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WHAT'S YOUR DIAGNOSIS

Answer: An incidental large arachnoid cyst associated with compensated hydrocephalus in a patient with hemophilia A

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MRI of the brain was obtained to better define the cystic lesion and adjacent structures impacted by its mass effects. Additional 3-dimension FIESTA sequences (fast imaging employing steady-state acquisition) were employed to obtain strong hyperintensity signals of fluid-filled tissues (Fig. 3A). These tests revealed a large lobulated sellar and suprasellar cystic lesion, most likely an arachnoid cyst. This finding was associated with mass effects on the third ventricle, foramina of Monro, optic chiasm and optic tracts (Fig. 3A and B). Compression of the pituitary infundibulum with expansion of the sella turcica and associated lateral ventriculomegaly (Fig. 3C) were also noted. There was no neuro-radiologic evidence of intracranial hypertension; no trans-ependymal oedema, as manifested by a periventricular core of interstitial cerebral T2 signal hyperintensity, was noted. Echogradiant MRI sequences revealed no cystic or intracranial hemorrhages. Spontaneous resolution of the patient's symptoms, absence of papilledema and absence of signs of intracranial hypertension prompted the neurosurgical service to elect observation follow-up clinical examinations and interval neuro-radiology assessments. Surgical intervention will be considered with the development of new focal neurologic symptoms and signs or interval progression of optic atrophy with corroborative neuro-radiologic features.

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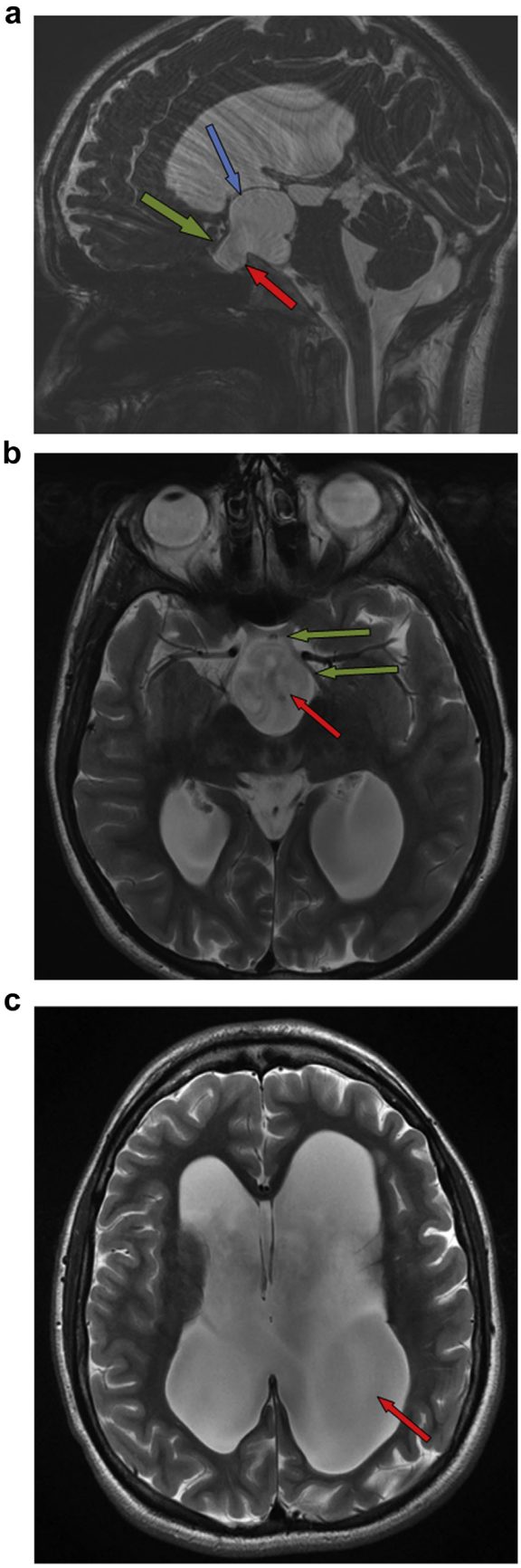

Figure 3 **(A)**: 3D Fiesta T2-weighted sagittal MRI of brain: Large sellar and suprasellar lobulated expansive cyst compressing the floor of the third ventricle (blue arrow). There is anterior displacement with mild mass effects on the optic chiasm and pituitary infundibulum (green arrow) and expansion of the sella turcica (red arrow). **(B)**: Axial T2-weighted MRI of brain: Supra-sellar arachnoid cyst (red arrow) compressing optic chiasm and optic tracts (green arrows). **(C)**: Axial T2-weighted MRI of brain: bilateral marked lateral ventriculomegaly secondary to arachnoid cyst obstruction of foramen of Munro. No trans-ependymal CSFpermeation suggests compensated hydrocephalus.

Discussion

Hydrocephalus associated with hemophilia is expected to occur in the presence of an intracranial hemorrhage. Either a non-communicating hydrocephalus caused by an obstruction of CSF circulation or the communicating form through disruption of CSF resorption by arachnoid granulations may occur. Although rare in hemophilia, an intracranial hemorrhage may arise spontaneously or from minor injuries [2]. Arachnoid cysts are anomalous dilatations of the arachnoid mater, which contain CSF-like fluid, but do not communicate with the ventricular system. The majority are small, benign and incidental neuro-radiologic discoveries. Most are found in the middle cranial fossa and are typically asymptomatic [3—5]. Progressive growth or an intracystic hemorrhage, which often occur after trauma [6], may cause hydrocephalus and neurologic deficits. Here, we present a rare, incidental finding of significant obstructive hydrocephalus caused by a large arachnoid cyst without intracranial or an intracystic hemorrhage in a patient with hemophilia A.

Although often sporadic, arachnoid cysts may arise secondary to infection, inflammation, neoplasm or intracranial hemorrhage [4]. An antecedent, asymptomatic intracranial hemorrhage in this patient who had hemophilia A, may have resulted in the development of this arachnoid cyst. However, without a history or neuro-radiologic evidence of prior hemorrhages, it seems more likely that this is an incidental congenital developmental defect. Gradual enlargement of the lobulated cyst resulted in the distortion and cephalad displacement of the third ventricle, obstruction of foramina of Munro, progressive disruption of CSF circulation and absorption with resultant accumulation of CSF fluid and expansion of lateral ventricles. Acute uncompensated hydrocephalus is usually associated with papilledema, intraventricular hypertension and trans-ependymal interstitial cerebral oedema. However, the absence of papilledema and segmental optic nerve atrophy manifested by temporal optic nerve pallor and RNFL thinning [1] is evidence of a chronic mechanism with a slow gradual compression of the anterior visual pathway (Fig. 2). Further absence of trans-ependymal oedema in T2 weighted images (Fig. 3C) is evidence that this hydrocephalus is chronic and compensated without associated intracranial hypertension. This case demonstrates the rare occurrence of an incidental large lobulated sellar and suprasellar arachnoid cyst causing chronic compensated hydrocephalus and mild segmental optic atrophy in a patient with hemophilia A.

Author contributions

The authors are aware of and have complied with the international code of ethics in publishing.

1. Joyce N Mbekeani: Data collection and write-up (neuro-ophthalmology), technical compilation, final approval for submission and corresponding author.
2. Manzoor Ahmed: Data collection and write-up (neuro-radiology), editing and final approval of submission.

Disclosures

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