

The Giant Grape-Like Enterogenous Cyst Extending from the Upper Cervical Canal to the Ambient Cistern: Case Report and Literature Review

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ABSTRACT

Objective and Importance: The authors report the first known case of a giant multiloculated grape-like enterogenous cyst extending from the upper cervical canal to the ambient cistern. **Clinical Presentation/Methods:** We report the case of a 40-year-old male who had a prior transmastoid craniotomy at an outside facility 14 months prior with an indeterminate diagnosis, who presented to the University of New Mexico with recurrent headaches and nausea. Scans demonstrated a giant multiloculated cystic lesion in the right cerebellopontine angle that extended superiorly and inferiorly with brainstem compression and hydrocephalus. **Intervention/Results:** We took the patient to the operating room for a retrosigmoid suboccipital craniectomy for tumor resection. Post-operatively, the patient improved but required ventriculoperitoneal shunting for continued communicating hydrocephalus. **Conclusion:** This is the first known case of a giant multiloculated grape-like enterogenous cyst extending simultaneously from the upper cervical canal to the ambient cistern. Enterogenous cysts should be considered on the differential diagnosis of giant grape-like lesions extending from the cervical canal to the prepontine cistern.

Keywords: Enterogenous; Giant; Cervical; Pre-Pontine; Grape

1. Objective and Importance

The authors report the first known case of a giant multiloculated grape-like enterogenous cyst extending from the upper cervical canal to the ambient cistern.

2. Clinical Presentation

We report the case of a 40-year-old male who had a prior craniotomy at an outside facility 14 months prior with an indeterminate diagnosis, who presented to the University of New Mexico with new onset headaches and nausea. Scans demonstrated a giant multiloculated cystic lesion extending from the upper cervical canal, into the cerebellopontine angle, and finally up into the ambient cistern. This mass was causing brainstem compression and obstructive hydrocephalus (**Figures 1-6**).

3. Intervention

We took the patient to the operating room for a retro-

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sigmoid suboccipital craniectomy for tumor resection. We identified at least 10 separate loculated compartments filled with xanthochromic looking fluid that we fenestrated and resected. The inferior portion attached to the upper cervical cord and medulla was not resected. Pathology demonstrated the presence of abundant mucin secretion and glandular epithelium. CAM 5.2 highlights the presence of abundant glandular epithelium. EMA was also positive highlighting the presence of fibrotic underlying leptomeninges. CDX-2 was positive in this glandular epithelium. PAS positivity highlights the presence of abundant mucin secretion and glandular epithelium (**Figures 7 and 8**). Post-operatively the patient improved neurologically but required ventriculoperitoneal shunting for continued communicating hydrocephalus and to allow for healing of his leaking wound (**Figure 9**).

4. Discussion

Enterogenous cysts, also known as endodermic or neuroepithelial cyst, are benign congenital lesions that were first described by Puusepp in 1934 [1]. They arise from

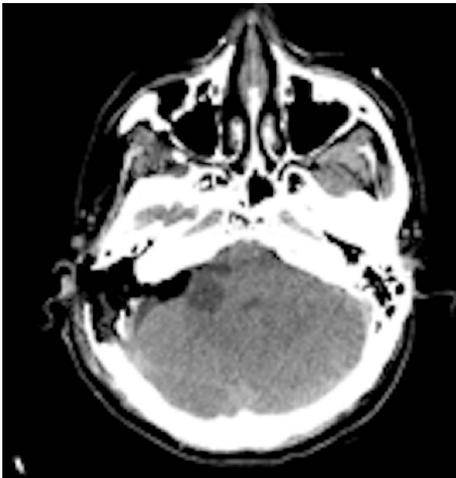


Figure 1. Post-operative CT scan after initial transmastoid surgery at outside hospital showing minimal residual cyst remaining. Diagnosis was not made from pathological reports at that time.

