Case Report

Suprasellar dermoid cyst associated with colloid cyst of the third ventricle: Disordered embryogenesis or a mere coincidence?

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ABSTRACT

Intracranial dermoid cyst and colloid cysts of the third ventricle are rare benign congenital lesions of early adulthood. Both lesions are thought to be congenital in origin however association is rare. Only one case of this association has been reported. We report a 22-year-old male with suprasellar dermoid cyst and colloid cyst of the third ventricle presenting simultaneously. Embryogenesis of this association has been discussed.

Key words: Anterior neuropore closure, colloid cyst, neural tube defects, suprasellar dermoid

Introduction

Intracranial dermoid cysts and colloid cysts are rare congenital lesions generally occurring along the midline.[1-3] Dermoid cysts are known to be ectodermal in origin while debate still exists about the embryological origin of colloid cysts.[3] Here, we report a 22-year-old young male with suprasellar dermoid cyst associated with a colloid cyst of the third ventricle and discuss the possibility of their unified embryological derivation. To best of our knowledge, only one similar case has been reported previously.[4]

Case Report

A 22-year-old male patient presented with on and off frontal headaches and occasional blurring of vision for 1 year. Patient was operated initially for suprasellar dermoid cyst 5 years back. Bifrontal craniotomy and partial excision of dermoid cyst was performed during previous surgery. Patient was asymptomatic 4 years after first surgery. Clinical examination was normal with visual acuity of 6/6 in both eyes without evidence of any field defects on perimetry. Routine hematological and biochemical investigations were normal. Hormone status of the patient was normal. Magnetic resonance imaging (MRI) revealed mainly cystic lesion in the suprasellar region with small solid component anteriorly which was hyperintense on T1-weighted sequences without any contrast enhancement. Cyst wall did not show any enhancement on contrast. Simultaneously a colloid cyst of 1.2 × 1.5 cm was seen at the foramen of Monro, mainly occupying third ventricle with evidence of mild hydrocephalus [Figure 1a]. An old bifrontal craniotomy was re-explored and complete excision of suprasellar dermoid cyst was achieved through bilateral subfrontal approach along with transcortical transventricular excision of colloid cyst in the same setting. Intraoperatively, suprasellar cyst was containing a whitish fluid with fat globules with small solid vascularized fatty nodule. Post-operative scans showed complete excision of both the lesions [Figure 1b]. Histopathological findings confirmed the diagnosis [Figure 2].

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Discussion

Intracranial dermoid cysts generally occur along the midline and are derived from the trapped somatic ectoderm during embryological development during third to fifth week of gestation.[3] These cysts are generally

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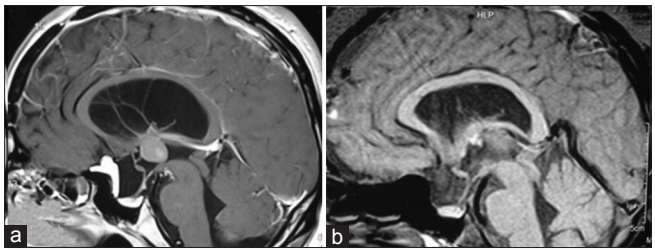

Figure 1: Sagittal sections of contrast-enhanced magnetic resonance images of brain showing cystic lesion in the suprasellar area with small solid part anteriorly. Cyst displaced the pituitary stalk posteriorly. Pituitary gland is seen compressed in the sella. Colloid cyst of the third ventricle is evident with mild ventriculomegaly (a). Post-operative scans (48 hrs) scans showed complete excision of dermoid and colloid cyst with decreased mass effect on the optic chiasm (b)

associated with dermal sinus tracts and other neural tube closure defects; however, dermal sinus tract may be absent in suprasellar and cerebello-pontine angle dermoids.[3,4] These lesions mainly presents with headache, local sings of mass effect, or neural compression. Spontaneous rupture and chemical meningitis, hydrocephalus are rare presentations.[3,4] Surgical excision is preferred treatment; however, complete excision is seldom achieved.

