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Autoimmunity and cystatin SA 1 deficiency behind chronic mucocutaneous candidiasis in autoimmune polyendocrine syndrome

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Patients with the monogenic disease autoimmune polyendocrine syndrome type I (APS I) develop autoimmunity against multiple endocrine organs and suffer from chronic mucocutaneous candidiasis (CMC), a paradoxical complication with an unknown mechanism. We report that saliva from APS I patients with CMC was defective in inhibiting growth of *C. albicans in vitro* and had reduced levels of a salivary protein identified as cystatin SA1. In contrast, APS I patients with no CMC expressed salivary cystatin SA1 and could inhibit *C. albicans* to the same extent as healthy controls. We evaluated the anti-fungal activity of cystatin SA1 and found that synthesized full length cystatin SA1 efficiently inhibited growth of *C. albicans in vitro*. Moreover, APS I patients exhibited salivary IgA autoantibodies recognizing myosin-9, a protein expressed in the salivary glands that also produce cystatin SA1, thus linking autoimmunity to cystatin SA1 deficiency and CMC. This data suggests an autoimmune mechanism behind CMC in APS I and provide rationale for evaluating cystatin SA1 in anti-fungal therapy.

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