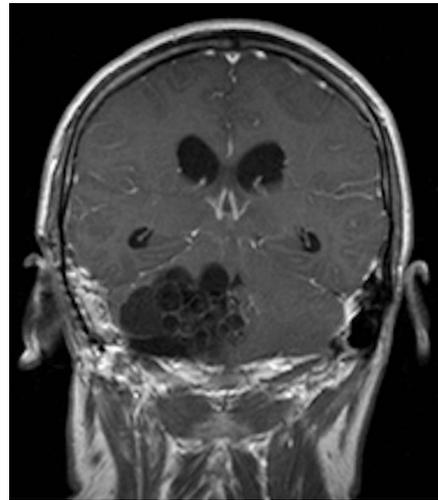


Figure 4. Coronal MRI post contrast showing this giant grape-like mass with multiple different compartments.



Figure 2. Patient presents to our facility 14 months later with recurrent symptoms and giant mass with brainstem compression and obstructive hydrocephalus.

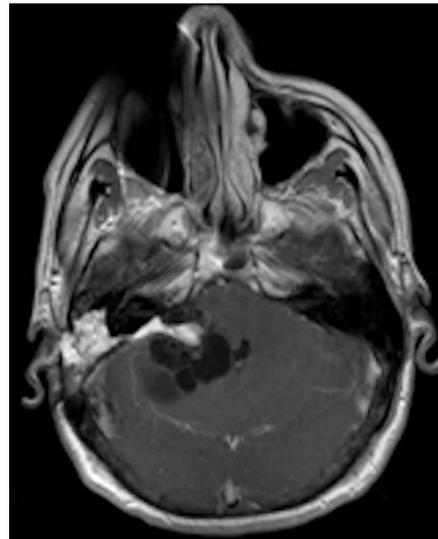


Figure 5. Axial MRI post contrast showing this giant grape-like mass.

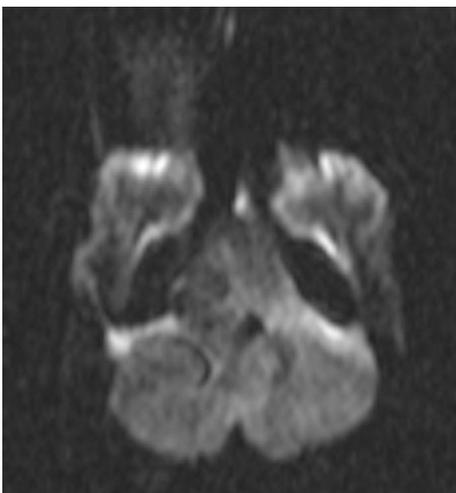


Figure 3. Diffusion MRI showing no restricted diffusion.



Figure 6. Sagittal MRI post contrast showing this giant grape-like mass.

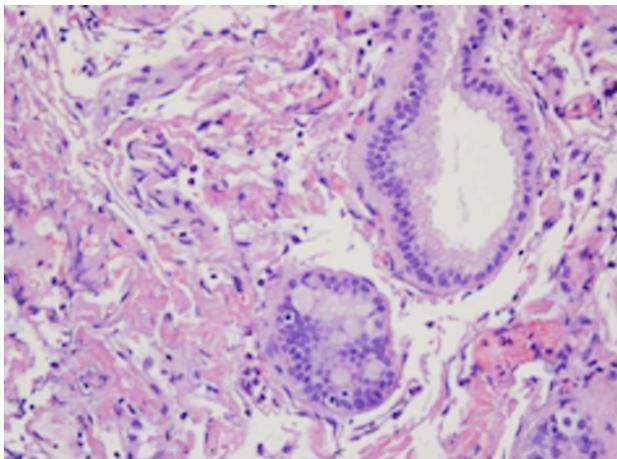


Figure 7. Histological analysis demonstrated presence of abundant mucin secretion and glandular epithelium.

