

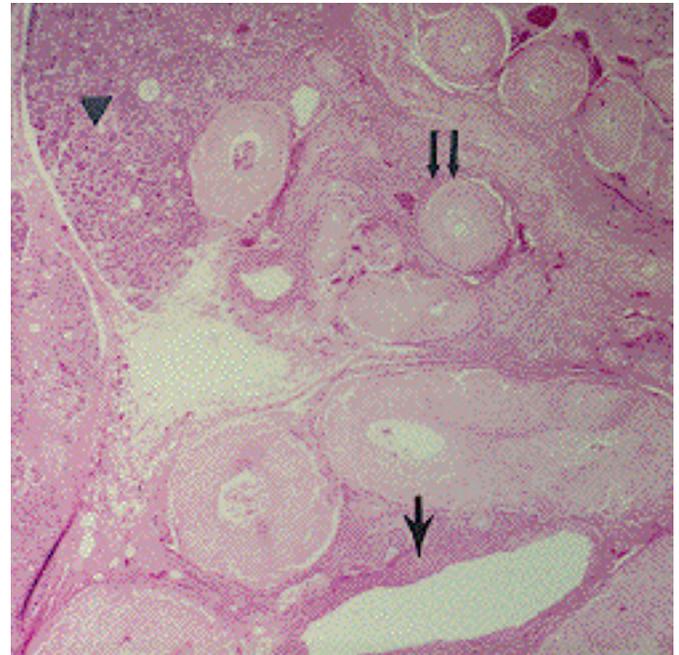
AMYLOIDOSIS

Amyloidosis is the name given to a deposition of extracellular proteinase material found with many different diseases. Not all amyloids are the same, but they show common morphologic properties. Grossly, the lesions are mucosal covered when they appear in the laryngeal or lingual areas or are covered by nonulcerated skin when they are cutaneous.

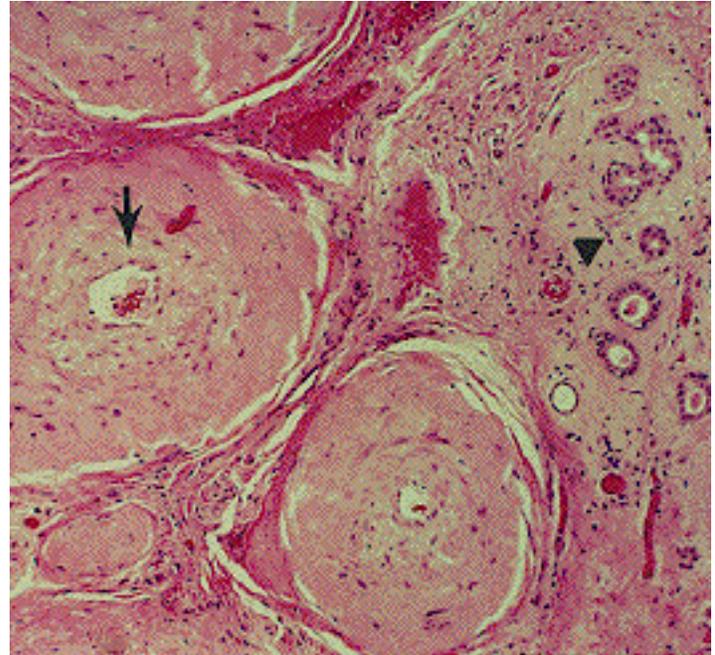
Systemic amyloidosis may be of a primary type or appear as a myeloma—associated disease, especially in older adults. So-called secondary amyloidosis characteristically results from long standing wasting diseases such as osteomyelitis and tuberculosis and, therefore, was more common formerly than now. There also is a hereditofamilial form and a hemodialysis-associated type.

Microscopically, there is eosinophilic acellular amorphous material often arranged perivascularly or spread throughout the specimen. Foreign body giant cells are occasionally present and they contain amyloid. Using Congo red stain, amyloid stains red and becomes more apparent. When the same stain is viewed under polarized light, there is a yellow-green birefringence.

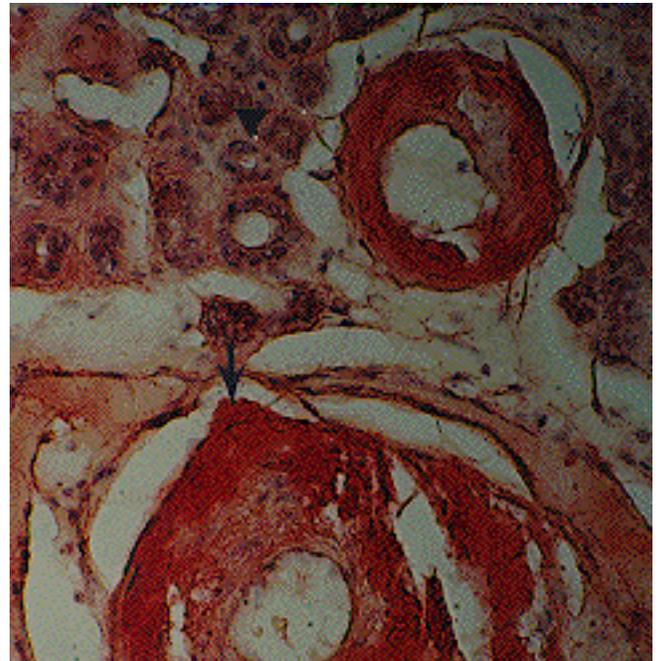
Amyloidosis, submandibular gland. A deposit of amyloid around ductal walls (double arrows) and in the walls of blood vessels is seen (single arrow). There is an eosinophilic appearance to the amyloid and very few cells are present. Much of the submandibular gland (triangle) has been replaced by amyloid deposits.

