

WHAT IS ALECT2 AMYLOIDOSIS?

LECT-2 Amyloidosis, also known as ALECT2, is a rare form of systemic amyloidosis that most commonly affects the kidneys. It is caused by the misfolding of the leukocyte chemotactic factor-2 (LECT-2) protein. LECT-2 is a protein that is produced in the liver that has several functions in inflammation and metabolism. Initially thought to be limited to the kidneys, it has since been seen in the liver, and less commonly in the spleen, bowel, adrenal glands, lung, prostate, gallbladder, pancreas and parathyroid gland. The disease is slow to progress and often leads to end-stage kidney disease (ESKD).

WHO DOES IT AFFECT?

ALECT2 is most often seen in patients with Hispanic ethnicity, Native Americans, and those with Mexican, North-African and South-East Asian descent. It is mostly seen in patients over the age of 60, though there have been some cases diagnosed earlier.

WHAT CAUSES ALECT2?

It is still unclear what causes ALECT2, though we know the protein is associated with cell growth, immune function, and tissue repair following an injury. ALECT-2 predominantly presents as chronic kidney disease (CKD) and renal impairment. Hypertension and diabetes are common, though likely not the cause of the renal disease.

SIGNS AND SYMPTOMS

ALECT2 amyloidosis may be associated with unexplained weight loss, fatigue, and non-specific symptoms. Patients tend to present with signs or symptoms of impaired kidney function and are found to have protein in the urine (proteinuria) and/or a decline in the kidney's ability to filter blood (glomerular filtration rate). Patients may also have elevated serum creatinine levels.

DIAGNOSIS

It is incredibly important to rule out AL and AA, as treatment for these conditions is different and treatment delay can lead to worse outcomes. Kidney biopsy with Congo Red staining and Laser Dissection Mass Spectrometry to type the protein are necessary and the gold standard for confirming the diagnosis of ALECT2 amyloidosis.

PREVENTION AND TREATMENT

ALECT2 amyloidosis tends to progress slowly. Currently, there are no specific treatments for ALECT2 amyloidosis. Most management is supportive in nature, such as treating other conditions that can also affect kidney function (e.g., hypertension and diabetes). For ESKD, management may include dialysis and kidney transplantation.