Hemorrhagic Familial Colloid Cyst

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No disclosures

BACKGROUND

- Colloid cysts are slow-growing benign lesions located in the third ventricle.
- Estimated prevalence of 1 in 5800.[1]
- They have been associated with a variety of symptoms and neurological deficits, most common being headache.
- Rarely, colloid cysts have also been reported to cause sudden death secondary to acute obstructive hydrocephalus.
- Hemorrhagic colloid cysts are rare. Of the few reported cases in the literature, the majority have been diagnosed on histopathological examination following resection with a minority showing convincing pre-operative imaging findings of hemorrhage.[1]
- A minority of cases are familial.[2]

CLINICAL PRESENTATION

A 55-year-old male with a known colloid cyst status post ventriculoperitoneal shunting at age 13 years with additional ventriculoperitoneal shunt placements at age 32 years presented to the ED with 5 days of right frontal headache.

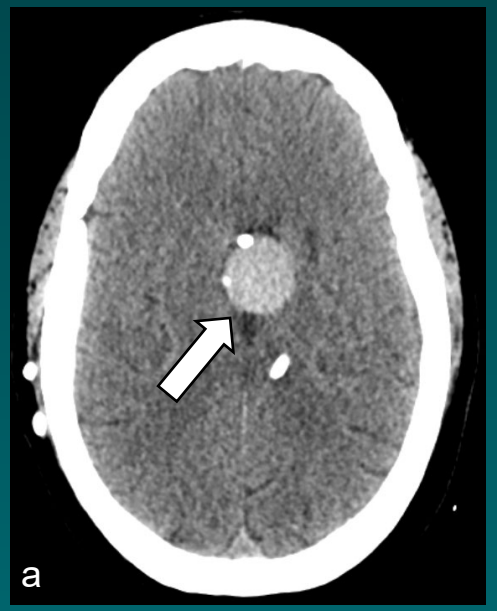
<u>Family History:</u> Patient's 51-year-old brother presented at age 47 years after minor trauma. He had bifrontal headaches attributed to rhinosinusitis. Non-contrast head CT revealed a 1.9 cm hyperattenuating mass at the third ventricle causing obstructive hydrocephalus. He underwent transcallosal resection, and pathology confirmed a colloid cyst.

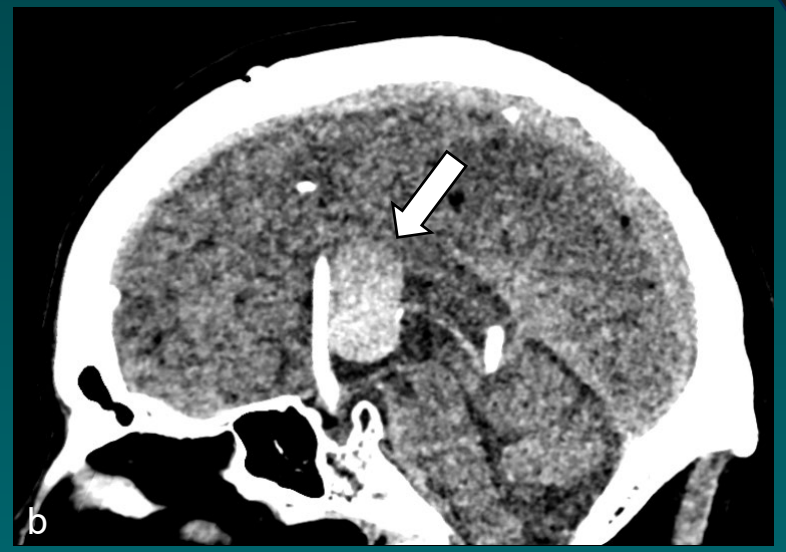
Neurological examination was non-focal.

