# Localized Amyloidosis in the Nasopharynx and Neck: Case Report

## Nasofarinks ve Boyunda Lokalize Amiloidoz

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**ABSTRACT** Amyloidosis results from the deposition of amyloid proteins in organs and tissues. Localized amyloidosis in the head and neck is a rare and benign process. Amyloidosis involving the nasopharynx or neck is even rarer. In this paper, a case of localized amyloidosis of the nasopharynx and neck mimicking nasopharyngeal carcinoma with neck metastasis was reported with a literature review. The only complaint was nasal stuffiness. The diagnosis was made through nasopharyngeal biopsy and fine needle aspiration biopsy (FNAB) of the neck masses. Subsequent nasopharyngeal curettage was undertaken. No further treatment was given, and there was no evidence of disease in the 20th postoperative month. Amyloidosis, although rare, should be considered in the differential diagnosis of head and neck masses.

**Key Words:** Amyloidosis, nasopharynx, head and neck neoplasms

**ÖZET** Amiloidoz, amiloid proteininin doku ve organlarda birikmesi sonucu oluşur. Baş ve boyundaki lokalize amiloidoz nadir ve benin seyirlidir. Amiloidoz nazofarinks veya boyunda nadir görülür. Boyuna metastaz yapan nazofaringeal karsinomu taklit eden lokalize amiloidoz için literatürde tek vaka mevcuttur. Tek şikayeti burundan nefes alma güçlüğü olan hastada nazofaringiyal biyopsi ve boyundaki kitleden yapılan ince iğne aspirasyon biyopsisi sonuçlarına dayanılarak amiloidoz tanısı konuldu. Daha sonra nazofaringeal küretaj yapıldı, ek bir tedavi verilmedi. Yapılan takipler sonucunda, postoperatif 20 ay sonra hastalığın progresyon göstermediği görüldü. Amiloidoz nadir görülmesine rağmen baş boyun kitlelerinin ayırıcı tanısında düşünülmelidir.

**Anahtar Kelimeler:** Amiloidoz; nazofarinks; baş boyun neoplazmları

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Amyloidosis is a syndrome characterized by the deposition of an insoluble proteinaceous material in the extracellular matrix of one or several organs.[1,2] Rokitansky was the first to describe amyloid deposition in 1842; then Virchow, in 1853, termed the lesions as 'amyloid' in the belief that carbohydrate was the chief constituent.[3] Amyloid deposits have fibrous appearance with apple-green birefringence on polarized light microscopy after staining with Congo red.[1]

Several classifications of amyloidosis have been proposed. Systemic amyloidosis reflects the involvement of multiple organ systems such as the cardiovascular and gastrointestinal systems together with the lymph nodes, spleen, liver, kidneys and adrenal glands.[4] Systemic amyloidosis includes

three distinct forms: AL amyloidosis, associated with primary systemic amyloidosis; AA secondary amyloidosis associated with neoplastic or inflammatory conditions; and familial amyloidosis, which shows recessive inheritance.[5]

Localized amyloidosis in the head and neck is a rare and benign process. The most common sites of involvement are the larynx, subglottis and thyroid. Other sites include the orbit, salivary glands, paranasal sinuses, and oral cavity.[6]

A case of localized amyloidosis in the head and neck affecting both the nasopharynx and neck was reported. The presentation mimicked nasopharyngeal carcinoma with neck metastasis. To date, there has only been one similar case report in the literature.[7] This rare case was discussed with a review of the literature

## CASE REPORT

A 67-year-old woman presented to our department with nasal stuffiness within the last 3 years and bilateral neck masses for 2.5 years. The masses were 1-1.5 cm in diameter and have not grown larger for the past two years. Otoscopic examination, pure tone thresholds and tympanogram were normal. Nasal endoscopy and computerized tomography (CT) scan of the nasopharynx and the paranasal sinuses revealed a granular mass with irregular surface on the posterior wall of the nasopharynx (Figure 1). The histological examination of the nasopharyngeal biopsy and the fine needle aspiration biopsy (FNAB) of the neck masses revealed amyloidosis (Figure 2). Further examination for systemic diseases failed to demonstrate any evidence of systemic amyloidosis or any predisposing cause. The patient was on antihypertensive drugs for chronic hypertension and had no other additional medical problems. Cardiologic evaluation, including an electrocardiogram revealed normal results. A chest X-Ray, urinalysis, liver function tests, blood urea nitrogen, serum creatinin, complement factors, coagulation tests and serum protein electrophoresis were all within normal limits. Rectal biopsy and gastric polypectomy were negative for systemic involvement.

