Understanding Amyloidosis: Unraveling the Complexities and Therapeutic Approaches for Oncology Nurses

Teresa Fogaren, MSN, AGPCNP-C, Anthony C. Shelton, RN, OCN®, Douglas V. Faller, MD, PhD, Dawn Marie Stull, PharmD, BCOP, Sara Thuenemann, RN, MSN, and Lisa Mendelson, MSN, AGPCNP-BC

BACKGROUND: Primary systemic light-chain (AL) amyloidosis is a rare clonal plasma cell disorder characterized by the production of abnormal immunoglobulin fragments, which form insoluble fibrils that aggregate as amyloid deposits in organs and tissues, leading to organ dysfunction and death.

OBJECTIVES: The aim of this literature review is to increase awareness of AL amyloidosis and educate nurses on the care of this patient population.

METHODS: This overview is based on a literature search of AL amyloidosis, including its pathogenesis, prognosis, and presentation. Guidance for nursing assessment, intervention, and patient education throughout the disease trajectory is presented.

FINDINGS: AL amyloidosis is a rare disease resulting in organ impairment and death if untreated. Nursing management includes knowledge of key assessment, monitoring, intervention, and education strategies with goals to preserve organ function and improve survival and quality of life in patients with AL amyloidosis.

KEYWORDS
AL amyloidosis; oncology nursing; treatment; multiple myeloma; MGUS

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AMYLOIDOSIS IS A BROAD TERM THAT ENCOMPASSES multiple, rare protein-misfolding disorders characterized by the extracellular deposition of insoluble amyloid proteins in organs and tissues (Merlini, 2017; Merlini & Bellotti, 2003). There are at least 36 different types of amyloidosis (Sipe et al., 2016), which may present as localized or systemic disease (Merlini, 2017; Merlini & Bellotti, 2003). The most common presentation of systemic amyloidosis is immunoglobulin light-chain (AL) amyloidosis. AL amyloidosis is a serious, incurable, but treatable disease, accounting for roughly 70% of amyloidosis cases (Milani et al., 2018; Palladini & Merlini, 2016).

Light chains are normally part of immunoglobulins (or antibodies) and are produced by a type of mature white blood cell, or B-lymphocyte, called a plasma cell, which originates in the bone marrow. Similar to multiple myeloma (MM), AL amyloidosis is a disease that arises from the overproduction of abnormal light chains and their fragments by a small subset of genetically identical (clonal) plasma cells (Milani et al., 2018; Myeloma UK, 2018). In AL amyloidosis, the abnormal light chains and their fragments have a nonconventional structure that causes them to misfold and interact with other proteins to form insoluble thread-like strings called fibrils. These fibrils then aggregate in various organs and tissues throughout the body as protein deposits known as amyloid (Zhang et al., 2017). Amyloid deposition results in progressive disruption of normal tissue structure, and can ultimately lead to organ failure and death (Bianchi & Kumar, 2020; Milani et al., 2018). Multiple organs are affected in AL amyloidosis, but dysfunction in one or two organ systems normally predominates. The kidney and the heart are the most commonly affected organs, with dysfunction in each of these organs observed in roughly 70% of cases. In addition, the liver, nervous system, gastrointestinal tract, lungs, and soft tissue may also be affected (Grogan et al., 2017; Merlini et al., 2016).

In 2012, the annual global incidence of AL amyloidosis was reported to be about nine cases per one million people (Desport et al., 2012), although the true figure may be higher because the condition is frequently unsuspected (Lousada et al., 2015; McCausland et al., 2017). According to the Amyloidosis Foundation (2020), about 4,500 new cases of AL amyloidosis are diagnosed each year in the United States, and it has been estimated that nearly 12,000