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Perspective

Brief Note on Amyloidosis

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DESCRIPTION

Amyloidosis is an uncommon disorder that occurs when an abnormal protein, called amyloid, develops in organs and inhibits the normal function. Organs that might be affected includes the heart, kidneys, liver, spleen, nervous system and digestive tract. Depending on the type of organ, the signs and symptoms varies. Amyloidosis includes swelling of ankles and legs, shortness of breath with minimal exertion, constipation, skin changes, such as thickening or easy bruising, and purplish patches around the eyes, difficulty in swallowing, irregular heartbeat, low red blood cell count, sudden weight loss, numbness, tingling or pain in hands or feet, especially pain in wrist, enlarged tongue, which sometimes looks rippled around its edge, severe fatigue and weakness.

Types of amyloidosis

Various proteins lead to amyloid deposits, but only a few have been related to major health problems. Different types of amyloidosis include:

Amyloid light chain amyloidosis (immunoglobulin light chain amyloidosis): It is the most common type and also called as primary amyloidosis. The cause is unknown but it occurs when bone marrow releases abnormal antibodies. It is associated with a blood cancer called multiple myeloma. It may affect kidneys, heart, liver, intestine, and nerves.

AA amyloidosis: AA stands for Amyloid Type-A. Earlier known as secondary amyloidosis, this condition is the consequence of another chronic infectious or inflammatory disease such as rheumatoid arthritis, crohn's disease, or ulcerative colitis. It generally affects kidneys, but it may also affect digestive tract, liver, and heart. Amyloid type-A protein is responsible for this.

Dialysis-related amyloidosis: This is most common in adults and people who have been on dialysis for more than 5 years. This form of amyloidosis is affected by deposits of beta-2 micro-globulin that develops in the blood. Deposits can build up in different tissues, but it commonly affects bones, joints, and tendons.

Age-related systemic amyloidosis: This is affected by deposits of normal Transthyretin in the heart and other tissues. It is more common in adults especially men.

Organ-specific amyloidosis: It is affected by deposits of amyloid protein in single organ, including the skin (cutaneous amyloidosis).

Hereditary amyloidosis: This is an genetic disorder. It affects the liver, nerves, heart and kidneys. Various genetic defects are related to a higher chance of amyloid disease.

Men are more likely affected by amyloidosis when compared to women. It affects 15% of patients with multiple myeloma. It may occur in people with kidney disorder who are on dialysis for a longer period of time. Factors that increase risk of amyloidosis include age, family history, sex, kidney dialysis and other diseases like chronic infectious or inflammatory disease may increase risk of AA amyloidosis. Amyloidosis can damage heart, nervous system and kidneys.

Amyloid decreases heart capability to fill with blood during cardiac cycles. When less amount of blood is pumped during each cardiac cycle, it leads to shortness of breath. Amyloid-related heart complications can become life-threatening.

Pain, numbness or tingling of fingers, burning sensation in toes or the soles of feet can be observed in amyloidosis. If amyloid disturbs the nervous system in intestine, it may lead to constipation or diarrhoea. Amyloid can damage the kidneys filtering system, causing protein leaking from blood to urine. The capability of kidneys to remove waste products from body is reduced, which finally leads to kidney failure or dialysis.

Diagnosis of amyloidosis can be performed by various tests such as urine test, blood test, biopsy, genetic tests, etc. Urine test and blood test may be followed by one or more imaging processes to take an appearance at body's internal organs, such as an echocardiogram, nuclear heart test or liver ultrasound. A genetic test can be required to check if amyloidosis is of hereditary type. Biopsy is performed by taking a sample of bone marrow.

The main goal of amyloidosis treatment is to reduce the impact of symptoms and prolong life of the patient. It has a poor prognosis and the average existence without treatment is only 13

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months. Cardiac participation has the worst prognosis and consequences in death in about 6 months after onset of congestive heart failure. Only 5% of the patients with primary amyloidosis survive beyond 10 years.

CONCLUSION

Amyloidosis is a multifaceted disease with several organs affects and dissimilar manifestations. The clinicians required to have a high degree of suspicion to make an early and correct diagnosis and initiate the suitable treatment. Treatment of these convoluted patients requires cautious coordination among a wide assortment of subspecialists. This has become considerably more significant as compelling mediations have arisen for those with AL and different sorts of amyloidosis, and the progress of remedial intercession is predicated on the earliest conceivable analysis.