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Huge Localized Amyloidosis of the Sinonasal Cavity: A Rare Case Report

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Abstract

Background: Amyloidosis is the general term for deposition of fibrous abnormal protein aggregates (amyloid) in organs or tissues that leads to dysfunction of the affected organs. Treatment and follow-up of localized amyloidosis are particularly difficult because its pathogenesis is still unclear. Localized amyloidosis of the head and neck is rare and that of the nose is rarer still. Here, we report a case of localized amyloidosis of the nasal cavity, which affected an area larger than those in previous reports.

Case presentation: A 60-year-old woman presented with the chief complaints of headache, proptosis and nasal congestion. There was a lack of response to chemotherapy and endoscopic sinus surgery was performed. As of 18 months after surgery, subjective symptoms have been alleviated and are consistent with the objective findings. There have been no signs of recurrence.

Conclusions: Cases of localized amyloidosis of the nasal cavity are rare, and standard treatment has not yet been established. In this case, proactive surgical intervention was successful for this condition.

Keywords: Amyloidosis; Sinonasal; Treatment; ESS

Introduction

Amyloidosis is the general term for deposition of fibrous abnormal protein aggregates (amyloid) in organs or tissues that leads to dysfunction of the affected organs. Treatment and follow-up of localized amyloidosis are particularly difficult because its pathogenesis is still unclear. Localized amyloidosis of the head and neck is rare and that of the nose is rarer still. Here, we report a case of localized amyloidosis of the nasal cavity, which affected an area larger than those in previous reports.

Case Report

A 60-year-old woman was referred to our department with a suspected tumor nasal cavity and sinus. She had no significant past history such as rheumatoid arthritis, and her family medical history was negative for conditions such as multiple myeloma. She had been receiving conservative treatment for nasal blockage and discomfort around the eyes since 2005 at nearby otolaryngology clinic with the diagnosis of chronic sinusitis. Swelling of the upper right eyelid and eye pain occurred around 2010. After a sinus tumor was suspected based on imaging findings, she was referred to our department in November 2013. Initial CT showed a destructive and expansive tumor in the nasal cavity advancing across the sinuses with punctate calcification in some parts (Figures 1a and 1b). The tumor extended into the left orbit, resulting in eye movement disorder and proptosis. Hertel proptosis meter measurements were 13.55 for the right eye and 18.51 for the left eye. Both visual acuity and intraocular pressure were normal. Both nasal cavities were filled with masses accompanied by crusting and partially yellow granular nodules under the mucosa. Adhesions were also present, so the nasopharynx could not be seen even with an endoscope (Figure 1c). Nasal endoscopic biopsy revealed direct fast scarlet-positive deposits in the stroma, and a many eosinophilic amorphous deposits that had a greenish-yellow tone under polarized light (Figure 2). These findings were consistent with amyloidosis. Based on the tissue liquid chromatography-tandem mass spectrometry findings, AL-kappa amyloidosis was diagnosed. No abnormalities were detected on the following tests performed in the internal medicine department: digestive tract random biopsy using a gastro scope and colon fiber; echocardiography; neck, thoracic, abdominal and pelvic CT; abdominal fat biopsy; free light chain analysis; and

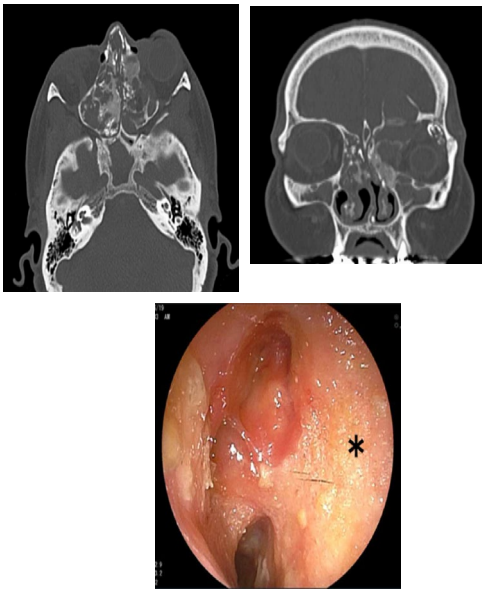

Figure 1: Preoperative computed tomography scan (a) axial and (b) coronal views. Endoscopic view (preoperative): (c).
(a) and (b) Soft shadows are evident in sinuses on both sides with calcification in some parts. Partial bone destruction and a tumor extending into the left eye socket are apparent, as is a projection towards the front of the left eye.
(c) Both nasal cavities are filled with masses accompanied by crusting and partially yellow granular nodules under the mucosa. In addition, intranasal adhesion is present, preventing observation of the nasopharynx by an endoscope. Asterisk indicates the nasal septum.

