Cholesterol granuloma of the lateral ventricle

Case report

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Cholesterol granulomas (CGs) are benign lesions resulting from an inflammatory reaction to cholesterol and hemosiderin. These masses most often arise within the temporal bone or nasal sinuses; intracerebral CGs are extremely rare. In this report the authors present an unusual case of a CG arising within the lateral ventricle.

The patient presented with transient hemiparesis and numbness. Computed tomography and magnetic resonance imaging demonstrated a cystic partially enhancing midline mass within the right lateral ventricle, expanding the ventricle and displacing the septum pellucidum. The patient underwent an interhemispheric, transcallosal resection of the lesion. Microscopic examination revealed a granulomatous inflammatory lesion containing cholesterol clefts, macrophages, and hemosiderin. Embedded within the granulomatous response were foci of tiny cystlike structures lined by nonciliated flattened cuboidal epithelium, consistent with the diagnosis of CG.

To the authors’ knowledge this is the first reported case of CG presenting as an intraventricular mass. The origin of this lesion is unclear, but it may relate to prior traumatic brain injury. The authors describe the presentation, imaging findings, histopathological characteristics, and surgical treatment of this rare lesion and related pathological entities.

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Key Words • brain neoplasm • cerebral ventricle neoplasm • cholesterol granuloma • lateral ventricle

Cholesterol granulomas are benign lesions resulting from a robust inflammatory response to cholesterol and hemosiderin. Intracranially, these lesions most often arise within the pneumatized temporal bone or at the petrous apex, and they can extend posteriorly into the cerebellopontine angle. These lesions are usually associated with pathological conditions of the inner ear including infection, trauma, and hemorrhage and are thought to be caused by insufficient drainage of the mastoid air cells. Intracerebral CGs are extraordinarily rare.

In this report we describe the case of a 52-year-old man referred for treatment of an intraventricular mass that was found to be a CG. We also discuss surgical treatment of this unusual entity, as well as possible causes of a large, aggressive granulomatous reaction within the lateral ventricle.

Case Report

History and Examination. This 52-year-old African-American man with a history of diabetes, hypertension, hyperlipidemia, and chronic renal insufficiency presented to an outside hospital with an acute episode of transient hemiparesis and numbness. Computed tomography scanning demonstrated a midline intraventricular mass in the region of the septum pellucidum (Figs. 1 and 2). The patient’s symptoms resolved spontaneously and, on referral to our center, he did not exhibit any neurological deficits. Of note, the patient sustained a significant closed head injury 4 years prior to this presentation; however, no imaging of the patient’s brain was performed at that, or any other time, before this presentation.

Preoperative imaging confirmed a 2.5 × 3.3 × 2.5-cm midline mass that expanded the bodies of the lateral ventricles and depressed the roof of the third ventricle (Figs. 1 and 2). The lesion was isodense on CT scans and appeared to be located within the right lateral ventricle, displacing

Abbreviations used in this paper: CG = cholesterol granuloma; CPXG = choroid plexus xanthogranuloma; CT = computed tomography.
the septum pellucidum medially. The signal characteristics of the mass suggested a proteinaceous cystic lesion. Magnetic resonance imaging demonstrated a hyperintense mass with heterogeneous signal characteristic of proteinaceous material, calcium, or cholesterol. The posterolateral aspect of the lesion enhanced with contrast; otherwise, there was only a rim of enhancement that represented displaced vascular structures. There was no sharp delineation on the posterior margin, suggesting that the lesion had possibly arisen from the medial thalamus. There was no associated hydrocephalus. Preoperative considerations for a lesion in this location included a germ cell tumor, intraventricular meningioma, central neurocytoma, and an intraventricular oligodendroglioma; however, in this case, the imaging findings favored an intraventricular craniopharyngioma.

Operation and Postoperative Course. The patient underwent a bifrontal craniotomy with an interhemispheric transcallosal approach. Upon opening the lateral ventricle, the mass, which had the external appearance of a colloid cyst, was opened and the contents along with a large portion of the cyst wall were sent for histological examination. The mass was completely resected except for a small rim on the posterior edge of the lesion that was densely adherent to the thalamus and choroid plexus. Postoperatively, the patient was awake and alert and was without focal neurological deficits.