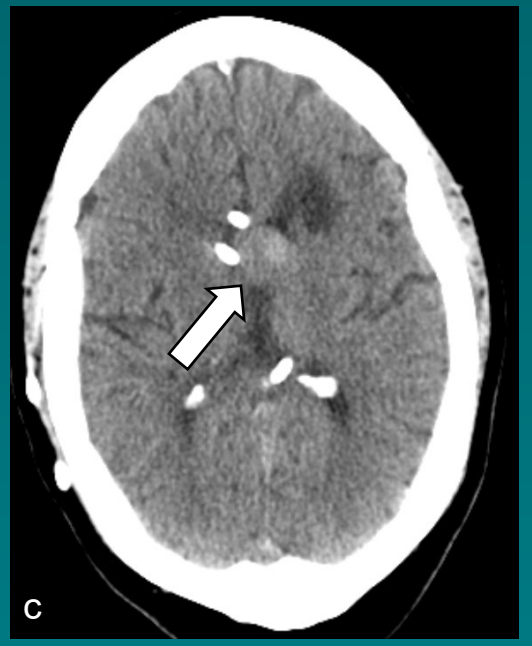
Non-contrast head CT was ordered.

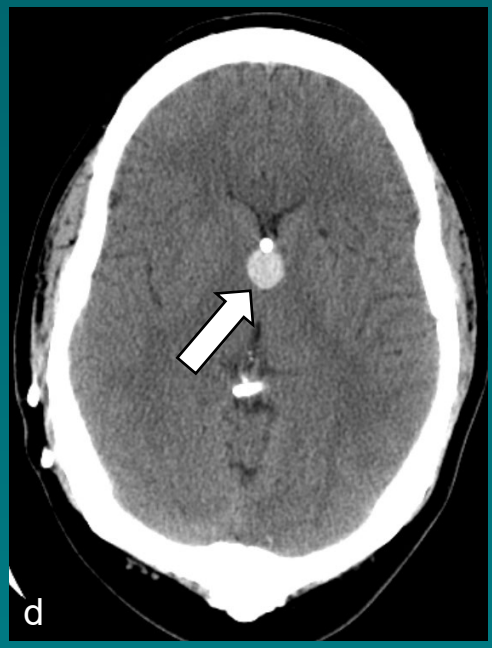
IMAGING

Non-contrast head CTs. **(a)** Axial and **(b)** sagittal images at presentation show a well-marginated hyperattenuating lesion (arrows) centered at the anterosuperior third ventricle measuring up to 2.8 cm AP, multiple shunt catheters, and decompressed ventricles. **(c)** Axial image from 1 year prior showed a less hyperattenuating lesion (arrow) measuring up to 2.1 cm AP. **(d)** Axial image from 22 years prior showed the lesion (arrow) measured up to 1.4 cm AP.