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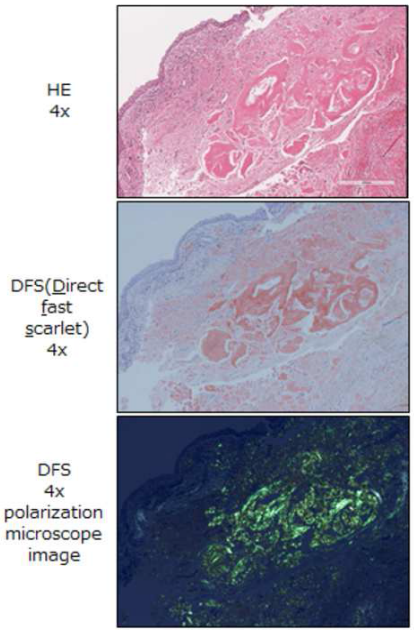

Figure 2: Pathological examination. Direct fast scarlet (DFS)-positive deposits in the stroma, and a significant amount of eosinophilic amorphous deposits in a greenish-yellow tone under polarized light.

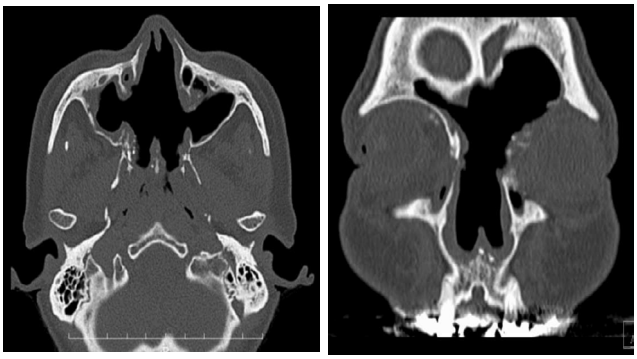

Figure 3: Postoperative computed tomography scan: (a) axial and (b) coronal views. (a) and (b) Mucosal thickening is evident in some parts of the sphenoid sinuses, but there are no signs of obvious recurrence. The paranasal communication is preserved.

immunofixation electrophoresis. No obvious amyloid deposition was found elsewhere in the body, indicating the strong possibility of localized amyloidosis in the nasal cavities. Further, the patient showed no elevations of plasmin-α2-plasmin inhibitor complexes (PIC) and thrombin-antithrombin (TAT) levels, which have been reported in systemic amyloidosis only and which are considered useful markers in differentiating systemic from localized amyloidosis [1]. The options of craniotomy to repair spinal fluid leakage and resection to remove frontal sinus lesions were explained to the patient, together with possible esthetic problems (e.g. saddle nose and displacement of the eye ball), visual loss and diplopia. The patient refused surgery. Based on a small number of cases of transition to localized amyloidosis during the follow-up of systemic amyloidosis [2], and the difficulty in distinguishing local and systemic amyloidosis at the early stage of the disease, she underwent chemotherapy in the Department of Collagen Disease, Internal Medicine Division, in January 2014. The regimen consisted of four 28-day cycles of melphalan 8 mg and dexamethasone 40 mg on days [1-3] .The patient did not respond to chemotherapy, and gradual growth of tumors and aggravation of headache, double vision and nasal blockage were observed. The patient then agreed to undergo surgery in our department. Due to the possible risk of leakage of spinal fluid, endoscopic surgery was performed by otolaryngology surgeons with neurosurgical backup.

Intraoperative Findings

The nasal walls of both sides of the maxillary sinuses were drilled with a diamond burr with preservation of the nasolacrimal duct. Maxillary sinuses were filled with pus, probably due to secondary sinusitis. Amyloid deposits were scattered under the mucosa, and they were removed as much as possible. Frontal sinus surgery was performed using a navigation system, and interseptal sinus cells were removed, thereby creating a large communication (according to the modified Lothrop procedure). There were aggregates of bones and soft tissue, inside of which were yellow amyloid deposits that collapsed easily by drilling. The frontal sinuses and interseptal sinus cells were also filled with pus, probably due to secondary sinusitis. The periosteum and dura mater were preserved, albeit with partial bone thinning. A cranial communication was absent. Then, the drill was moved through both the anterior and posterior ethmoid sinuses, which were filled with amyloid deposits, using the navigation system because the anatomical landmarks were lost. Obstacles collapsed easily during the procedure. Both sides of the anterior ethmoid artery were identified and preserved. A communication between all paranasal sinuses was created. The pathological mucosa with amyloid deposits underneath was removed as much as possible. Both sides of the nasal turbinates were not identifiable due to adhesion to the nasal septum, and thus they were removed en bloc.

