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Giant Supratentorial Acutely Hemorrhagic Enterogenous Cyst: Case Report and Literature Review

Paul E. Kaloostian*, Han Chen, Franklin Westhout, Howard Yonas
University of New Mexico Neurosurgery, Albuquerque, New Mexico, USA
Email: *pkaloos1@jhmi.edu

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ABSTRACT

The authors report the first case of a giant supratentorial enterogenous cyst presenting with acute symptomatic hemorrhage within the cyst. We report the case of a 78-year-old Philipino female who was found to have a large right fronto-parietal mass after a minor fall. She had a small amount of hemorrhage within the cyst but was stable for discharge the following day. She was readmitted 5 days later with acute onset severe headache and left-side weakness. On repeat imaging, her cyst had grown in size and had large acute hemorrhage within it. She was taken to the operating room for craniotomy and cyst resection. She recovered well post-operatively. This is the first known case of a giant supratentorial enterogenous cyst presenting with symptomatic enlargement due to large hemorrhage within the cyst. Enterogenous cysts should be considered on the differential diagnosis of hemorrhagic supratentorial giant cysts.

KEYWORDS

Hemorrhagic; Enterogenous; Cyst; Giant; Supratentorial

1. Introduction

The authors report the first case of a giant supratentorial enterogenous cyst presenting with expanding acute hemorrhage within the cyst.

2. Case Presentation

We report the case of a 78-year-old Philipino female who was found to have a large 6 × 3 × 7 cm right fronto-parietal mass after a minor fall (**Figures 1** and **2**). She was found to have a small amount of hemorrhage within the cyst and was then discharged home. She was readmitted 5 days later with severe headache, altered mental status, and left-sided weakness.

3. Intervention

On repeat imaging, her cyst had grown in size and had acute hemorrhage within it (**Figure 3**). She was taken to the operating room for craniotomy and cyst resection (**Figure 4**). Intra-operatively, this mass was full of xanthochromic fluid with a large amount of acute clot located posteriorly. No mural nodule was identified. The cyst was drained and the lining was removed until we identified normal looking white matter at all angles. She suffered some minor cardiac insults post-operatively but her neurological status recovered back to baseline. Pathology demonstrated multiple stretches of cyst lining which are largely composed of cuboidal cells, overlying a continuous basement membrane. No cilia or goblet cells were noted. Cytokeratin CAM 5.2 was positive, with GFAP negative and S-100 negative. CAMP 5.2 was positive and PAS was positive (**Figure 5**).

4. Discussion

Enterogenous cysts are extremely rare, comprising only 0.01% of all central nervous system tumors [1]. Only about 100 known cases have been described in the world literature [2]. They are noted to originate during the third or fourth week of embryonic development. Several names exist for these lesions such as neuroenteric cysts, enteric

*Corresponding author.

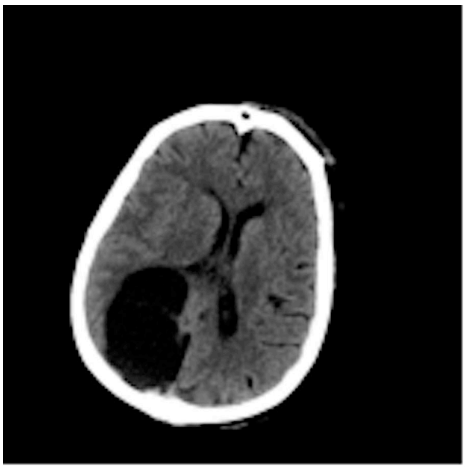

Figure 1. CT head showing giant right fronto-parietal enterogenic cyst with mass effect on the occipital horn.

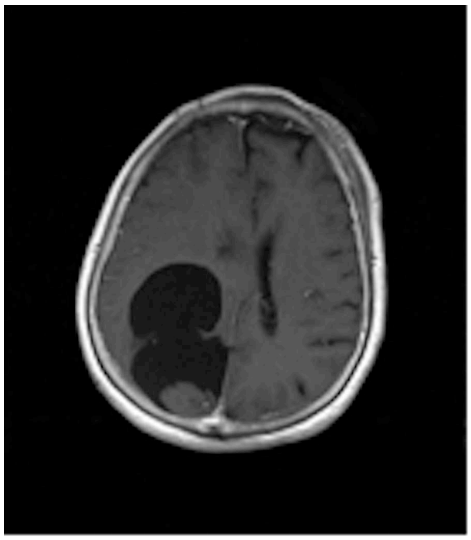

Figure 2. MRI brain showing giant right fronto-parietal enterogenic cyst with mass effect on the occipital horn.