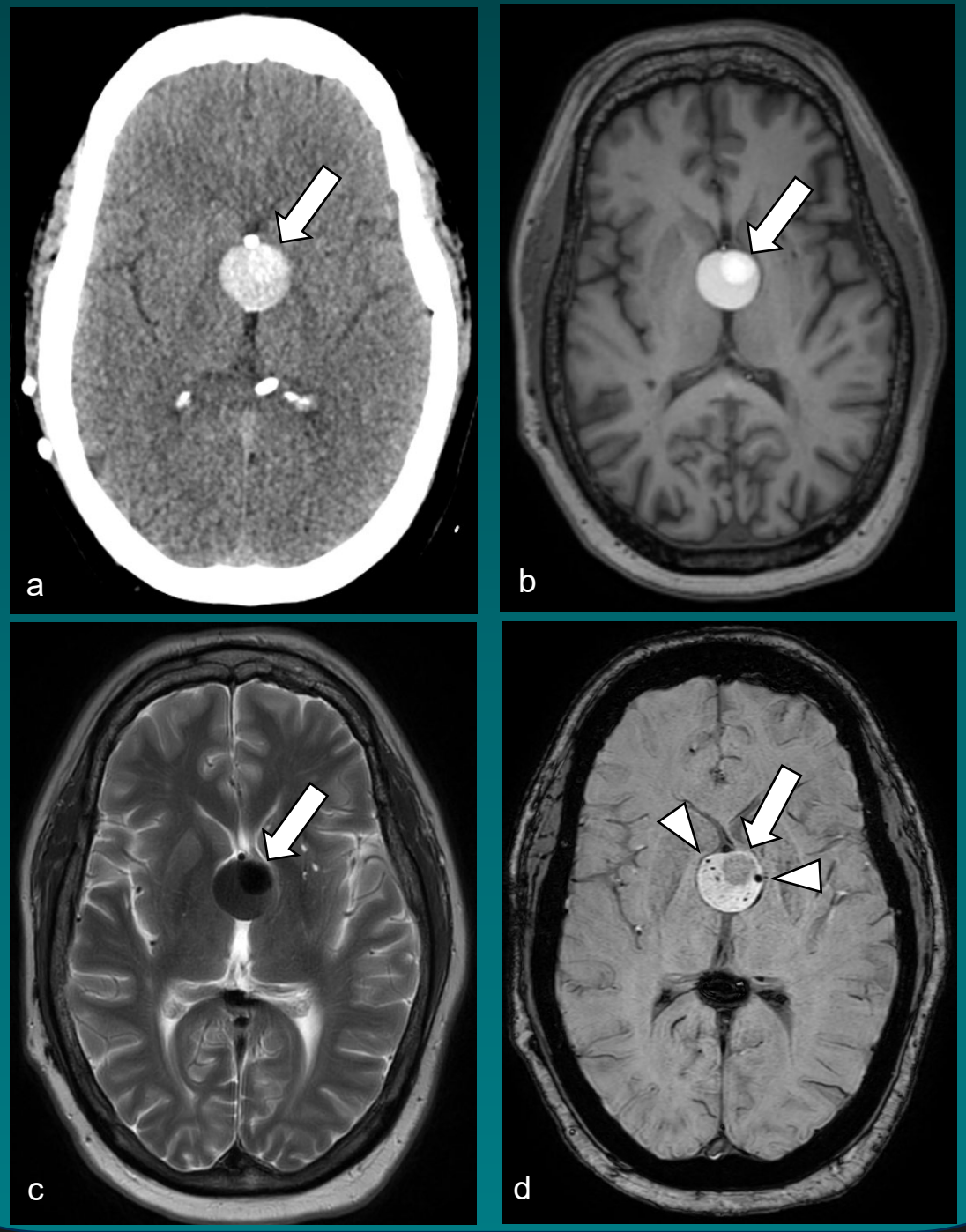






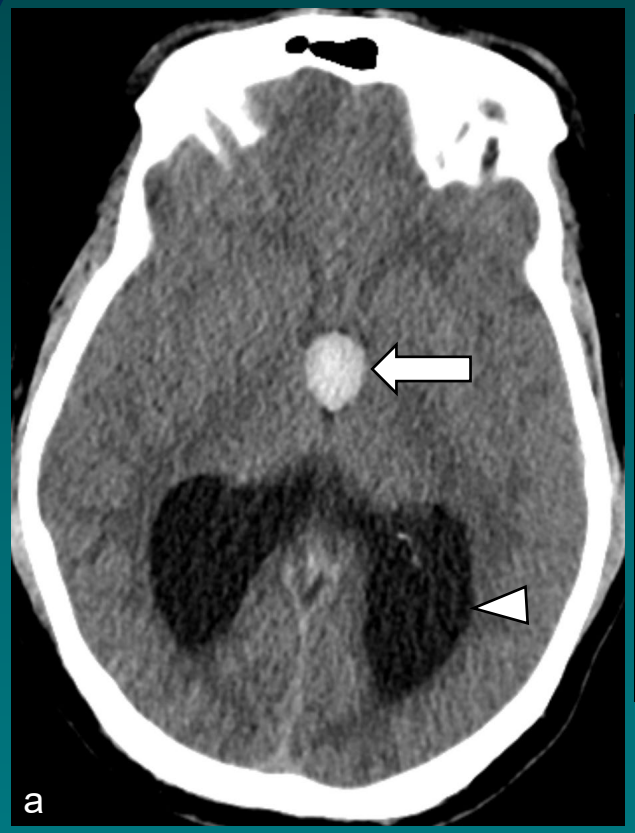
IMAGING

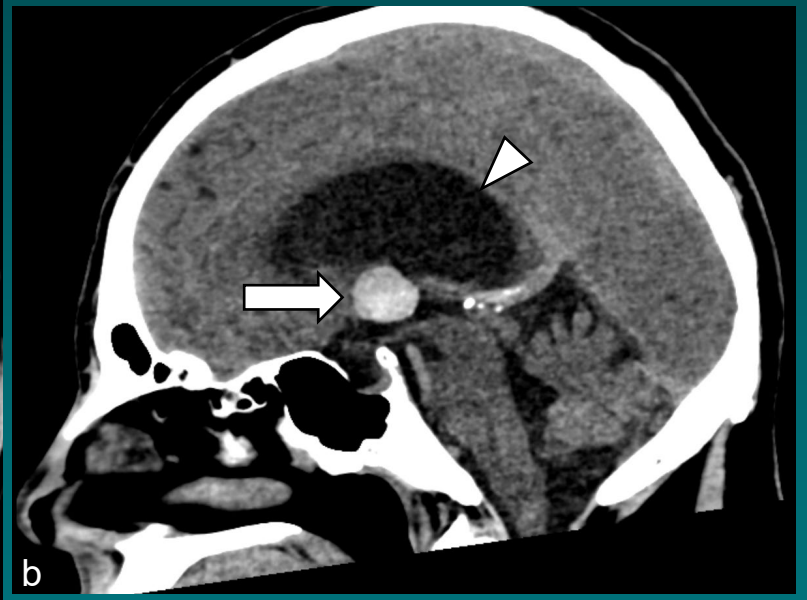
(a) Axial non-contrast head CT shows a faint round focus of intralesional increased attenuation (arrow) relative to the rest of the lesion. **(b)** Axial T1-weighted image shows lesion hyperintensity, greater in the round focus (arrow). **(c)** Axial T2-weighted image demonstrates lesion hypointensity, greater in the round focus (arrow), which is compatible with the dot sign and should not be confused with hemorrhage. **(d)** Axial susceptibility-weighted image shows