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Case Report

Localized nasopharyngeal amyloidosis: A case report ☆

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ABSTRACT

Amyloidosis is a rare condition on its own but finding localized amyloidosis of a site such as the nasopharynx is an extremely rare condition with very few cases described in the literature. The condition occurs due to the accumulation of misfolded proteins in the extracellular space disrupting the cell architecture and causing eventual dysfunction. In this case report we discuss the pathophysiology, symptoms, and imaging findings of a patient initially thought to have giant cell arteritis, but who was found on CT to have a mass later determined to be localized nasopharyngeal amyloidosis. Evaluation will require biopsy as it is the gold standard, but there are many other tests and even reasons to consider the use of interventional radiology to sample other tissues rather than the primary target site for amyloid deposition in conjunction with nuclear imaging.

Introduction

Amyloidosis is a rare disease with an incidence of approximately 1300 new cases in the United States every year, yet its effects on patients are far-reaching and quite consequential [1]. This disease stems from an extracellular accumulation of amyloid, proteins of divergent shape and structure, either systemically or in a certain target organ, often including the kidneys and liver depending on specific amyloid composition [1]. The amyloid formation may occur through many pathways, but 2 of the most common pathways involve AL and AA amyloid. AL amyloid, the more common of the 2, signifies the amyloid is of light chain origin while AA signifies that the amyloid comes from the amyloid A protein [1]. Therefore, risk factors for these forms of acquired amyloidosis are related to the causes of their subtype protein, thus AL is associated with plasma cell accumulation such as multiple myeloma and AA is associated with causes of chronic inflammation such as various rheumatologic diseases [2]. Risk will also increase with age as cells become more prone to error in protein synthesis [3]. Differentiation between subtypes is obtained by immunohistochemistry [2]. Localized amyloid deposition is rare, but the believed reason for why it may occur is the same as that of AL amyloidosis, but due to local processes or genetic factors only affecting the local tissue environment the extent of abnormal light chain build-up is limited [4]. The damage from amyloid once accumulated in a target organ is thought to be direct toxicity from the amyloid components or the disruption of the very structural constitution of the cells [5].

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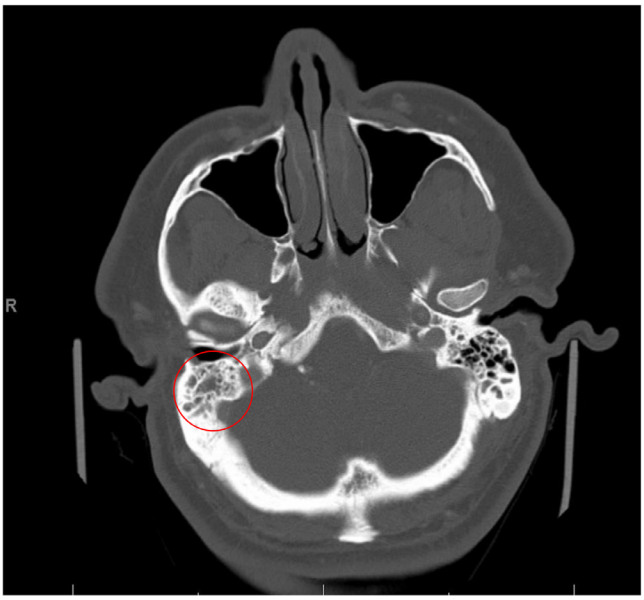

Fig. 1 – Opacified right mastoid air cells shown by red circle as seen on initial axial view of CT maxillofacial study.

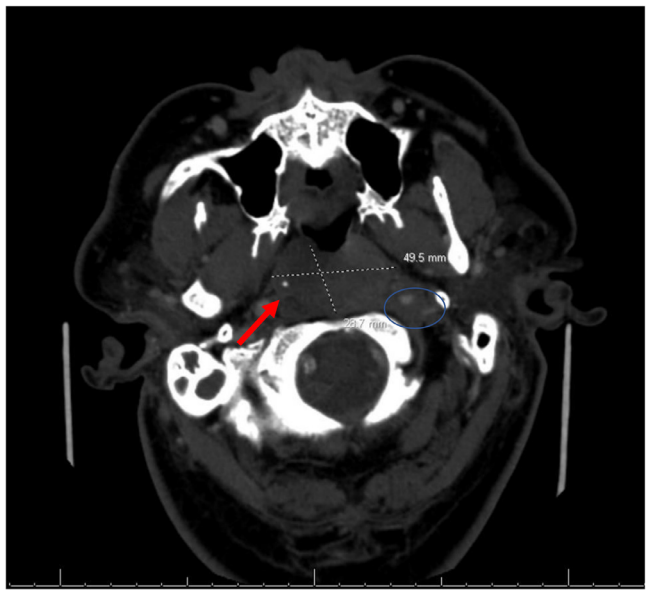

Fig. 2 – Axial view of the head from CT maxillofacial study which first identified the nasopharyngeal soft tissue thickening pointed out in red. The blue circle highlights the involvement of the carotid canal and bony erosion of the skull.