Colloid cysts of the third ventricle are also rare benign midline lesions presenting during early adulthood. Colloid cyst generally become symptomatic in early adulthood and mainly presents with headache and hydrocephalus.[2] Sudden death is most feared presentation which is fortunately rare.[2] Although controversy exists about timing of surgery of asymptomatic colloid cysts, symptomatic ones are excised surgically at the earliest. With advent of endoscopy, complete or partial resection of colloid cyst has become the preferred treatment option.

Congenital origin of these cysts is accepted universally; however, cell of origin is highly debated.[1,4,5] Initial reports suggested their neuroepithelial origin as the derivatives of paraphysis or ventricular ependyma and these were further supported later by other studies.[6,7] However, Mackenzie[8] in 1991 suggested the endodermal derivation of these cysts on the basis of ultrastructural features which was supported by other authors.[5,9] Until recently endodermal derivation of these cysts enjoyed the favor when Nagaraju *et al*.,[10] in 2010 reignited the debate by claiming colloid cysts as paraphysis remnants in human.

Colloid cysts are associated with various developmental malformations like corpus callosal agenesis, anterior cranial fossa encephalocels, cavum veli interpositi, and multiple neuroepithelial cysts and aqueductal stenosis.[1] These malformations represent a spectrum of neural tube closure defects and associated anomalies. Although isolated sporadic form of colloid cyst is

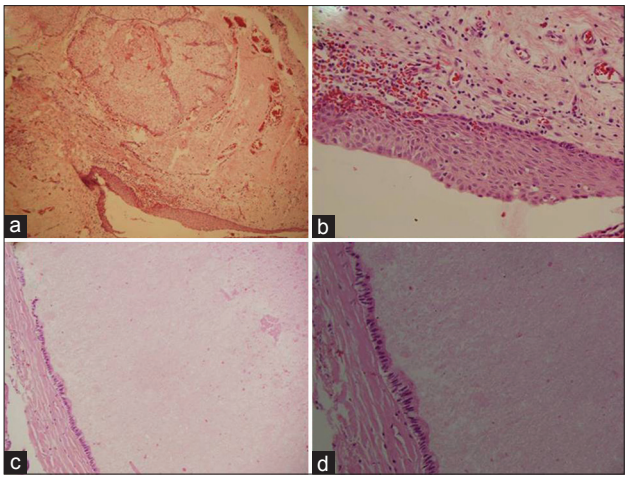

Figure 2: Photomicrographs showing stratified squamous epithelium with underlying sebaceous glands (a). Higher magnification of the lining epithelium (b). Colloid cyst wall comprising fibrocollagenous wall and is lined by columnar epithelium (c) with high-power view of lining epithelium (d). (a, b: HandE ×100; c, d: H and E ×200)

most common occurrence, these associations suggest disordered embryogenesis as a possible mechanism in their formation. Colloid cyst has also been reported to be associated with frontal epidermoid cyst.[11] Cheng *et al*.,[4] in 1999 reported a case of nasal dermoid sinus cyst associated with colloid cyst of third ventricle and proposed a fascinating theory of "anterior neuropore corridor defects" to explain the spectrum of these associated rare midline lesions. According to this theory, defective closure of situs neuroporicus promotes the neuroepithelium adjacent to commissural plate derivatives to form a vacuolated paraphysis growing slowly as colloid cyst of the third ventricle. Similar derivation from paraphysial remnants was also reported by Nagaraju *et al*., in 2010 supporting Cheng's hypothesis.[10] Although lack of substantial evidence remained main drawback of this theory, disordered embryogenesis appears to be the most plausible explanation in our case rather than mere coincidence.

Conclusions

Suprasellar dermoid cyst associated with colloid cyst of the third ventricle is a rare presentation. This association appears more than coincidence and can be partially explained by anterior neuropore corridor defect theory. This unique case provides the insight to the embryogenesis of other midline congenital lesions.

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