Unusual presentation and rare location of enterogenous cyst in the cerebellopontine angle: Two cases and review of the literature

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Abstract

Enterogenous cysts (EC) in the central nervous system are rare endodermal lesions. Here, we present two cases of EC in the cerebellopontine angle (CPA), along with a comprehensive review of the literature.

Introduction

Enterogenous cysts (EC) in the central nervous system are rare endodermal lesions. They are three times more common in the spine than in the brain. These cysts originate from aberrant embryological development of the notochord in which the notochord fails to completely separate from the endoderm. This results in a persistent abnormal connection forming the cyst. The usual location of these cysts is extra-axial, and midline anterior to the brainstem. They can present as an incidental finding or with mass effect depending on their location. The radiologic features of enterogenous cysts can mimic other cystic lesions making its diagnosis difficult.

Methods and Materials

After reviewing our hospital records within the last 10 years, we found two cases of histopathologically confirmed EC of the CPA. Their clinical, radiological, and pathological features are reviewed, along with the present literature, which revealed seventeen reported cases.

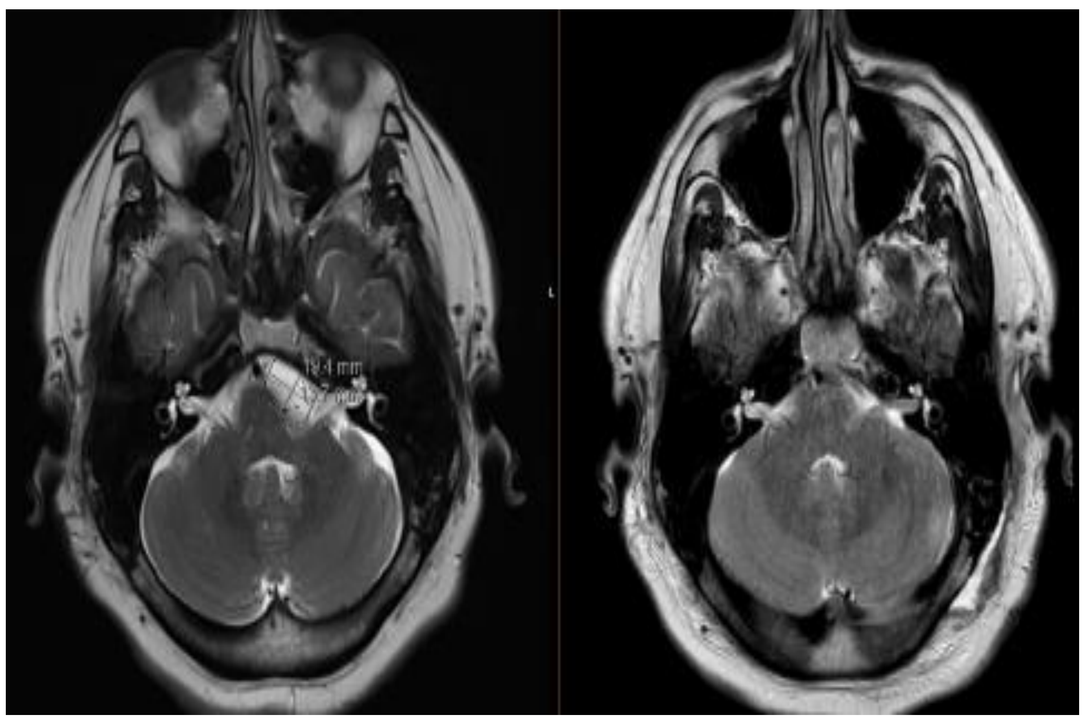

Figure 1 A. T2 axial MRI pre and post-left retrosigmoid craniotomy.

