

Case Report

Suprasellar epidermoid cyst: a rare cause of painless progressive bilateral vision loss-case report with clinico-radiological correlation

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Received: 27 June 2018

Accepted: 26 July 2018

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ABSTRACT

Intracranial epidermoid cysts are relatively rare lesions. They result from inclusion of ectodermal elements during time of neural tube closure. This lesion could rarely be acquired due to post-surgical or post traumatic implantation of the ectodermal components. They typically present in middle age group patients with evidence of loco-regional mass effect on adjacent structures. We present a clinico-radiological case report of the 27-year-old female patient who presented with painless progressive bilateral vision loss for last 4 months.

Keywords: Epidermoid cyst, Radiology, Suprasellar, Vision loss

INTRODUCTION

Basically, epidermoid cysts are benign inclusion tumours comprising of ectodermal elements (keratin and cholesterol crystals resulting from desquamation of epithelial cells) during the period of neural tube closure. They comprise around 1% of central nervous system tumours.¹⁻³

Usual intracranial location for these lesions include cerebellopontine angle (up to 40-50%), fourth ventricle (up to 17%), followed by parasellar region (up to 15%). Less common regions include interhemispheric fissure (less than 5%) and calvarium (10%).⁴⁻⁶

Radiologically they present as a suprasellar lobulated cystic lesion following CSF signal intensity and can pose a diagnostic challenge.⁷ Authors report a classical radiological (CT scan and MRI) features of a case of suprasellar epidermoid cyst in an adult female.

CASE REPORT

27-year-old female patient was referred for radiological evaluation to rule out the cause of bilateral progressive vision loss (left > right) for last 4 months. There was no history of any previous trauma, assault or seizure episodes. On neurological examination the patient was conscious and co-operative. No features suggested raised intracranial pressure. No evidence of any facial hypoesthesia or asymmetry. Bilateral pupil sizes were symmetrical (3.5 mm). Visual acuity was decreased in both eyes (right-patient could only do finger count close to the face, left-negative perception to light). Perception to light was present in the right pupil but note was made of relative afferent papillary defect on left side. Non contrast CT scan (Figure 1A) showed a large lobulated hypodense lesion in suprasellar region insinuating along the anterior inter-hemispheric fissure, along bilateral basifrontal lobe (left>right) and bilateral parasellar regions.

MRI showed a lobulated T2 hyperintense (Figure 1B) and T1 hypointense (Figure 1C) lesion showing incomplete suppression on T2 FLAIR sequence (Figure 1D). No obvious enhancement noted on post contrast T1 sequence (Figure 1E). The mass lesion is seen to show DWI restriction (f Figure 1F). On MR angiography (Figure

1G) the mass is seen to encase the circle of Willis vessels. On coronal reformatted images (Figure 1H and I) the mass is seen to compress the optic chiasma, thus explaining the vision loss. The lesion is also seen to compress the left proximal optic nerve (Figure 1J), thus explaining the left sided relative afferent pupillary defect.

