CASE REPORT

Bilateral Meckel’s cave amyloidoma: A case report

Amyloïdome bilatéral du cavum de Meckel : à propos d’un cas

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Summary Primary solitary amyloidoma of Meckel’s cave is rare, and a bilateral location is even more rare. To the best of our knowledge, only 12 cases in the literature have described such a primary lesion, including one case of bilateral involvement of Meckel’s cave. We report here on the case of a 57-year-old woman presenting with pseudotumor masses involving both Meckel’s caves and responsible for trigeminal neuropathy. The final diagnosis of amyloidoma was made on the basis of histological examination of surgical biopsy specimens.

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Introduction

Tumors of Meckel’s cave are uncommon, representing only around 0.5% of all intracranial neoplasms. The most frequent pathologies at this location include meningioma and schwannoma [1,2]. Amyloidoma of Meckel’s cave is rare, with just 12 documented cases of primary amyloidoma of the gasserian ganglion [1–8]. We report here only the second case of bilateral Meckel’s cave amyloidoma, and discuss the magnetic resonance imaging (MRI) findings as well as the differential diagnosis for this rare type of lesion.

Case report

A 57-year-old woman presented with a three-year history of progressive numbness and neuralgia in the territory of the second, third and, finally, first divisions of the right trigeminal nerve. Eight months prior to admission, the same progressive symptoms began on the left side of her face. She also noted dryness of the right eye and altered sensation on the right side of the tongue. Her previous medical history was unremarkable. General examination disclosed no abnormality. Neurological examination revealed hypoesthesia in all three divisions of both trigeminal nerves, with a right-sided dominance as well as right corneal hypesthesia. No other neurological abnormality was detected. Moreover, this patient had no known primary cancer.

MRI of the brain revealed extra-axial pseudotumor masses within Meckel’s caves that were filling and expanding the Meckel’s caves entirely on the right side and partially on the left. The masses were isointense with the cortex on T1-weighted images (Fig. 1), and hypointense on T2-weighted images (Fig. 2). After the administration of contrast (Fig. 3), intense and homogeneous enhancement was seen. Magnetic resonance angiography (MRA) ruled out a vascular lesion, while computed tomography (CT) of the...
head demonstrated the absence of any bony erosion of the skull base, especially at the level of the perisellar region. CT of the neck was normal. Whole-body fluorine-18-fluoro-2-deoxy-d-glucose (FDG) positron emission tomography (PET) demonstrated no abnormal uptake. Cerebrospinal fluid (CSF) analysis was normal.

Under MR-guided neuronavigation, the patient underwent a biopsy of the right-sided lesion, using an intradural subtemporal approach. The dura mater delimiting the lateral aspect of Meckel’s cave together with the Meckel’s cave contents were sampled.

The biopsy specimen consisted of fibrous tissue with large acellular and eosinophilic deposits of amorphous material (Fig. 4). The amorphous material was congophilic and showed apple-green birefringence under polarized light (Fig. 5). Amyloid deposits were resistant to potassium permanganate pre-treatment. Immunostaining for kappa and lambda light chains (Dako, Glostrup, Denmark) revealed that the amyloid deposits consisted predominantly of lambda light chains. The diagnosis of amyloidoma was made.

Biopsies of abdominal fat, rectal mucosa and bone marrow were eventually performed to rule out the possibility of a primary disease. There was no apparent sign of systemic amyloidosis, nor any sign of malignant or chronic inflammatory disease. Serum and urine immunofixation showed no monoclonal gammopathy. The skeletal survey was normal.

Six months later, the patient’s symptoms were unchanged, and the decision to undertake surgical resection of the right-sided lesion was taken. Through a subtemporal extradural approach, the double layer of dura mater covering the V2 and V3 trigeminal branches was divided, as well as the origin of the V1 branch in

Figure 1  Axial T1-weighted magnetic resonance image shows bilateral mass lesions within Meckel’s cave (arrows): the signal seen in the lesions is similar to that of the cortex.

Figure 2  Axial high-resolution T2-weighted magnetic resonance image shows that the lesions are hypointense. The left Meckel’s cave (arrow) is only partially filled by the lesion.

Figure 3  Gadolinium-enhanced (a) axial and (b) coronal T1-weighted magnetic resonance images show intense and homogeneous enhancement of the lesions, involving both Meckel’s caves (arrows), although the right side is more extensively involved in the disease.