multiple intralesional punctate hypointensities (arrowheads), representing blood products. Correlation with CT confirms that they are not dense enough to be calcifications. The dot sign (arrow) is not hypointense enough to represent hemorrhage.



IMAGING

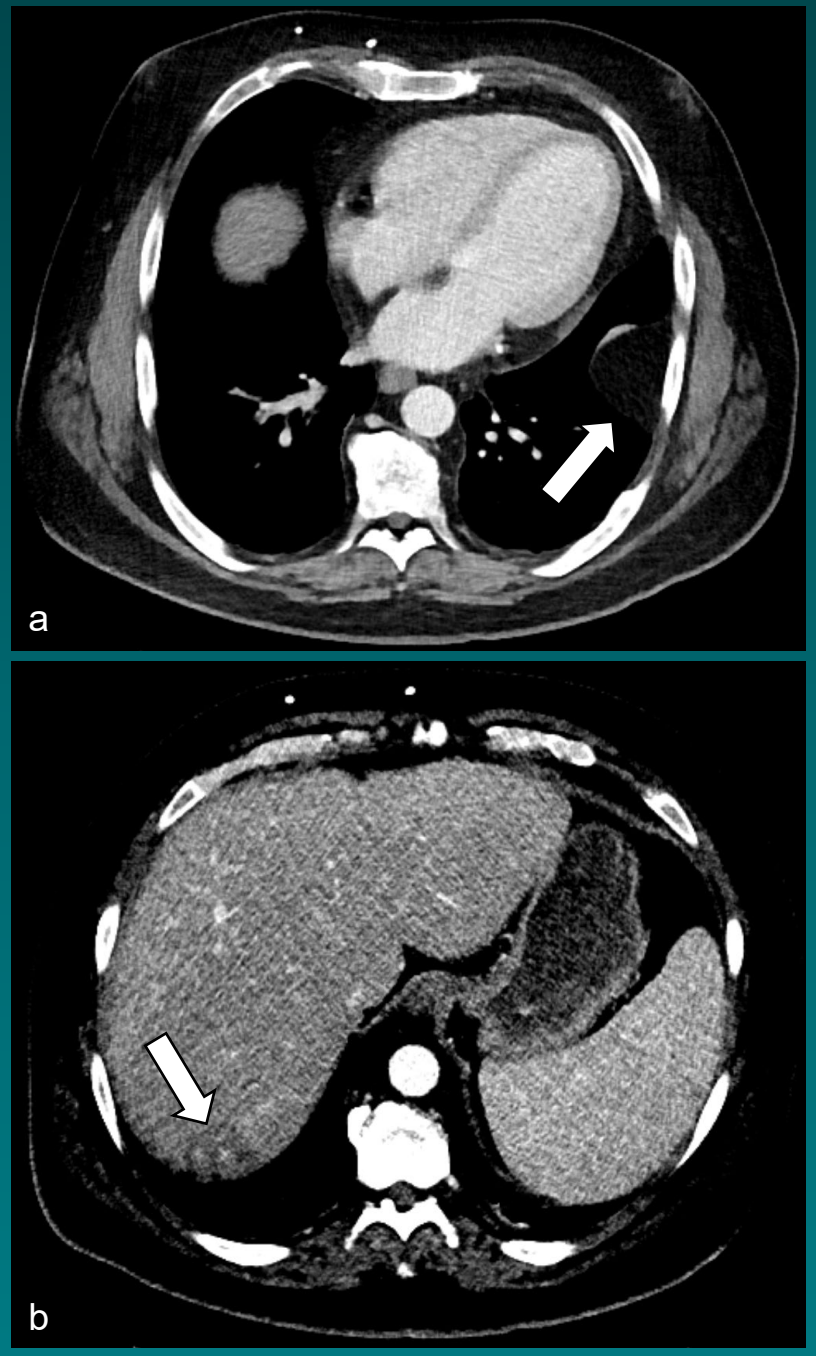
Non-contrast head CT of patient's brother. **(a)** Axial and **(b)** sagittal images show a hyperattenuating lesion (arrows) at the anterosuperior third ventricle with obstructive hydrocephalus (arrowheads). Surgical resection confirmed colloid cyst. Hemorrhage was not identified on imaging or histology.