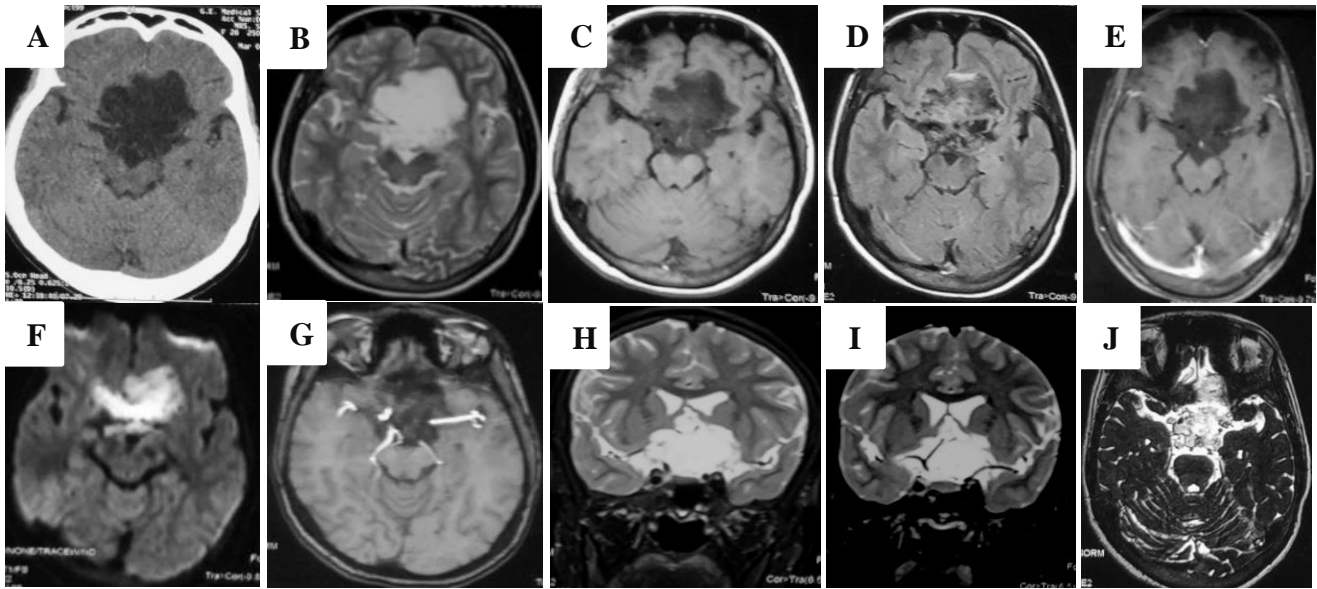


Figure 1: Non contrast CT scan. (A) A large lobulated hypodense suprasellar lesion which follows t2 hyperintense (B) T1 hypointense (C) Signal with incomplete suppression on T2 flair sequence (D) Non enhancement on post contrast T1 sequence (E) The lesion shows DWI restriction (F) Encasing circle of willis on MR angiography (G). coronal T2 images. (H, I and J) Compression of the optic chiasma and axial fiesta sequence shows mass encroachment along left proximal optic nerve.

DISCUSSION

Epidermoid cysts are slow-growing, benign tumours, and although they can reach a great size, they are usually very well circumscribed. Epidermoid cysts are inclusion cystic lesions formed by incorporation of ectodermal elements between 3rd to 5th week of embryogenesis during neural tube closure. They are also termed ‘pearly tumours’ and are mostly noted in 4th-5th decade individuals with no or slightly higher predilection for male gender.⁸ The most common locations of cranial epidermoid cysts are cerebellopontine cistern followed by suprasellar/parasellar region and fourth ventricle. Rare locations include cerebral parenchyma, intraventricular, pineal region and brainstem. Lesions in the cerebello-pontine angle and parasellar location could be due to proliferation of multipotent embryogenic cell rests or lateral displacement of ectodermal cells by the developing otic vesicles.⁹⁻¹⁰ They grow by accumulation of keratin and cholesterol, which are breakdown products created by the desquamation of epithelial cells. They have tendency to grow along available cisternal spaces, so there is no mass effect initially and they may remain

asymptomatic for many years. Slow growth over the years is typical and these cysts possess a linear growth pattern rather than an exponential one. Clinical features vary depending upon the location of the lesion. Symptoms are mainly due to mass effect or compression of cranial nerves.¹⁰

Typically, they are irregular, insinuating lesions that encase adjacent neurovascular structures. Suprasellar/sellar lesions usually present with non-specific headache and visual disturbances. In other locations, headache (either non-specific or that of intracranial hypertension), seizures and motor/sensory deficits have been noted. Hydrocephalus is rare and is usually a late manifestation.¹¹

On CT, they usually appear homogeneously hypodense, non-enhancing lesions but can rarely appear hyperdense. On MRI, they appear hypointense on T1, hyperintense on T2 sequences, hypointense on constructive interference in steady state (CISS) images. They uniformly restrict on diffusion sequences and this remains the most sensitive and specific MR sequence for epidermoid cyst.