Amyloidosis after sufficient amyloid deposition has occurred may manifest as cardiac, renal, and liver damage/ failure as well as macroglossia, easy bruising, periorbital ecchymosis, or even carpal tunnel [1].

Case presentation

Our patient is a 60-year-old male with a past medical history of coronary artery disease, hyperlipidemia, type 2 diabetes, and hypertension who initially presented with a reported 1-month long history of intractable left-sided temporal headaches. For 3 weeks, our patient has also experienced intermittent nasal congestion and foul-tasting nasal discharge that was responsive to Fluticasone nasal spray. He was believed to have potentially had giant cell arteritis given headache distribution and was given prednisone which provided transient pain relief, however, the headaches soon returned. Ophthalmologic exam was unremarkable and ESR and CRP values were not significantly elevated. CT head revealed mucosal thickening of his sphenoid sinuses and ethmoid air cells and complete opacification of the right mastoid air cells (Fig. 1) with a possible mass in his nasopharynx. Endoscopy revealed irregular tissue in the nasopharynx with mucopus in the area previously noted as a possible mass by CT. Repeat CT reveals opacification of the right mastoid air cells, consistent with previous scans, with partial opacification on the left mastoid air cells and an enhancing soft tissue mass with calcifications in the nasopharynx measuring 5 × 2.9 cm (Fig. 2) and left lower cervical lymphadenopathy, 0.96 cm (Fig. 3) in the short axis. It was believed that the mass may be the cause of the opacification of the mastoid due to the mass effect on the eustachian tube. Two months later, the patient presents to the hospital due to feeling that he cannot move the left side

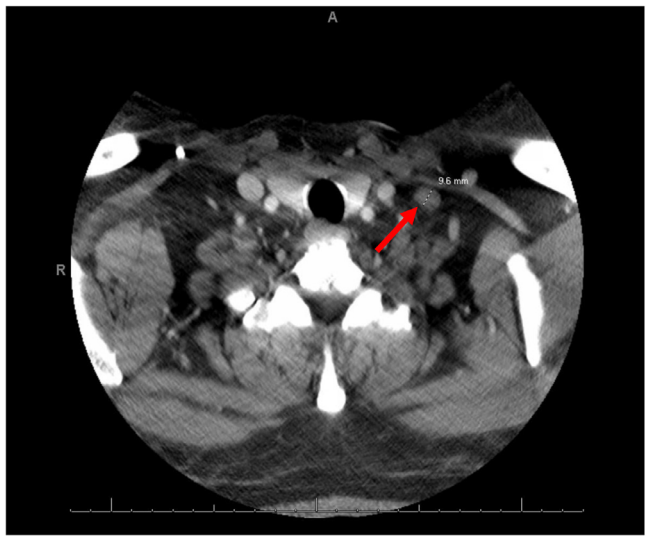

Fig. 3 – Axial view from CT Neck study showing enlarged cervical lymph node measuring 0.96 cm.

of his tongue accompanied by otalgia, tinnitus, and hearing loss on his left side. Laryngoscopy at that time reveals the mass is located at the right fossa of Rosenmuller. The patient later underwent laryngoscopy and biopsy with pathology revealing amyloidosis with a sample showing apple-green birefringence on Congo red staining and visualization of calcified retropharyngeal lymph nodes and CT showed skull base erosion and inclusion of the left cervical internal carotid artery

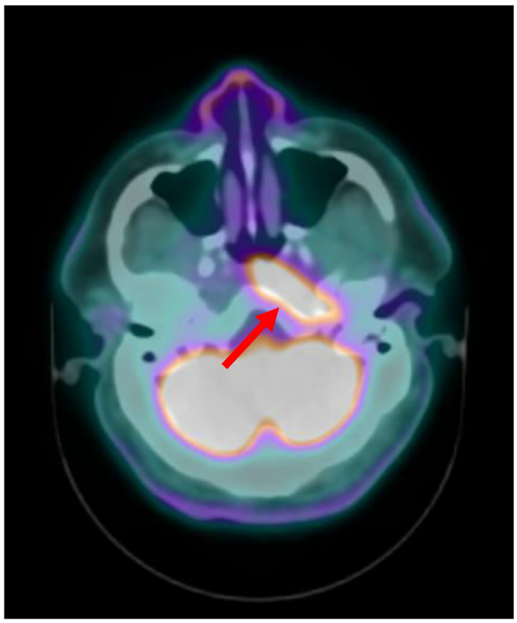

Fig. 4 – Axial view of the skull from the PET imaging study showing the mass pointed out by the red arrow in the left nasopharynx.

