

Supplemental Figure 1: Proteomic Detection of Lysozyme in Congo Red Positive Intestine Biopsy Specimens.

(A) Congo red (CR) stained areas in the intestine biopsy specimen (same sample as in Figure 1E) were subjected to LMD-LC-MS/MS using a QExactive mass spectrometer (Thermo-Fisher, Waltham, MA). PreBlank refers to a blank injection to ensure clean analysis of the patient sample. Amyloidogenic lysozyme protein is highlighted with blue star. Proteins highlighted with yellow stars represent universal amyloid markers (PubMed ID:24720917). Numbers in green cells represent total MS/MS matched to the protein of interest in the respective sample. (B) Sequence coverage of lysozyme from sample#1 shows confident detection of the protein with multiple independent and unique peptides. (C) MS/MS spectra showing the peptide containing the mutation p.Leu102Ser. Arrow highlights the location of the mutation and the mutant amino acid. A total of 12 MS/MS spectra were detected for the mutant peptide.

(A)

#	Visible?	Starred?	Probability Legend:	Accession Number	Molecular Weight	Protein Grouping Ambiguity	PreBlank	Sample#1 (GI Biopsy)	Sample#2 (GI Biopsy)
Bio View: Identified Proteins (76/77) Including 6 Decoys									
1	<input checked="" type="checkbox"/>	★	over 95%	LYSC_HUM...	17 kDa			121	124
2	<input checked="" type="checkbox"/>	★	80% to 94%	APOA4_HU...	45 kDa			110	112
3	<input checked="" type="checkbox"/>	★	50% to 79%	SAMP_HUM...	25 kDa			38	42
4	<input checked="" type="checkbox"/>	★	20% to 49%	APOE_HUM...	36 kDa			15	21
5	<input checked="" type="checkbox"/>	★	0% to 19%	CO6A3_HU...	344 kDa			103	144
6	<input checked="" type="checkbox"/>	★		ACTC_HUM...	42 kDa	★		84	92
7	<input checked="" type="checkbox"/>	★		ALBU_HUM...	69 kDa			59	66
8	<input checked="" type="checkbox"/>	★		VTNC_HUM...	54 kDa			42	64
9	<input checked="" type="checkbox"/>	★		CO6A1_HU...	109 kDa			36	40
10	<input checked="" type="checkbox"/>	★		CO6A2_HU...	109 kDa			30	42

(B) LYSC_HUMAN (100%), 16,536.9 Da Sample#1 (GI Biopsy)

Lysozyme C

9 exclusive unique peptides, 11 exclusive unique spectra, 121 total spectra, 73/148 amino acids (49% coverage)

MKALIVLGLV LLSVTVQGVK FERCELARTL KR LGM DGYRG I SLANW MCLA
 KWESGYNTRA TNYNAGDRST DYGIFQINSR YWCNDGKTPG AVNACHL SCS
 ALLQDNIADA VACA KRVVRD PGGIRAWVAW RNRRCQNRDVR QYVQGC GV