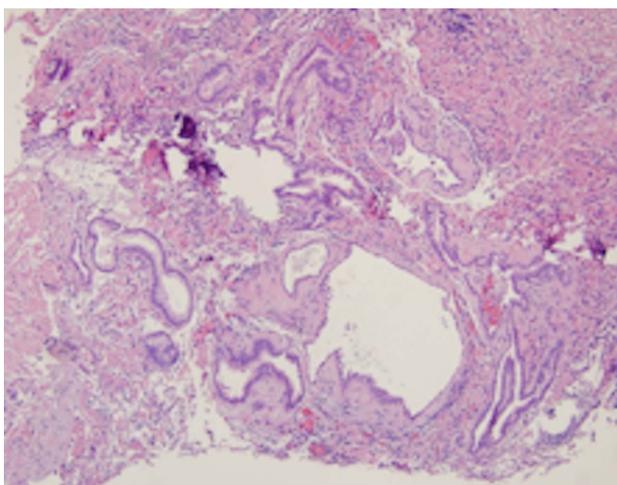


Figure 8. Histological analysis demonstrated presence of abundant mucin secretion and glandular epithelium.

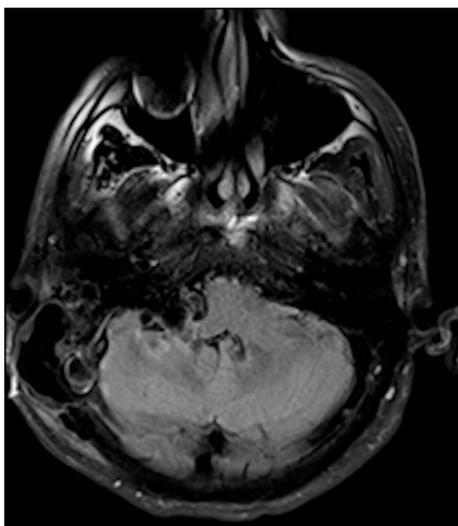


Figure 9. Post operative MRI showing complete resection and resolution of obstructive hydrocephalus.

misplaced epithelium of the nasopharynx, respiratory tree, or intestinal tract and are characterized by their simple columnar, ciliated, or goblet cell epithelium [2]. They develop during the third or fourth week of embryonic development [2]. Only about 100 of these lesions have been described in the world literature [3]. These lesions are found in all age groups, although more commonly diagnosed in young adulthood [4]. Most of these lesions have been described to occur in the lower cervical spine and thoracic spine in an intradural-extramedullary fashion [5]. Intracranial enterogenous cysts are extremely rare with most of them located in the posterior fossa. The literature notes locations of cysts are as follows: within 4th ventricle [6], cerebellopontine angle [7], ventral brainstem [8], cerebellar vermis [9], within the medulla [10], and at the foramen magnum [11]. Supratentorial cysts are exceedingly rare with only 20 cases reported worldwide [12]. No previous reports of simultaneous upper cervical spine to ambient cistern giant multiloculated lesions have been described. Additionally, this lesion was very unusual in that it had at least 10 separate loculated compartments that were distinct and separate from one another.

These cysts are known to contain clear, gelatinous, mucoid or xanthochromic fluid [13]. In 1976, Wilkins and Odon classified these lesions into three groups based on their histological features. Type 1 cysts have a simple or pseudo stratified epithelium that can be cuboidal or columnar with or without cilia. Type 2 cysts have a more complex epithelium with respiratory or gastrointestinal tissues. Type 3 cysts are similar to type 2 cysts but also have glial or ependymal cells [14].

For giant intracranial cysts, the differential diagnosis should include glial tumors, epidermoid cysts, arachnoid cysts, dermoid cysts, infectious cysts, colloid cysts, metastatic tumors as well as enterogenous cysts. Treatment of choice for these lesions is total surgical resection of the cyst wall and drainage of the cyst contents. If the wall is not resected, these lesions may recur [15]. In cases where the wall cannot be completely excised due to difficulty separating arachnoid pial plane, partial resection is recommended with connection of the loculations to the subarachnoid space [16]. When partial resection is performed, symptomatic recurrence is documented at 31 % at 2 months to 14 years [17].

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