canal as well as the hypoglossal canal. Flow cytometry was also performed to rule out a possible plasma cell dyscrasia. Serum electrophoresis shows hypogammaglobulinemia and urine electrophoresis was unremarkable. Kappa quant free light chain showed elevated free light chains. PET-CT using F-18 radiotracer was ordered to evaluate for potential systemic disease which indeed showed our initial lesion (Fig. 4) came back negative for any indication of systemic involvement. Ultrasound-guided fine needle abdominal fat and fluoroscopic right posterior iliac bone marrow biopsy were also performed by interventional radiology to rule out systemic disease.

Discussion

There were a recorded 38 cases of pure nasopharyngeal amyloidosis in the literature as of 2019 [6]. Two more cases appear to be seen in the literature since that date based on a PubMed search. Biopsy and Congo red staining with an expected apple-green birefringence of the tissue is used to verify amyloid deposition [7]. While this is the gold standard, certain tests such as serum protein electrophoresis and urine collection and protein electrophoresis can help identify a majority of patients likely to have amyloidosis [8]. The hypogammaglobulinemia seen in our patient is actually seen to be a finding in amyloidosis; however, other more classic findings associated with amyloidosis are the detection of certain monoclonal proteins in either serum or urine [9].

If there is a concern for damage to the organ of deposition in tissue collection then certain areas may preferentially be sampled in its stead by interventional radiology such as the abdominal fat, rectum, and bone marrow; however, there is the tradeoff of variably decreased sensitivity as these are often quite removed from the site of interest [8]. Thus, when damage to the native tissue is a serious concern, sampling from the abdominal fat would be a reasonable first choice as it has the highest sensitivity of the options previously listed [8]. Systemic disease is treated based on amyloid type with AL being managed by treating the cause of abnormal light chain deposition such as multiple myeloma which may require chemotherapy, treating the chronic inflammatory condition often seen in AA amyloidosis, organ transplantation in hereditary amyloidosis, or in conjunction with the previous treatments we may consider immunotherapy or surgery [2].

Currently, the recommendation for a patient with localized amyloid deposition of the nasopharynx with significant interference in their quality of life is the surgical removal of the mass from the site of burden, however, this must be balanced carefully with patient wishes and possible complications due to the surgery itself as well as to the removal of significant amounts of parenchymal tissue [10]. When there is a significant mass or if there were even greater involvement of the surrounding structures than was seen in our patient then symptomatic treatment may be preferred as this is a very slowly progressive disease. While our patient does have involvement in the carotid and hypoglossal canal, it was still determined that surgical intervention was still viable.

PET-CT imaging may be used to evaluate for possible systemic disease as certain tracers may be able to identify disseminated amyloid throughout the body [11]. As in our case, the radiotracer did not identify any areas outside of the nasopharynx to have accumulated amyloid, but of course, there is not yet a gold standard test for evaluating systemic disease. However, this in conjunction with negative biopsy samples from abdominal fat rectal, or bone marrow helps greatly increase the likelihood that the disease is localized. In general, cross-sectional findings in amyloidosis include wall thickening if the intestines are involved, splenomegaly, enlarged colon, cardiac wall thickening, pulmonary interstitial thickening, and pulmonary as well as other organ calcifications [12]. As seen from these nonspecific findings, amyloidosis does not have clear and direct imaging findings. The radiologist must maintain a high degree of suspicion and a broad differential to include this rare condition as a possibility for these abnormal findings. Serum amyloid protein scintigraphy, if available, may be useful as it is fairly specific to amyloid, but it may be less readily accessible to both institutions and patients [13]. Thus, based on the options available PET-CT might be a more reliable study in conjunction with the other tests mentioned above.

As in previous cases of nasopharyngeal amyloidosis, if the mass can be safely removed then the general prognosis should be positive as there is not much concern for recurrence [10]. Consequentially, if our patient undergoes surgery to debulk the mass, they should be able to maintain a fairly high quality of life and, even if they were to decide not to go ahead with surgery due to personal reasons, the disease would be very slow to progress, and we may assist with treating symptoms instead.

Conclusion

Nasopharyngeal amyloidosis is an exceedingly rare condition with very few recommendations in the literature. Having a detailed evaluation as listed above and as supported by the literature will help to ensure the best care for our patients.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report.

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