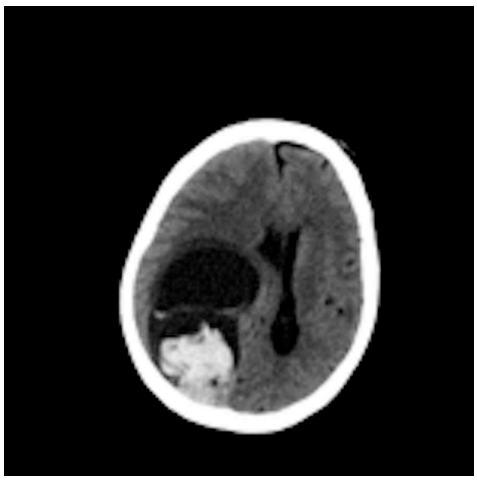

Figure 3. CT head showing spontaneous hemorrhage within the giant cyst with evidence of midline shift.

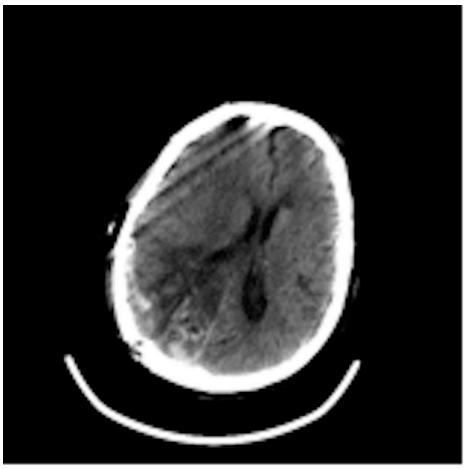

Figure 4. Post-operative CT head showing complete resolution of the mass effect and resection of the cyst.

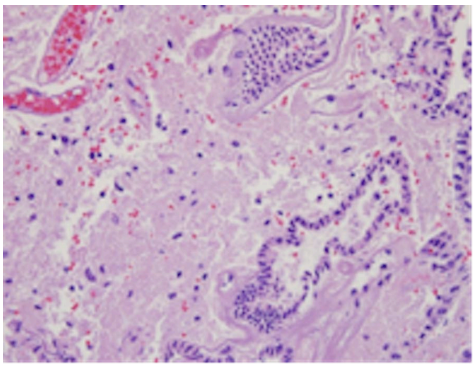

Figure 5. High power view hematoxylin and eosin staining demonstrating multiple stretches of cyst lining, which are largely composed of cuboidal cells, overlying a continuous basement membrane, a feature of Type I enterogenous cysts epithelial. No cilia or goblet cells were seen.

cysts, endodermic cysts, gastrogenic cysts, and bronchogenic cysts. Various theories exist to explain their pathogenesis. Perhaps the most accepted is that the cysts located between the diencephalon and mesencephalon revive from remnants of the Seesel's Pouch. This diverticulum arises caudal to Rathke's pouch and rostrodorsal to oropharyngeal membrane [3]. These lesions are documented in all age groups, though most commonly in children and young adults [4]. These cysts are most commonly located in the lower cervical and upper thoracic segments of the spinal cord, and tend to occupy the region anterior to the spinal cord or brainstem [5]. There are four documented cases of malignant transformation of these cysts [6].

The intracranial location is quite rare. The first reports of intracranial cysts were by Small in 1962 and Giombini in 1981 [7,8]. Reports of intracranial enterogenous cysts note the midline posterior fossa region anterior to the brainstem to be the most common site, as well as the fourth ventricular region [9]. Supratentorial enterogenous cysts are exceedingly rare, with only 20 described cases

[10]. These patients are usually older than patients with infratentorial cysts and much is not known about their natural history due to the paucity of cases worldwide. Giant supratentorial enterogenous cysts have only been described in 2 cases thus far [11]. Their natural history has yet to be documented. No reports of spontaneous hemorrhage within the cyst have been documented thus far.

On pathology, these cyst walls show immunoreactivity for EMA, CEA, and cytokeratin with absent reactivity for glioneuronal markers, and are lined typically by columnar mucin-rich epithelium resembling enteric or respiratory epithelium [12].

Treatment of choice for these lesions is total surgical resection of the contents and cyst wall, so as to prevent reaccumulation. However, if there is extensive scarring and adherence to the pia mater, it is recommended to leave the inner wall behind to avoid injuring the cortex [13]. Prevention of spillage of the contents of the cyst is critical to preventing dissemination [13].

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