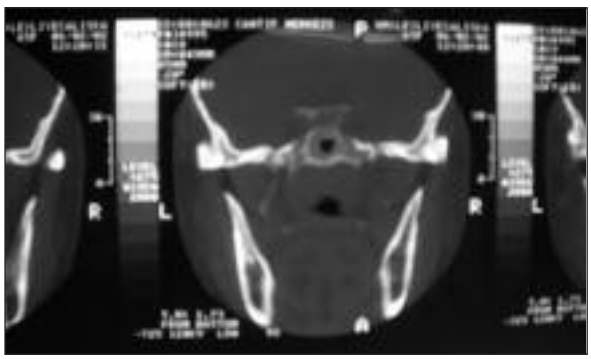


**FIGURE 1:** In the CT scan, the nasopharynx appeared to be obliterated by a granular mass with irregular borders.

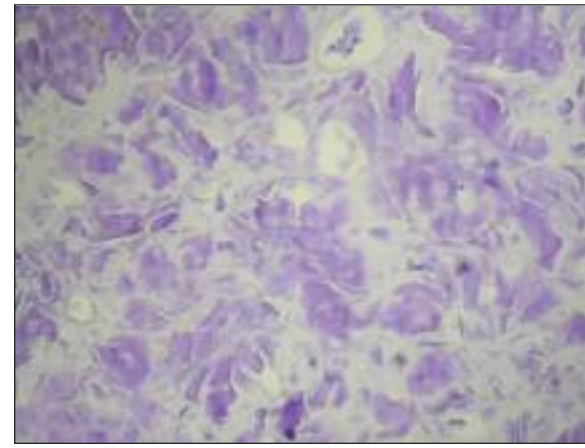

**FIGURE 2:** The fine needle aspiration biopsy from both cervical regions demonstrated an abundant amorphous homogenous acellular material, which stained positively with crystal violet (x40).

The nasopharyngeal mass was excised with sharp curettage and the pathological examination confirmed the diagnosis of amyloidosis. Microscopy revealed dense deposits of amyloid located in the submucosal region. Immunohistochemical analysis was positive for crystal violet and congo red indicating amyloidosis (Figure 3). No further treatment was given, and there has been no evidence of disease progression up to the postoperative month 20. The patient is still monitored regularly with nasopharyngeal endoscopy at 2-month-intervals.

## DISCUSSION

Amyloidosis can occur in almost every organ of the body. However, localized amyloid tumors in the head and neck region are extremely rare.[8] The lar-

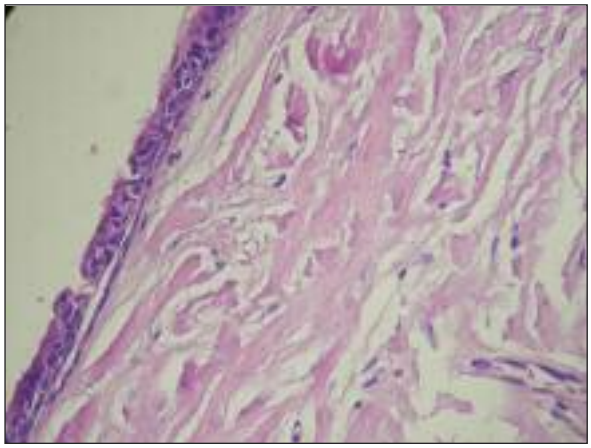

**FIGURE 3:** In excisional biopsy of the nasopharyngeal mass, submucosal material was observed which stained positively with Congo Red and Crystal Violet. This material was clearly seen in hematoxylin eosin (X40).

ynx is the most common site of involvement and amyloid deposits account for 0.2-1.5% of benign laryngeal tumors. Amyloidosis very rarely involves the nasopharynx or the neck.[7]

The clinical manifestations of amyloidosis are various and depend on the biochemical nature of the fibril protein and area involvement. In systemic amyloidosis, proteinuria is often the first symptom; other manifestations include peripheral neuropathies, dementia and cognitive dysfunction, and organ enlargement, especially of the liver, kidney, spleen and heart.[4,9] Macroglossia is most frequently noted in the head and neck region.[10]

Symptoms of localized nasopharyngeal amyloidosis include postnasal discharge, nasal obstruction, recurrent epistaxis, eustachian tube dysfunction and secondary ear problems such as conductive hearing loss due to middle ear effusion.[6] Our case presented with nasal stuffiness, but she had no symptoms or signs of ear disease. Interestingly, masses were found in the nasopharynx as well as the neck, mimicking nasopharyngeal carcinoma with multiple neck metastases.

In CT scans, amyloid deposits appear as relatively well-defined, submucosal, homogenous masses, usually demonstrating different forms of calcification. Calcification may also be seen within the lymphadenopaties in the neck.[11] In our case, CT scans showed no calcification within the nasopharyngeal mass or the adenopathies in the neck.

