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Review

Localised, Isolated Amyloidosis of the Nose and Paranasal Sinuses

Georgi Iliev[1], Polina Ivanova[1*], Kenan Kerimov[1], Petya Genova[1] and Kalin Kalchev[2]

[1]Department of ENT, University Hospital Saint Marina, Medical University of Varna, Varna, Bulgaria

[2]Clinic of General and Clinical Pathology, University Hospital Saint Marina, Medical University of Varna, Varna, Bulgaria

Abstract

Amyloidosis is mainly a systemic disease, hence primary localised amyloidosis of the nose and sinuses may occur. Symptoms may include epistaxis, nasal obstruction, facial deformity and vision changes.

We present a case of isolated, localised sinonasal amyloidosis in 81-year old woman and a brief literature review.

After an extensive evaluation process and a histology verification the final diagnosis was concluded – sinonasal amyloidosis. Treatment with radiotherapy showed excellent outcomes as the process was halted and the patient reported improved breathing.

Localised amyloidosis is a benign, slowly growing process. Although it is a rare condition, nasal amyloidosis must be considered in the differential diagnosis of nasal obstruction.

Keywords: Amyloidosis; Localised; Nasal obstruction; Radiotherapy

Introduction

Amyloidosis is mainly a protein deposition disease, diagnosed on clinical signs, symptoms and histopathology. Rarely the disease may occur on localised basis in the head and neck region affecting the larynx, pharynx, orbit, nasal cavity, sinuses and oral cavity. As one of the most uncommon locations is the nasopharynx and the sinuses, very few cases are reported in the literature. Our case described here is about isolated localised sinonasal amyloidosis. The purpose of this case report is to present a rare manifestation of an unusual systemic disease as a localised form and to draw attention to the recognition of this uncommon condition.

***Corresponding author:** Polina Ivanova, Department of ENT, University Hospital Saint Marina, Medical University of Varna, Varna, Bulgaria, Tel: +35 9896863941; E-mail: ppivanovax@gmail.com

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Case Report

An 81-year-old female presented to the ENT clinic at the University Hospital of Varna 'Saint Marina' due to difficulty nasal breathing since several months and a slight facial deformity regarding the nasal bridge. Her general condition was good and she did not have any other complaints, or any underlying known disease such as hypertension or diabetes mellitus. The patient's nasal obstruction coincided with the time of appearance of the facial deformity (Figure 1).

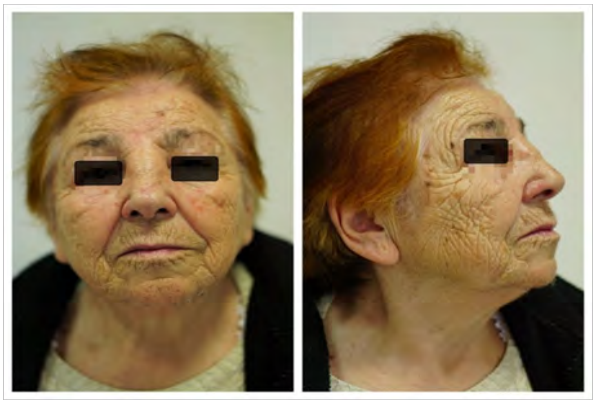

Figure 1: Image of patient's face deformity regarding the nasal bridge.

The evaluation process included routine blood test, which did not present anything abnormal. Anterior rhinoscopy revealed a bulging mass engaging the nasal meat uses. Further, a Computed Tomography (CT) scan was appointed. The findings showed an advanced neo-process, presenting as heterogeneous mass, engaging the nasal meat uses at the level of the middle and superior turbinates, spreading along the frontal sinuses, with destruction of the base of the latest, an osteolysis of the nasal bones bilaterally and the anterior walls of the anterior ethmoidal sinuses, and a suspicion for the zone of destruction in the medial wall of the right orbit (Figure 2). For further diagnostics, a biopsy was performed under endoscopic control and local anaesthesia with 2% Lidocaine. The histological examination displayed fragmented parts of tissue with respiratory epithelium, sub mucosal diffuse deposits of eosinophilic, amorphous fissured material, consistent with an amyloid structure; a periglandullar inflammation (Figure 3). Further investigations performed by the internal medicine specialists did not show systemic involvement.

Results

The histopathological conclusion confirmed the diagnosis - a localised amyloidosis. Based on the location and local engagement of the amyloidosis the patient was classified as non-operable. Due to that, it was decided to be used only radiation therapy for treating the process.

The patient received a full course of external beam radiotherapy with a total amount of 36 Grey. A following CT-scan was performed

9 months after the diagnosis and the treatment with radiation. The findings revealed no enlargement of the process. External radiotherapy appears to halt the progression of isolated localised sinonasal amyloidosis. The patient also reported improved nasal breathing.

