

Primary Amyloidoma of Meckels Cave: Case Report

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Abstract

BACKGROUND:
Amyloidoma within the central nervous system is a rare entity. This case presentation involves a 47 year old female with a left sided Meckels cave primary amyloidoma.

CLINICAL PRESENTATION:
A 47 year old female presented with four years of trigeminal pain and paresthesia in the V1-V3 distribution. She had a MRI, which revealed an intensely enhancing mass in the left cavernous sinus. She was referred to Neurosurgery and Otolaryngology for biopsy. Surgical pathology revealed a primary amyloidoma.

CONCLUSION:
This case presentation involves a patient who presented with signs of idiopathic trigeminal neuralgia and was later found to have a primary amyloidoma of Meckels cave. This patient presented without signs of systemic amyloidosis. It is rare to have a primary amyloidoma involving the trigeminal ganglion, however it must be kept on the differential even if the patient does not show signs of systemic disease.

Introduction

Amyloidoma within the central nervous system is a rare entity. Our review of literature showed only 52 reported cases of central nervous system amyloidomas. To have a lesion involve the trigeminal ganglion is even more rare; only 13 cases have been reported on our review. This case presentation involves a 47 year old female with a left sided Meckels cave primary amyloidoma.

Case Presentation

A 47 year old female presented with four years of trigeminal paresthesia in the V1-V3 distribution. She had multiple negative MRIs over that time span, and was started on gabapentin. She eventually developed pain leading her PCP to obtain another MRI. At this time MRI revealed a 8 x 15 x 20 mm intensely enhancing mass in the left cavernous sinus. She was referred to Neurosurgery and Otolaryngology for possible biopsy vs removal. Based on imaging and exam it was thought that she had a trigeminal nerve schwannoma. She underwent a combined procedure with a transsphenoidal approach by Otolaryngology and removal of Meckels cave mass by Neurosurgery. Surgical pathology was consistent with a primary amyloidoma (Figures 3, 4, and 5).

Imaging

MRI revealed an enhancing lesion of the left Meckel's cave with extension along V3 through the foramen ovale and along V2 through foramen rotundum. The lesion was hypointense on T2 as shown below. Figure 1 shows the T1 weighted MRI and Figure 2 shows the T2 weighted imaging.

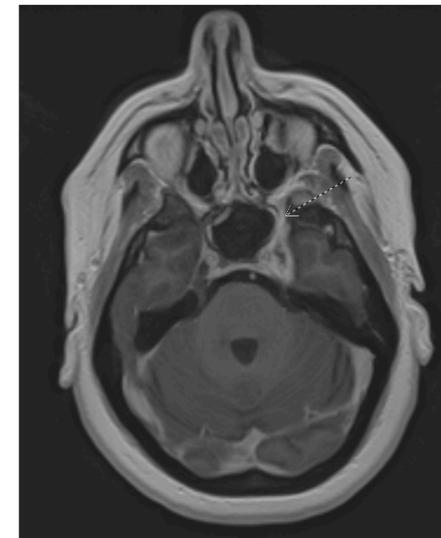


Figure 1. MRI T1 Axial

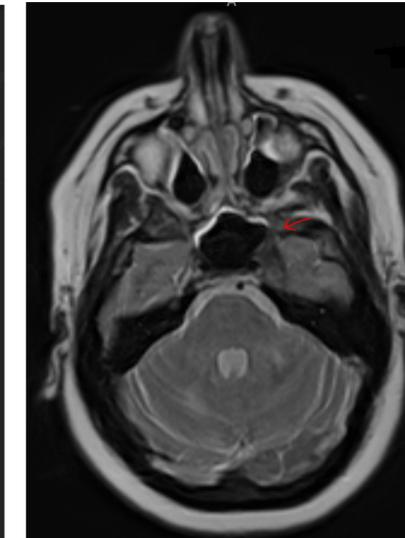
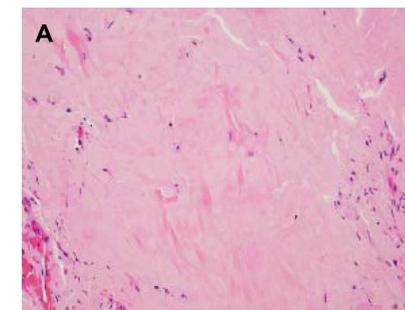
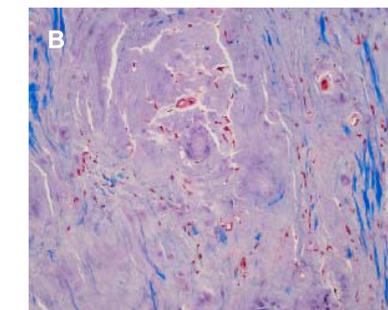


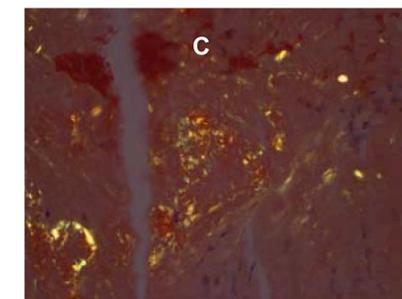
Figure 2. MRI T2 Axial



Amorphous material



Trichrome



Apple green birefringence

Figure 3. Pathology Slides

Pathology

Results from the surgical pathology specimen were reported as amyloidosis consistent with a primary amyloidoma. Figure 3 shows three different samples from the pathology specimen. Figure 3 A shows the amorphous material, B is the trichrome staining of the specimen, and C reveals the typical apple green birefringence.

Discussion

Amyloidosis is due to extracellular deposition of amyloid fibril proteins, which can occur systemically or as a localized deposit. Isolated deposition is referred to as an amyloidoma. Amyloid depositions rarely occur in the Central Nervous System. Deposits within the CNS mostly involve the walls of blood vessels or occur in senile plaques [1]. Even more rarely, they will accumulate into a tumor like deposit within the CNS [2]. There is a female predominance of CNS amyloidomas and mean age at presentation is 47.8 years [2]. This report describes a case of a primary amyloidoma of Meckles Cave, only 13 cases of such deposition have been described in the literature [4,5...13]. The most common lesions that occur within Meckles Cave are meningiomas and schwannomas [3]. However, when a patient has a lesion of this area on imaging and paresthesia, dyesthesia, or anesthesia of the trigeminal nerve, one must consider primary amyloidoma on the differential.

References

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