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Case report

Lymph node enlargement due to amyloid

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Summary. A case of a patient presenting with supraclavicular and cervical lymph node enlargement with demonstration of amyloid is presented. Histologic features are described. Amyloid deposition was not found in any other organ. Immunohistochemical typing with antisera raised against protein AA, Aλ, and Aκ showed a reaction only against Aλ. Thus, this case belongs to the immunoglobulin λ light chain-derived types that occur in many other parts of the body, particularly the respiratory tract.

Key words: Amyloid – Amyloidosis – Lymph nodes.

Report of a case

This 53 year old white male sought medical advice because of swellings noted for the previous 2 weeks in the right neck and supraclavicular area. Other signs or symptoms were denied. Enlarged lymph nodes were found on the right side lateral to the internal jugular vein and superior to the subclavian vein. No other lymph node enlargement and an otherwise entirely normal physical examination was found. Liver and spleen were not enlarged. A chest film was interpreted as normal. The hemoglobin, hematocrit, platelet and leukocyte counts, and erythrocyte sedimentation rate were all normal. A bone marrow examination was normal, and there was no increase in plasma cells. A serum protein electrophoresis was normal. No amyloid was found in a rectal biopsy.

On gross examination, the specimen consisted of seven oval tan segments of tissue, the largest measuring $6.0 \times 5.0 \times 3.0$ cm and the smallest 1.5×1 cm. On cut section, they had a waxy, lardaceous appearance. The overall configurations were those of lymph nodes.

Histologically, the lymphoid tissue was entirely replaced by an eosinophilic, amorphous, "soft"-appearing substance, deposited in irregularly-shaped masses of varying size (Figs. 1 and 2). Between these masses were small numbers of lymphocytes, and an occasional plasma cell, macrophages, and small vascular channels. There were no sheets of plasma cells and no atypical plasma cells. Perivascular deposition of amyloid was present. Occasional multinucleated macrophage giant cells were present. Congo red staining was positive and a birefringent emerald green with polarized light.

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Fig. 1. The thickened capsule of the lymph node is apparent. The amyloid masses are irregular in size and shape. H & E. ×135

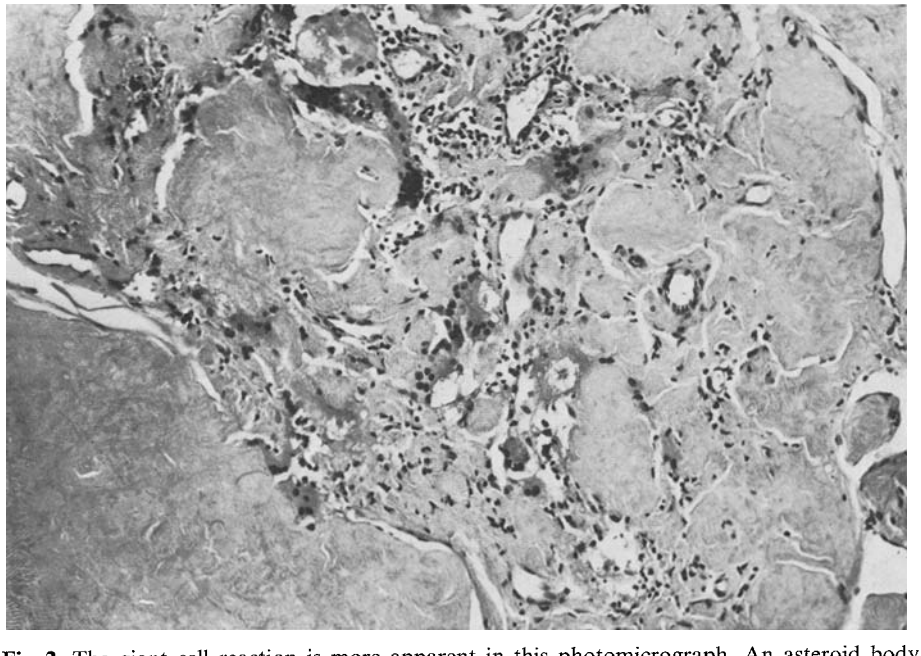

Fig. 2. The giant cell reaction is more apparent in this photomicrograph. An asteroid body in a giant cell is visible just to the right of center. H & E. ×350

Immunohistochemical typing was performed on formalin-fixed paraffin-embedded tissue sections using rabbit antisera against purified amyloid fibril proteins, e.g., Anti-AA, Anti-Aλ, and Anti-Aκ as described elsewhere (Linke and Nathrath 1980). A reaction was found only with the Anti-Aλ antiserum.

Discussion

Mackenzie (1963) described three patients who presented with lymph node enlargement as their chief complaint where the lymph nodes were found to contain amyloid. Two of the patients died of amyloidosis of unknown cause. In our experience, lymph node enlargement as the presenting feature of amyloidosis is rare. A report on all the cases of amyloid involving lymph nodes collected at the Lymph Node Registry in Kiel will be given in a separate article (Newland and Lennert, in preparation). Among a total of 40,000 lymph node biopsies in the files of the Registry, there were only 10 cases of amyloidosis in which lymph node enlargement was the presenting feature.

The microscopic appearance of amyloidosis involving lymph nodes is said to be variable (Symmers 1956; Robb-Smith and Taylor 1981). It may involve vessels in and around the node only, to replacement of the entire node by amyloid as in the presently described case. The giant cell reaction as seen in this case may sometimes be quite marked (Henry and Farrer-Brown 1981).

It has been stated that usually amyloid firstly appears in the cortical and medullary reticulin framework of lymph nodes (Symmers 1956). Follicles may or not be involved at this point. Eventually, the lymph node may become a mass of amyloid with little remaining recognizable architecture. Occasionally, the follicles may be surrounded by amyloid. Less commonly, the amyloid is said to initially be deposited in the walls of the sinuses and extend from there.

It is now known that amyloidosis is due to a variety of diseases and disease processes with different mechanisms for the deposition of amyloid (Glenner 1980). Review articles from several points in time indicate the progress made in the understanding of amyloidosis (Symmers 1956; Glenner et al. 1973; Glenner 1980). The ultimate outcome for the patient described here at this point in time is uncertain. Could there be a solitary lymph node amyloid deposition independent of generalized amyloidosis as in amyloid tumor of the lung? In this case, there was no accumulation in the other areas examined. The negative clinical history, physical examination, a normal peripheral blood and bone, a normal serum protein electrophoresis (performed in a routine laboratory), and the absence of amyloid in a rectal biopsy suggest that this patient has neither primary generalized amyloidosis nor amyloidosis secondary to some other disease. The identification of Aλ-amyloid suggests that the amyloid originated from a (benign or malignant) plasma cell proliferation. Since such a proliferation was not found, it is likely that the amyloid tumor described here arose locally as has been found in the lung (Glenner 1980) and trachea (Linke and Nathrath 1980).

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Note added in proof.

The patient is in good health 15 months after removal of the lymph node. No indications of a plasma cell neoplasm have been found.