal X software in ten species. (b) *in silico* analysis of the pathogenicity of substitution at position 1399 from M to L namely SIFT.