Postoperative Course

The patient was free from postoperative complications and discharged 1 week after surgery. 1 year after surgery, mucosal thickening was observed in some parts of the sphenoid sinuses, but signs of obvious recurrence were absent. CT did not show recurrence, and the preservation of a communication was confirmed (Figure 3a and 3b). Headache, the most prominent symptom among her chief complaints, was relieved. Protrusion and shifting of the eyeballs were alleviated, resulting in evenly balanced right-left eye positions. The results of the Hess test performed 5 months after surgery also showed no difference between the right eye (14.63) and left eye (15.54). The patient was very satisfied with the outcomes. No signs of recurrence have been observed as of 18 months after surgery.

Conclusions

Amyloidosis is a general term for the deposition of fibrous abnormal protein aggregates (amyloid) in certain organs or tissues that leads to the dysfunction of the affected organs [4]. It is classified into two types: systemic amyloidosis characterized by amyloid deposition in multiple organs, and localized amyloidosis characterized by amyloid deposition in a particular organ or tissue. It is generally believed that the prognosis of localized amyloidosis is favorable, whereas that of systemic amyloidosis is poor, as exemplified by an expected survival of 13 months after onset. Thus, differentiating whether amyloidosis is systemic or localized is extremely important [5]. Barteles et al. recommended systemic examination including a detailed medical history (including family history); physical examination to detect hepatosplenomegaly, heart failure, edema and lymphadenopathy; blood and urine tests (erythrocyte sedimentation rate, liver function, immunoglobulin, rheumatoid factor, serum monoclonal protein, urine

protein, urine monoclonal protein); and histological examination of the gastrointestinal tract and abdominal subcutaneous fat [6]. In this study, localized amyloidosis was diagnosed after a thorough systemic examination. The pathogenesis of localized amyloidosis is unclear, so providing treatment and care during follow-up is extremely difficult. Localized amyloidosis of the head and neck is rare. In particular, only 15 cases of localized amyloidosis of the nasal cavity have been reported [7,8] and its treatment has not been established. The area affected by amyloid deposition in our case was larger than those in previous reports. Amyloidosis is subdivided by the biochemical classification of accumulative amyloid proteins, such as the AL type (immunoglobulin light chain fragments are deposited) and the AA type (serum amyloid A protein fragments are deposited [1]. Many cases of amyloidosis of the head and neck are the AL type. Deposition can occur in any part of the head and neck but is most frequent in the larynx. Unlike the pathology of systemic AL amyloidosis, marked infiltration of plasma cells is often observed in localized AL amyloidosis [9]. Definitive diagnosis is based on direct evidence of the presence of amyloid in pathological specimens. However, special care needs to be taken to include the submucosa when collecting specimens, otherwise amyloid deposits that are usually under the mucosa will not be detected. Indeed, we performed biopsy several times before reaching the definitive diagnosis in our case.

In principle, surgery is recommended for localized amyloidosis. Recurrence of laryngeal amyloidosis, which is a more common type of head and neck amyloidosis than localized nasal amyloidosis, is not rare, and thus debulking was recommended for its treatment [10]. In our patient, localized nasal amyloidosis that affected an area larger than those in previous reports involved part of the cranial base. Conservative treatment was initially chosen, taking into account the patient's request. However, after a lack of response to chemotherapy and aggravation of headache, double vision, proptosis and nasal congestion, we finally opted for surgery. Open surgery was not required and all procedures were endoscopic. The navigation system was very useful since most of the anatomical landmarks had been destroyed. Extensive drilling with a 4-hands technique was required, and a good operative field was obtained. The modified Lothrop procedure was used to prevent possible recurrence and osteogenesis [8], and secondary sinusitis. The outcomes of the creation of a large communication between sinuses were favorable: signs of recurrence were absent 18 months after surgery, and the patient was satisfied with the alleviation of subjective symptoms (e.g., headache and visual field defects) and improvement of esthetic aspects (e.g., proptosis). We initially offered nonsurgical treatment, namely, chemotherapy based on the risk of surgical complications and the patient's preference. However, the therapeutic effect was poor, and early surgical intervention would likely have been more beneficial in this case. Long-term follow-up is necessary, considering the high recurrence rate and possible transition of localized amyloidosis to systemic amyloidosis [11].

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