Histological Examination. Microscopic examination of the pathological specimen revealed numerous cholesterol clefts, macrophages, chronic inflammation, and hemosiderin pigment. Embedded within the granulomatous response were several foci of tiny cystlike structures lined by nonciliated, low columnar, and flattened cuboidal epithelium (Fig. 3). No teratomatous component was seen. No keratin or skin adnexa was seen. The cyst epithelium was immunopositive for the epithelial marker CAM5.2 and immunonegative for glial fibrillary acidic protein, S100 protein, vimentin, and synaptophysin. The epithelium also stained positively for PAS and was resistant to digestion with diastase. The adjacent brain tissue demonstrated reactive gliosis, numerous Rosenthal fibers, and vascular calcifications.

Discussion

Intracranial CGs are rare lesions that typically arise within the temporal bone. These benign lesions are thought to occur following trauma or infection, causing hemorrhage in the pneumatized spaces of the temporal bone or sinuses and resulting in an inflammatory reaction to trapped hemosiderin and cholesterol. The inflammatory reaction to cholesterol crystals caused by erythrocyte and hemoglobin breakdown coupled with insufficient drainage then results in further hemorrhage, inflammation, and granuloma formation.2 Cholesterol granulomas are slow growing and can erode through bone into the surrounding areas where they can become symptomatic because of compression of the adjacent neurological structures. For example, typical petrous CGs can expand into the cerebellopontine angle causing facial weakness and lower cranial nerve dysfunction.8 Cholesterol granulomas not originating in the temporal bone or sinuses are extraordinarily rare.

On histopathological examination, the lesion in this case displayed several features consistent with CG including the typical inflammatory granuloma encompassing cholesterol clefts and hemosiderin-laden macrophages. Embedded within the lesion were several small foci of epithelium that were negative for glial fibrillary acidic protein, S100 protein, vimentin, and synaptophysin, suggesting that the epithelium was neither of ependymal nor choroid plexus origin. Additionally, the epithelium did not exhibit the typical stratified squamous epithelium with peripheral nuclear palisading classically seen with craniopharyngiomas.6 Another possibility is that this lesion may represent a CPXG, a related pathological entity that is an inflammatory lesion consisting of cholesterol crystals and fibrosis. These lesions are found within the choroid plexus of the lateral ventricles in 1–7% of autopsy studies but are rarely symptomatic.4 Additionally, CPXGs are almost exclusively confined within the choroid plexus, and demonstration of typical choroid plexus epithelium surrounding the lesion establishes the diagnosis.1,4,9 Histopathologically, CGs and CPXGs are quite similar, consisting of a granulomatous inflammatory response to cholesterol and hemosiderin.10 Mu-
enchau and Laas\textsuperscript{5} have suggested a similar origin of CGs and CPXGs. Given that the epithelium seen embedded within this lesion was negative for S100 and synaptophysin, it is not consistent with choroid plexus; therefore, it is unlikely that this lesion is a CPXG. Although it is possible that this lesion does represent a large CPXG that has overgrown the choroid plexus and expanded into the ventricle, this would also be very unusual, as CPXGs are almost universally confined within the choroid plexus.

Finally, the epithelium may represent a small residual component of a benign neurenteric cyst that ruptured into the ventricle resulting in a florid granulomatous response; however, the lesion displayed none of the classic features of a neurenteric cyst. We therefore believe that the most likely diagnosis is a typical CG appearing in a very atypical location.

To our knowledge, this is the first reported case of a lesion with a dominant CG-like appearance presenting in the lateral ventricle. The origin of this lesion remains unclear; however, several possibilities exist. As noted earlier, typical petrous apex CGs are thought to result from hemorrhage or infection trapped within the pneumatized air spaces of the temporal bone. Perhaps the lesion described in this report is related to an intraventricular or intraparenchymal hemorrhage that occurred as a result of the head trauma sustained by the patient 4 years prior to this presen-

Fig. 2. A: Unenhanced CT scan demonstrating an isodense midline mass in the body of the right lateral ventricle displacing the septum pellucidum to the left. B: Axial T2-weighted spin echo MR image demonstrating a cystic hyperintense lesion within the right lateral ventricle with a dark rim representing a fibrous capsule. C: Axial T1-weighted MR image without contrast demonstrating the hyperintense mass with an area of low signal intensity representing calcification. D: Axial Gd-enhanced T1-weighted MR image demonstrating no significant enhancement of the central cystic mass, with mild enhancement of the posterior solid portion, and an enhancing rim of displaced veins.
This lesion may then represent an inflammatory response to cholesterol crystals and hemosiderin deposited at the time of hemorrhage.

Conclusions

This report describes a large intraventricular lesion which, based on histopathological evaluation, was found to be a granulomatous inflammatory reaction to cholesterol crystals and hemosiderin most consistent with a diagnosis of CG. This is the first report of a CG arising within the lateral ventricle.

References


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