IMAGING

Patient's chest CT with contrast (performed for unrelated reasons). **(a)** Axial image through the lung bases reveals a 6 cm left pleural lipoma (arrow). **(b)** Axial image through the upper abdomen shows a probable hepatic "hemangioma"/venous malformation (arrow).



MANAGEMENT

- After neurosurgical evaluation determined that no acute intervention was needed, patient was discharged.
- Headaches were treated with acetaminophen and resolved after a few days.
- At follow-up in neurosurgery clinic, colloid cyst resection was recommended due to its growth.
- However, given headache resolution, patient opted for surveillance imaging instead.

DISCUSSION

- Colloid cysts have variable MR signal characteristics.
  - Retrospective review of 64 patients with colloid cysts seen on MR[3]
    - 50% T1 hyperintense to gray matter, 50% iso- or hypointense
    - 53% T2 hyperintense to gray matter, 47% hypointense

- Dot and black hole signs
  - Dot sign - a discrete T2 hypointense intracyst nodule,[4] as seen in our patient
  - Black hole sign - describes a dot sign with T2 hypointensity at least 50% of the cyst volume, has a T2 hyperintense rim[3]
  - Histopathologically, these represent viscous liquid motor oil-like amorphous colloid material or solid amorphous colloid material, but not hemorrhage[5,6]

- Hemorrhage/blood products
  - Strongly hypointense on susceptibility weighted imaging (SWI), as seen in our patient
  - While cases documenting SWI hypointense hemorrhage have been reported,[1] they are rare; in the retrospective review, none of the 64 cysts had SWI hypointensity.[3]
  - Acute hemorrhage has been suggested as a cause of cyst enlargement, which may have occurred in our patient, and is thought to be a cause of acute obstructive hydrocephalus.[7]

DISCUSSION

- Familial colloid cysts
  - In a retrospective analysis, 13 of 383 (3.4%) of patients with radiologically diagnosed colloid cysts had a family member with a colloid cyst.[2]
    - 9 occurred in mother and child (69.2%), 3 in siblings (23.1%), and 1 in father and child (7.7%)
  - Survey data has suggested a rate of 12%.[8]

- A gene responsible for familial colloid cyst has not yet been identified.
  - Possibly related to nasal and suprasellar dermoid cysts[9]
  - Unclear whether our patient's pleural lipoma and hepatic "hemangioma" are related to an underlying genetic mechanism associated with colloid cyst formation

- MRI screening
  - Recommended for first-degree relatives when two or more family members have colloid cysts[9]

TAKE-HOME POINTS

- Hemorrhage
  - Hemorrhage in a colloid cyst is rare and should not be confused with the “dot sign” - a T2 hypointense nodule representing viscous or solid colloid material
  - SWI should be used to document hemorrhage
  - Hemorrhage is believed to be a cause of acute cyst enlargement and acute obstructive hydrocephalus
- Familial
  - ~3-12% of cases
  - No causative gene has been identified
  - If two or more family members have colloid cysts, first-degree relatives should be screened with MRI

QUESTIONS OR COMMENTS?

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