Diagnosis of amyloidosis is made with histopathological examination of amyloid deposits in biopsy specimens. Our case was diagnosed by FNAB of the nasopharyngeal and neck masses. The FNAB material stained positively with crystal violet and characteristic apple-green birefringence was demonstrated with congo red. However, when localized amyloidosis is suspected, systemic involvement should be excluded. Systemic amyloidosis is a serious and sometimes fatal condition and life expectancy is markedly shortened in such patients.[8] Renal failure and cardiac arrhythmias are the major causes of death.[10] Therefore, a biopsy from the head and neck that reveals amyloid necessitates evaluation for systemic involvement either by rectal biopsy (75% positive) or fat aspiration of the anterior abdominal wall (90% positive).[6] Specific organ involvement may also be excluded by laboratory or radiological studies.[7]

Unlike systemic amyloidosis, localized amyloidosis has an excellent prognosis. Surgery is reported to be the best therapy for localized amyloidosis.[8] Transpalatal approach or nasopharyngeal currettage is preferred. Excision of the amyloid deposits should only be considered if they cause morbidity; for the remaining cases of localized amyloidosis, conservative treatment with careful observation is suggested.[3] In the present case, the nasopharyngeal mass causing nasal obstruction was excised with curettage. The removal of the asymptomatic neck masses showing amyloid deposits in FNAB was considered unnecessary.

Sometimes, amyloid tumors in the nasopharynx may be difficult to treat and may recur despite surgical excision.[3] Simpson et al have reported less recurrence following laser excision of localized amyloidosis.[5] Localized amyloidosis is a disease of great rarity and slow progression, for which spontaneous regression is known to occur; therefore, it is difficult to predict the outcome.[6] Recurrent masses and extensive lesions involving the head and neck should be handled conservatively.[7]

In conclusion, amyloidosis, although rare, should be considered in the differential diagnosis of head and neck masses. Amyloid deposits in the head and neck necessitate further evaluation for systemic involvement. Localized excision is a good treatment option for localized amyloidosis, also providing an accurate diagnosis. However, nasopharyngeal amyloidosis is difficult to treat and tend to recur, requiring a long follow-up period. Therefore, conservative management should be considered in cases causing minimal morbidity.

## REFERENCES


1. Cathcart ES. Amyloidosis. In: Kelley W, Haris ED, Ruddy S, Sledge CB, eds. Textbook of Rheumatology. 4th ed. Philadelphia, Pennsylvania: WB Saunders Company; 1993. p.1413-28.
2. Zindancı İ, Kocatürk E, Koç M, Büyükbabani N, Kavala M. [A case of biphasıc amyloidosis: response to narrow band UVB]. Turkiye Klinikleri J Dermatol 2006;16(2):76-80.
3. Domínguez S, Wienberg P, Clarós P, Clarós A, Vila J. Primary localized nasopharyngeal amyloidosis. A case report. Int J Pediatr Otorhinolaryngol 1996;36(1):61-7.
4. Sipe JD, Cohen AS. Amyloidosis. In: Braunwald E,ed. Harrison's Principles of Internal Medicine, 15th ed. New York: McGraw Hill; 2001. p.1974-9.
5. Simpson GT 2ND, Strong MS, Skinner M, Cohen AS. Localized amyloidosis of the head and neck and upper aerodigestive and lower respiratory tracts. Ann Otol Rhinol Laryngol 1984;93(4 Pt 1):374-9.
6. Patel A, Pambuccian S, Maisel R. Nasopharyngeal amyloidosis. Am J Otolaryngol 2002;23(5):308-11.
7. Zhuang YL, Tsai TL, Lin CZ. Localized amyloid deposition in the nasopharynx and neck, mimicking nasopharyngeal carcinoma with neck metastasis. J Chin Med Assoc 2005;68(3):142-5.
8. Pitkäranta A, Malmberg H. Localized amyloid tumor of the nasopharynx. Otolaryngol Head Neck Surg 2000;122(2):309-10.
9. Balal M, Paydaş S, Erdoğan Ş, Karayaylalı İ, Seyrek N. [Transthyretin amyloidosis and hypopotasemia:a case report]. Turkiye Klinikleri J Med Sci 2006;26(1):90-2.
10. Nandapalan V, Jones TM, Morar P, Clark AH, Jones AS. Localized amyloidosis of the parotid gland: a case report and review of the localized amyloidosis of the head and neck. Head Neck 1998;20(1):73-8.
11. Gean-Marton AD, Kirsch CF, Vezina LG, Weber AL. Focal amyloidosis of the head and neck: evaluation with CT and MR imaging. Radiology 1991;181(2):521-5.