Calcification is seen in 10%- 25% of cases. Further, the presence of protein or debris can change the homogeneous signal to a non-homogeneous one. The rare white or dense epidermoids show reversed signal intensity on T1 and T2 images.¹¹⁻¹²

Craniopharyngioma (adamantinomatous type), dermoid cysts, rathke cleft cyst and arachnoid cysts are the most important differential diagnoses. Adamantinomatous type of craniopharyngioma (more common 90% as compared to papillary type) usually present as the solid cystic suprasellar mass with few calcific specs within and the solid component shows vivid post contrast enhancement.¹³ Dermoid cysts appear as well defined low attenuating (fat density, negative Hounsfield unit on CT and T1 hyperintense) lobulated masses with occasional wall calcification and thin peripheral rim enhancement.¹⁴ Rathke cysts are classically believed to arise from failure of obliteration of the lumen of rathke pouch and usually follow CSF signal intensity on CT and MRI, however they can show T1 hyperintensity due to high protein content, but usually never show restriction on diffusion weighted MRI. They usually arise in the cleft between anterior and posterior pituitary lobe.¹⁴ Most suprasellar arachnoid cysts are congenital, secondary to an imperforate membrane of Lilliequist and follow CSF signal intensity on all sequence, without any evidence of post contrast enhancement or DWI restriction.¹⁴ Complete surgical excision is the treatment of choice with consistent good excellent long-term results being reported in various series. However, as portions of the capsule may become densely adherent to surrounding structures, total resection could be extremely difficult and subtotal resection may have to be performed in such cases to prevent neurological damage. Recurrences are uncommon and mostly seen after subtotal resections.¹⁵

CONCLUSION

Epidermoid cysts need to be considered in the differential diagnosis of suprasellar cystic lesions, especially those showing diffusion restriction. We report a case of a suprasellar epidermoid cyst in 27-year-old female patient presenting with bilateral progressive painless vision loss with typical radiological findings.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Chowdhury FH, Haque MR, Sarker MH. Intracranial epidermoid tumor; Microneurosurgical management: An experience of 23 cases. Asian J Neurosurg. 2013;8:21-8.

2. Ren X, Lin S, Wang Z, Luo L, Jiang Z, Sui D, et al. Clinical, radiological, and pathological features of 24 atypical intracranial epidermoid cysts. J Neurosurg. 2012;116:611-21.
3. Schiefer TK, Link MJ. Epidermoids of the cerebellopontine angle: A 20-year experience. Surg Neurol. 2008;70:584-90.
4. Kato K, Ujiie H, Higa T, Hayashi M, Kubo O, Okada Y, et al. Clinical presentation of intracranial epidermoids: A surgical series of 20 initial and four recurred cases. Asian J Neurosurg. 2010;5:32-40.
5. Osborn AG, Preece MT. Intracranial cysts: Radiologic-pathologic correlation and imaging approach. Radiology. 2006;239:650-64.
6. Kallmes DF, Provenzale JM, Cloft HJ, McClendon RE. Typical and atypical MR imaging features of intracranial epidermoid tumors. AJR Am J Roentgenol. 1997;169:883-7.
7. Russell D, Rubinstein LJ. Congenital tumours of maldevelopmental origin. In: Pathology of the nervous system. Baltimore: Williams and Wilkins. 1977:24-64.
8. Bauman CH, Bucy PC. Paratrigeminal epidermoid tumors. J Neurosurg. 1956;13:455-68.
9. Abramson RC, Morawetz RB, Schli HM. Multiple complications from an intracranial epidermoid cyst: Case report and literature review. Neurosurg. 1989;24:574-8.
10. Alvord EC Jr. Growth rates of epidermoid tumours. Ann Neurol. 1997;2:367-70.
11. Hakyemez B, Aksoy U, Yildiz H, Ergin N. Intracranial epidermoid cysts: Diffusion-weighted, FLAIR and conventional MR findings. Eur J Radiol. 2005;54:214-220.
12. Russell DS, Rubinstein LJ. Pathology of Tumors of the Nervous System. 4th ed. Baltimore: Williams and Wilkins; 1977:29-32.
13. Sartoretti-Schefer S, Wichmann W, Aguzzi A, Valavanis A. MR differentiation of adamantinous and squamous-papillary craniopharyngiomas. Am J Neuroradiol. 1997 Jan 1;18(1):77-87.
14. Osborn AG, Preece MT. Intracranial cysts: Radiologic-pathologic correlation and imaging approach. Radiology. 2006;239(3):650-64.
15. Yamakawa K, Shitara N, Genka S, Manaka S, Takakura K. Clinical course and surgical prognosis of 33 cases of intracranial epidermoid tumours. Neurosurgery. 1989;24:568-73.

Cite this article as: Chauhan G, Singh V, Phadke RV, Yadav S. Suprasellar epidermoid cyst: a rare cause of painless progressive bilateral vision loss- case report with clinico-radiological correlation. Int J Res Med Sci 2018;6:3186-8.