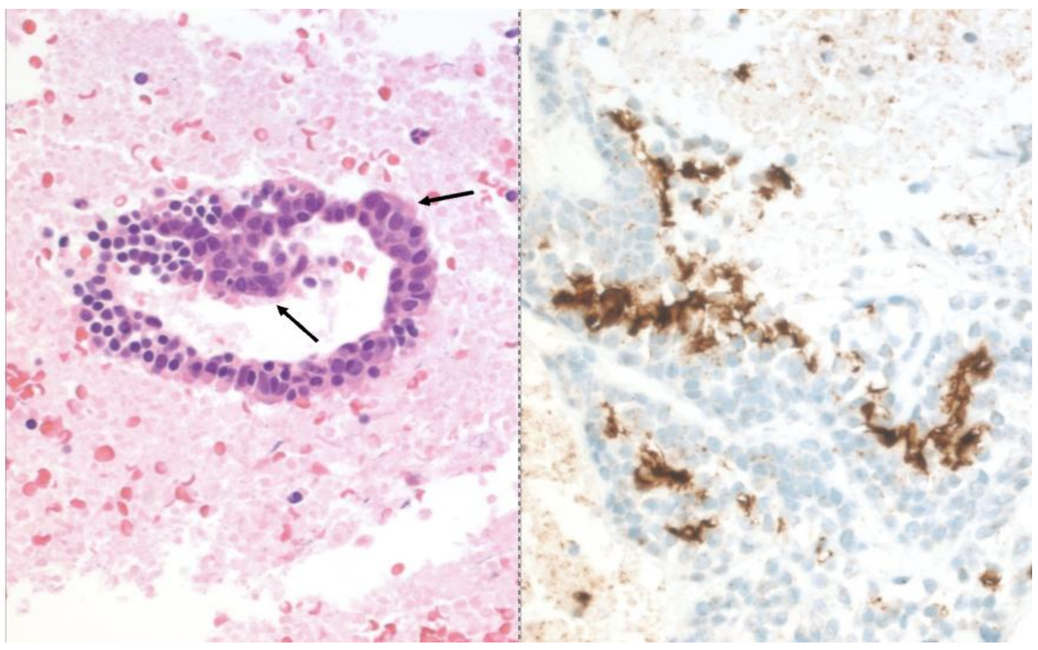

Figure 1B. Histopathology Case 1: Left: 400x, ciliated epithelium. Right: 400x, BRAF IHC showing cilia as a nonspecific staining artifact

Results

Case 1

A 54-year-old male initially presented to our neurosurgical service at age 49 for Bell's palsy from the left CPA mass presumed to be an epidermoid cyst. He was initially treated with steroids with improvement in his facial weakness. He returned to our clinic 5 years later with hemifacial spasm and numbness in the V2 distribution, as well as hearing loss. MRI showed increased size of the left CPA angle mass from 0.7cm to 1.9 cm, with compression of the Cranial nerves VI and VII. The patient underwent a left retrosigmoid craniotomy with fenestration of the cyst. Pathologic examination identified scattered groups of ciliated epithelial cells confirming EC diagnosis—figure 1 AB.

Case 2

A 38-year-old female presented with incidentally found right CPA cystic mass after a fall. The patient returned to the emergency department 2 years later with an onset of left arm numbness. She had an absent gag reflex on the physical exam. The lesion increased in size from 2.5 cm to 3.3 cm. The patient underwent a right retrosigmoid craniotomy for cyst fenestration with a preliminary diagnosis of an epidermoid cyst. Pathologic examination identified EC. — figure 2AB

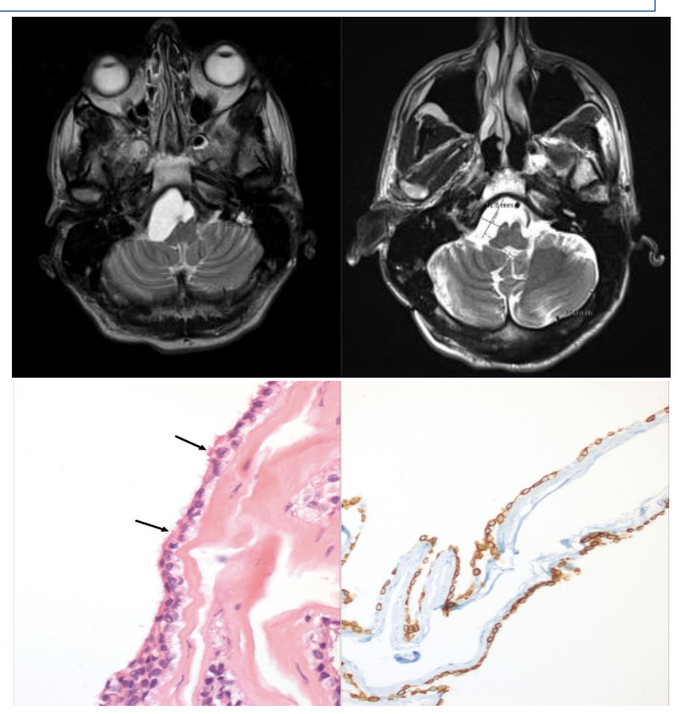

Figure 2 A. T2 axial MRI pre and post-right retrosigmoid craniotomy

Figure 2 B. Histopathology Case 2: Left: 400x, ciliated epithelium. Right: 200x, CK7

Discussion

We present two cases of rare EC at the CPA which were both mistaken for another benign lesion due to similar radiologic findings and unusual location. The differential diagnosis of cystic lesions in this location includes epidermoid cyst, arachnoid cyst, schwannoma with cystic degeneration, dermoid cyst. EC are non-enhancing lesions with T1 isointense and T2 hyperintense, no nodular component, and no restricted diffusion on magnetic resonance imaging. EC are congenital lesions derived from endodermal lining epithelium. The key histopathologic findings include ciliated or non-ciliated columnar, cuboidal or squamous epithelium, goblets cells, mucin producing cells suggesting enteric origin, typically positive for cytokeratin and carcinoembryonic antigen. A literature review found 17 reported cases; of these cases one underwent a malignant transformation. The mean age was 43.7 years (range 25-69); M: F=6/7

Conclusions

Our two cases add to the body of literature showing the importance of including these in the differential diagnosis and the aim to completely resect these tumors given the potential risk of malignant transformation.

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