



AL Amyloidosis

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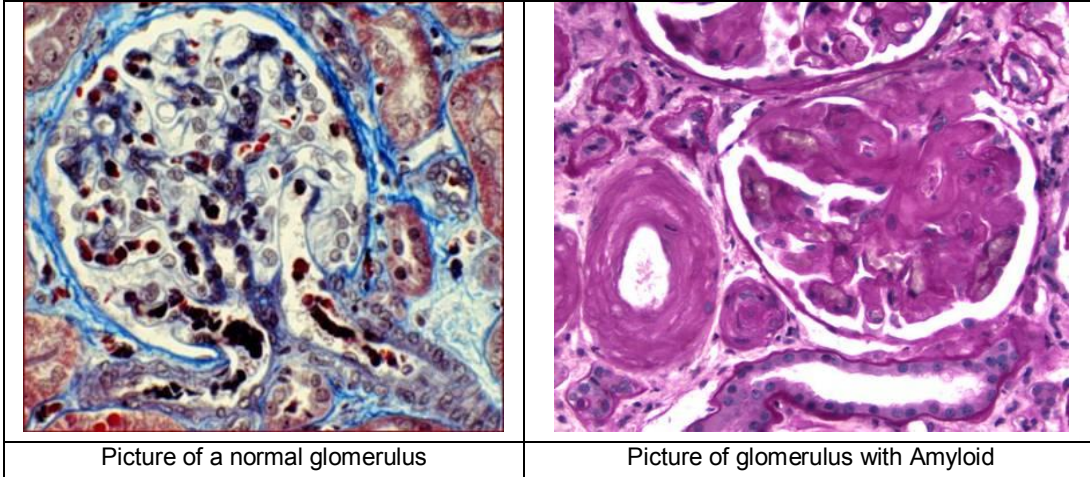
What is AL-Amyloidosis?

AL Amyloidosis (also called “primary” amyloidosis) is a blood disorder in which a certain protein accumulates in various tissues throughout the body. This protein, called “M-protein”, is actually composed of pieces from immunoglobulins (also called antibodies) which occur naturally in the body and fight off infection. In the case of people with amyloidosis, too many of these immunoglobulin fragments are present in the circulation. They can lodge in almost any organ in the body, such as the brain, the lungs, the liver, the heart, the skin, or the **kidneys**. As these proteins accumulate, they begin to affect the function of whichever organ they are in. Although AL amyloidosis can be suspected from many of its different symptoms (see below), the only way to be certain of the diagnosis is with a biopsy.

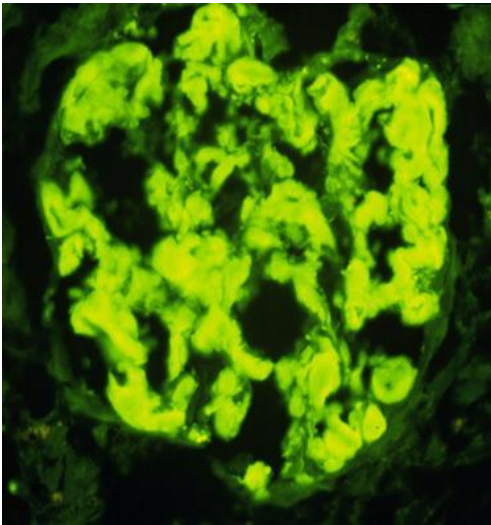
Renal, or kidney, amyloidosis occurs when the abnormal proteins mentioned above began to lodge in the kidneys. As the amyloid accumulates, the kidneys are no longer able to work properly. The kidneys normally filter and clean the blood, excreting the body’s natural waste products as urine. If they are unable to do this, these waste products build up in the blood. This is referred to as Renal Failure. Eventually renal failure can also cause problems with anemia, high blood pressure, and fluid build up.

What does it look like?