Figure 4  The biopsy specimen is characterized by fibrous tissue with eosinophilic deposits of amorphous material and a minimal inflammatory reaction (hematoxylin and eosin stain; original magnification: ×100).
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The amyloid protein shows apple-green birefringence under light polarization of a Congo-red-stained histological section (original magnification: ×200).

The cavernous sinus, thereby allowing Meckel’s cave to be attained. The amyloidoma consisted of an avascular brownish-colored mass. The lesion was completely removed with preservation of the trigeminal nerve rootlets. The immediate postoperative course was uneventful. Trigeminal pain disappeared, and sensory deficits improved partially after two months.

Discussion

Amyloidosis is a rare disorder, characterized by extracellular deposition of an amorphous hyaline material [2,4,6–8]. It can involve almost any organ system, and may be seen as either a systemic process or a focal mass lesion—a so-called amyloidoma [1,6,8]. In the central nervous system (CNS), amyloid deposits are commonly found within cortical senile plaques and in blood vessel walls, a state known as “amyloid or congophilic angiopathy”. Amyloid deposits have also been described along peripheral nerves, in autonomic ganglia and in neurofibrillary tangles in association with dementia [1,2,4,8]. Space-occupying amyloidoma of the CNS, on the other hand, is a rare entity characterized by solitary or multiple slow-growing masses, with little or no mass effects [6,8] and mostly involving the cerebral parenchyma adjacent to either an ependymal or dural surface [8]. Other locations have included the Meckel’s cave, skull base, cerebellopontine angle, pituitary gland, temporal and orbital bone and spinal canal [1,4,6,8].

Cases of amyloidoma of Meckel’s cave have predominantly occurred in adults in their late 40s or older, with a slight female predominance [2,8]. The clinical presentation of amyloidoma is similar to that of slow-growing neoplasms [2,4,5,8], with symptom duration ranging from three to ten years. Common clinical symptoms have included trigeminal neuralgia, progressive numbness and dysesthesia [2,5,8]. The MRI aspect of Meckel’s cave amyloidoma is somewhat variable, depending on the density of protein deposition. It can appear as low, intermediate or high signal intensity areas on both T1- and T2-weighted images and are often heterogeneous. After administration of contrast, intense enhancement can be seen [1,2,6–8].

Our present case showed MRI appearances similar to those reported by Vorster et al. [1] and Yu et al. [7], with isointensity relative to the cortex on T1-weighted images and hypointensity on T2-weighted images. The differential diagnoses for hypointense T2-weighted images of Meckel’s cave are sarcoidosis and lymphoma. Lymphoma grows rapidly, may extend to the skull base and is usually responsible for erosive bony changes. Sarcoidosis can occur in the nervous system commonly involving the second, seventh and eighth cranial nerves, even in the absence of evidence of systemic disease. On the other hand, schwannoma and meningioma demonstrate relative hyperintensity on T2-weighted images [2,6,7] and, therefore, might be differentiated from amyloidosis. In our present case, an intracavernous aneurysm was easily ruled out by the absence of such typical MRI features on T2-weighted images as the flow void phenomenon or intra-aneurysmal high-signal thrombus, as well as the lack of calcification on CT. MRA was also clearly normal. In addition, a cavernous hemangioma of the cavernous sinus should have been considered on the basis of female gender (86–94% predilection) and age of our patient. Although a rare lesion, a location within the cavernous sinus has been typically reported [9,10]. Still, the hypointense appearance on T2-weighted images and the shape of the lesion allowed us to rule out this diagnosis. Moreover, our lesions were not hyperdense on CT. Metastasis were ruled out by clinical history and the absence of any associated bony lytic lesions. However, the final diagnosis of amyloidoma remains difficult to make, especially in the absence of other systemic manifestations. It usually requires biopsy for histological confirmation.

Surgical removal of amyloidoma is recommended for a proper diagnosis and pain treatment. However, in many cases, nerve deficits persist after surgery. The use of focused radiotherapy, steroids and/or colchicine has no favorable effects [4,8].

Conclusion

Although extremely rare, amyloidoma should be included in the differential diagnosis of mass lesions of Meckel’s cave, especially when the lesion shows hypointense signal intensity on T2-weighted images. The absence of associated bony lesions and a normal FDG PET scan are additional findings favoring a diagnosis of amyloidoma. The final diagnosis of this extremely rare condition is made by pathological examination of biopsy samples.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References