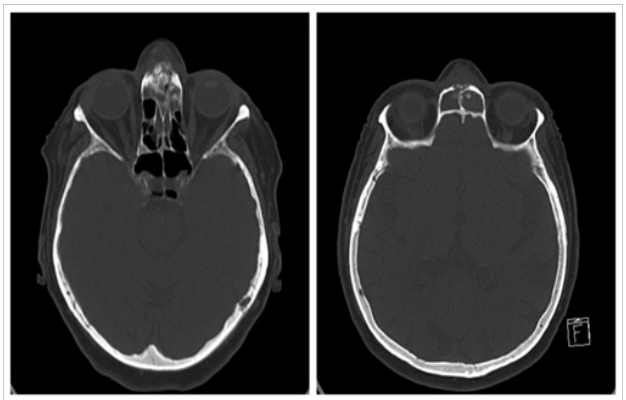

Figure 2 (left): Axial CT image of the patient showing engagement of the nasal meat uses.

Figure 2 (right): Axial CT image of the patient showing engagement of sinus frontal is with osteolysis.

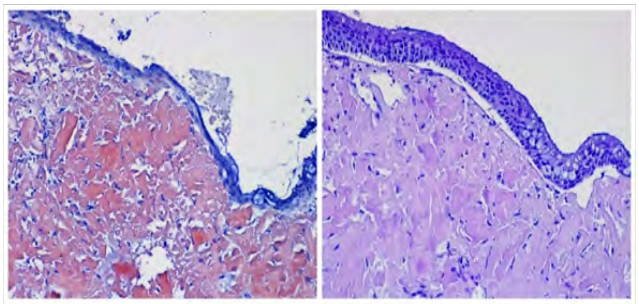

Figure 3 (left): Histology of amyloidosis from tissue section from the nasal meat uses after straining with Congo × 100.

Figure 3 (right): Histology of amyloidosis from tissue section from the nasal meat uses after straining with H&E × 100.

Discussion

Amyloidosis is the term for a group of protein folding disorders characterised by the extracellular deposition of insoluble polymeric protein fibrils in tissues and organs. Amyloid diseases, defined by the biochemical nature of the protein composing the fibril deposits, are classified according to whether they are systemic or localised, whether they are acquired or inherited, and their clinical patterns [1].

Amyloidosis is a rare disease with an incidence of 12 cases per million populations per year [2]. The main subtypes of amyloidosis are primary AL amyloidosis, secondary Amyloid A (AA) amyloidosis, familial amyloidosis, and ß2-microglobulin-related amyloidosis. The diagnosis of amyloidosis is based on clinical organ involvement and histological evidence of amyloid deposits [3].

Rarely, however, amyloidosis may be present in a localised form. When occurring on a localised basis in the head and neck region, it can involve the orbit, sinuses, nasopharynx, oral cavity and larynx, the latter of which is the most common head and neck presentation of localised amyloidosis.

Isolated local nasal amyloidosis is extremely rare and idiopathic. In the literature are reported very few cases regarding localised nasal amyloidosis. The youngest patient described in the literature that we occurred with localised nasal amyloidosis was 8 years old, whereas our patient is one of the oldest with the age of 81 years [4]. Another young female patient was at the age of 14 years [5]. A clear definition, in reference with age group or gender predominance, has not been found yet. As seen from the previous cases every age group may be affected by the disease.

With so few information from the literature review, little is known of this specific condition, its aetiology and the initial factors. Given that the most common localization of localised amyloidosis is at mucous membranes in contact with the environment, it has been suggested there is an antigenic induction of amyloidogenic plasma cell clones [6]. A chronic inflammation has often been encountered also as an etiological factor [7].

Localised amyloidosis usually presents as one solitary volume occupying lesion, although multiple nodules may appear. The symptoms may vary depending on location of the formation. In its localised form amyloidosis resembles a malignant process. Calcifications are also common, radiologic appearance may include micro calcifications, increasing the suspicion of malignancy. In sinonasal amyloidosis symptoms usually are unspecific such as nasal obstruction, recurrent epistaxis, mucoid discharge, Eustachian tube disorder, facial deformity, as in our case the patient the complaints were nasal obstruction and facial deformity. Localised AL amyloidosis can be treated with radiation therapy and depending on the localisation; some forms may be amenable to surgery. The overall survival of localised AL amyloidosis is excellent and strikingly different from systemic AL amyloidosis. Progression to systemic disease is an exceptionally rare occurrence [8].

Conclusion

We present the case of an exceptionally rare, isolated, idiopathic, localised form of sinonasal amyloidosis with destruction of the bony parts of both the frontal and ethmoidal sinuses. The diagnosis of such atypical condition is difficult and troubled due to the unspecific signs and symptoms and their similarity to other more common conditions affecting the nasal region. Extensive evaluation process should be performed as biopsy is the only confirmative method. Localised amyloidosis does not respond to medications, but depending on the patient's condition. Treatment options include surgical intervention and radiotherapy. In our case, as the patient was inoperable, radiotherapy was applied. Radiotherapy can be used in order to halt the process and get the disease into remission.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Conflict of interest

None declared. The authors have no financial, consultative, institutional, and other relationships that might lead to bias or conflict of interest.

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