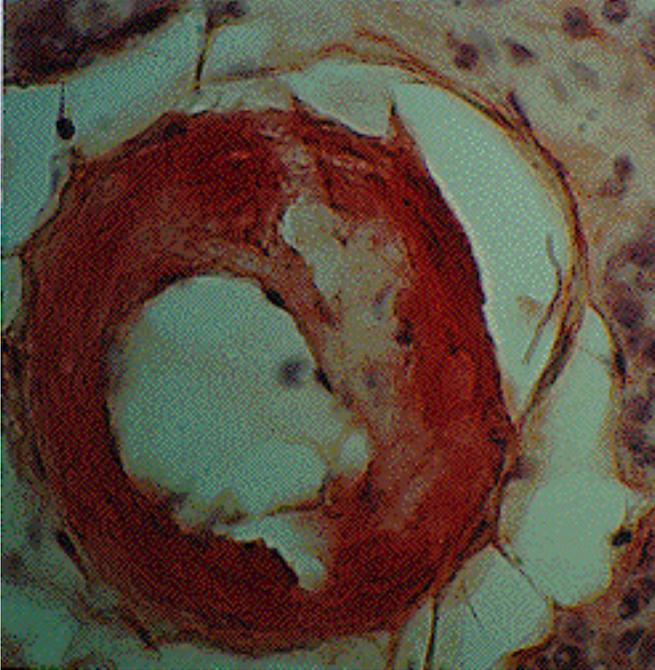


Amyloidosis, higher power view of upper left. Amyloid surrounding blood vessel (arrow) and atrophic gland (triangle).

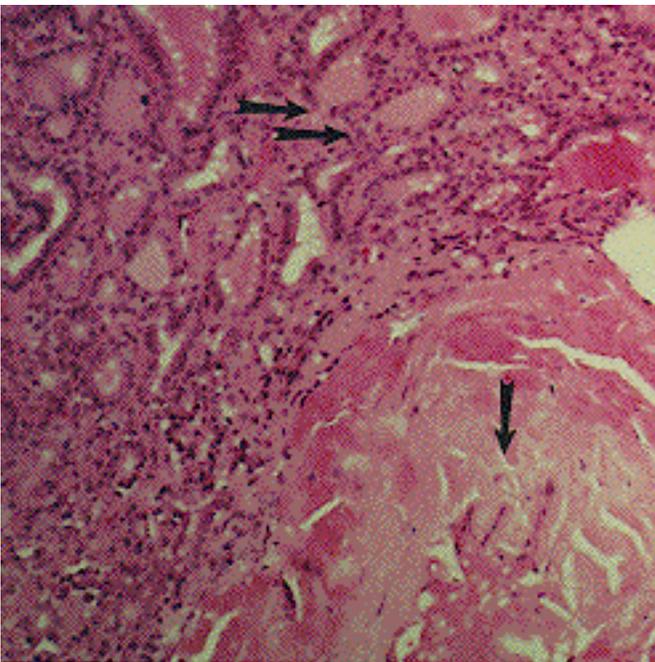


Amyloidosis, submandibular gland, Congo red stain showing deposition of amyloid in wall of blood vessels (arrow). Under a polarizing light this same material would show a green-yellow birefringence. Triangle indicates glandular tissue.





Amyloid. Note endothelium of blood vessel with red-staining amyloid in vessel wall. Amyloid typically produces a “smudged” appearance to any structure it affects.



Amyloidosis, thyroid; in addition to the circular deposit of amyloid (single arrow), normal thyroid follicles (double arrows) are also seen.

CLINICAL ASPECTS:

Amyloidosis in the head and neck area affects chiefly the larynx and tongue but may affect any site. It tends to be of the solitary rather than the systemic form. In the larynx, the most common complaint is hoarseness with a submucosal swelling most often along the true vocal cord, but sometimes in other areas of the larynx or hypopharynx. The tongue may become so infiltrated with amyloid that it cannot be retracted and part of the tongue may need to be resected. Local deposits of amyloid are also seen in the skin of any part of the body and in the head and neck area are most common in the periocular region. The lips and the neck and intraoral areas are also affected. In addition to head and neck sites, the kidney, spleen, liver, and heart frequently are involved. Also any area of the gastrointestinal tract from the tongue downward may be the site of amyloid deposition.

The classic method of diagnosing amyloidosis when a nodule of the disease was not readily available, was by rectal biopsy. It has also been found that labial salivary gland tissue shows deposition of amyloid in a high percentage of cases. Once the diagnosis is established, the patient must be evaluated to determine the type of disease and whether amyloidosis is present in any other part of the body.

There is no effective medical treatment. In the head and neck areas surgical treatment may consist of palliative measures to reduce swelling of the tongue or resection of a portion of the larynx to improve the airway.