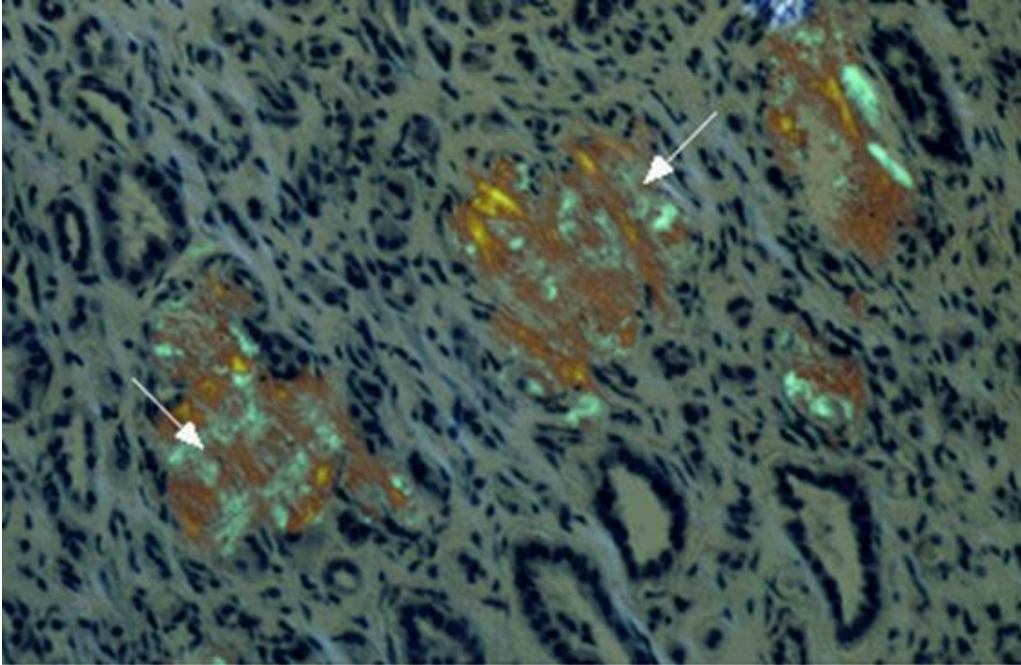
First a quick overview of the kidney- most people have two kidneys, one on each side of their lower back. All of the blood in your body passes through your kidneys many times during the day, and each time blood goes through some of it gets filtered by the gomeruli (singular = glomerulus). This filtering is how your body cleans the blood (and removes extra water), and some of the filtered blood becomes your urine. Urine isn’t red (like blood) because the red blood cells, which give blood it’s color, are too big to fit through the filters. A glomerulus is just a tiny bag of blood vessels through which blood gets filtered, and all of the filtered out blood (urine) runs into tubes (ureters) which eventually lead to your bladder.



Shown above are two glomeruli, or kidney filters, as they look under a microscope. The normal filter is on the left, and the filter affected by amyloidosis is on the right. Notice that in the normal glomerulus, there are many open “loops” scattered throughout- these are all small blood vessels seen in cross-section. In the biopsy with amyloidosis, almost all of these loops have collapsed because of all the deposited M-protein.



Shown above is another biopsy specimen. This time a “stain” has been added that turns the amyloid fluorescent green.



This is another stain that is used in diagnosis, called “Congo Red”. When exposed to amyloid, this stain turns “bright apple green”.

How did I get it?

We don't yet know why some people get AL amyloidosis, even though 1,000-3,000 new cases are diagnosed each year in the US. About 2/3 of these patients are male, and almost all of them are over the age of 40. Although this type of amyloidosis can occur by itself, it is often associated with other blood disorders, such as multiple myeloma and Waldenstrom's Macroglobulinemia. Both of these are diseases of the type of blood cells that produce immunoglobulins.

What are the symptoms?

Amyloidosis can affect many organs other than the kidney, and its symptoms can therefore be quite varied. Some of the most common are listed below, along with the organ system involved.

Kidney	Renal Failure, fluid retention, swelling, shortness of breath
Heart	Heart failure, arrhythmias, heart attack
Gastrointestinal	Liver/spleen swelling, GI bleeding, malabsorption

Neurologic	Pain and numbness
Musculoskeletal	Swollen (but weakened) muscles, swollen tongue, joint pain
Blood	Bleeding, anemia
Lung	Shortness of breath, fluid in the lung
Skin	Thickened skin, easy bruising

Amyloidosis can be suspected from any of the symptoms listed above. Both the blood and the urine can also be tested for the M-protein that causes amyloidosis. However, an actual diagnosis of AL Amyloidosis requires biopsy of an effected organ. Although this can be any effected organ, such as skin or bone marrow, Renal Amyloidosis can only be diagnosed by a kidney biopsy.

Treatment

Because AL amyloidosis is caused by the overproduction of immunoglobulins, which come from blood cells, it is best treated by a doctor specializing in blood and blood cancers (Hematologist/Oncologist). Treatment usually involves some sort of chemotherapy; some of the most common drugs used are- Steroids, Melphalan, and Thalidomide. Sometimes a bone marrow transplant is required as well.

Prognosis

Because primary amyloidosis can affect so many different organs, its course varies widely from person to person. The most life threatening complications are usually not associated with the kidney, but rather with heart involvement or infection. Patients most at risk have one or more of the following-

- High numbers of circulating plasma cells
- Significant bone marrow infiltration
- Cardiac involvement

Patients that respond to therapy usually have stabilization of their renal disease as well. When they do not, renal replacement therapy (dialysis) may become necessary.

