

# A Treatable Cause of Progressive Visual Loss

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## Abstract

Sphenoethmoidal encephalocoele is a rare clinical entity with varying clinical presentation. It can be seen in association with corpus callosal agenesis and midline facial defects. We are reporting a case of 44 year old female presented with progressive vision loss of right eye, and surgical correction of meningoencephalocoele and the defect prevent further visual detiortion.

## Case Discussion

Reporting a case of 44 year old female presented with history of gradually progressive painless visual loss of right eye for past 6 months which was insidious in onset .She had nasal block on the right side, no nasal discharge. No history of diplopia, headache, vomiting or fever. Past history was remarked with pthisis bulbi of left eye following a trauma at 13 years and history of surgical correction of cleft lip and palate. Her right eye vision was fairly normal till 6 months back; the only functioning eye. She is a diabetic and hypertensive on treatment. On examination her higher mental function was normal, visual acuity was only counting finger at 1 feet on right eye, no vision left eye, fundus showed pale disc on right side with features of diabetic retinopathy. VEP showed reduced P100 amplitude on right side. MRI had been done which showed sphenoethmoidal encephalocoele with corpus callosal agenesis. She was referred to higher centre for transnasal endoscopic repair, following which her visual acuity on the right side became CF at 3 feet on 7 th day post surgery and no further progression of symptoms and improved to 6/60 at one month. Postoperative CT scan showed no evidence of hemorrhage infarct or midline shift.<sup>1</sup>

## Images and description

MRI showed parallel running lateral ventricles and colpocephaly(Racing Car appearance), large central sphenoethmoidal meningoencephalocoele with large bony defect in anterior sellar floor, planum sphenoidale and ethmoid bone. Herniated sac (4.1x3.5x2.6) contains bilateral optic nerves olfactory nerves, infundibulum of pituitary gland and csf. Pthisis bulbi of left eye noted.

Encephalocoeles can be of spontaneous /congenital or traumatic in origin. Transphenoidal encephalocoele is a rare clinical entity and may have varying clinical presentation. Prompt diagnosis and surgical treatment is required in such patients. Reports of association of sphenoethmoidal meningoencephalocoele with corpus callosal agenesis have been there in the literature. sphenoethmoidal encephalocoele causing progressive visual loss have not been reported yet.<sup>2</sup>

## Case Report

## Open Access &

## Peer-Reviewed Article

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## Keywords:

MRI, Sphenoethmoidal encephalocoele, visual deterioration.

**Received:** February 23, 2023

**Accepted:** October 30, 2023

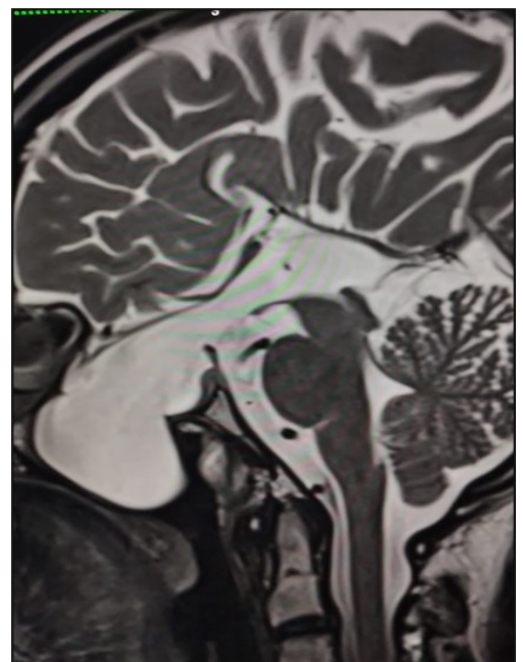
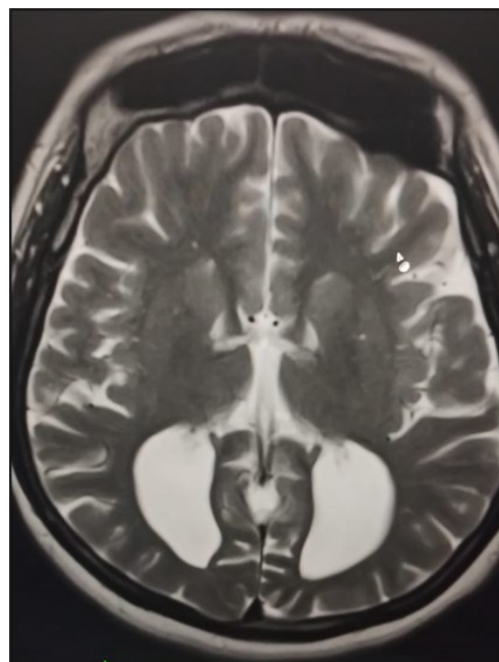
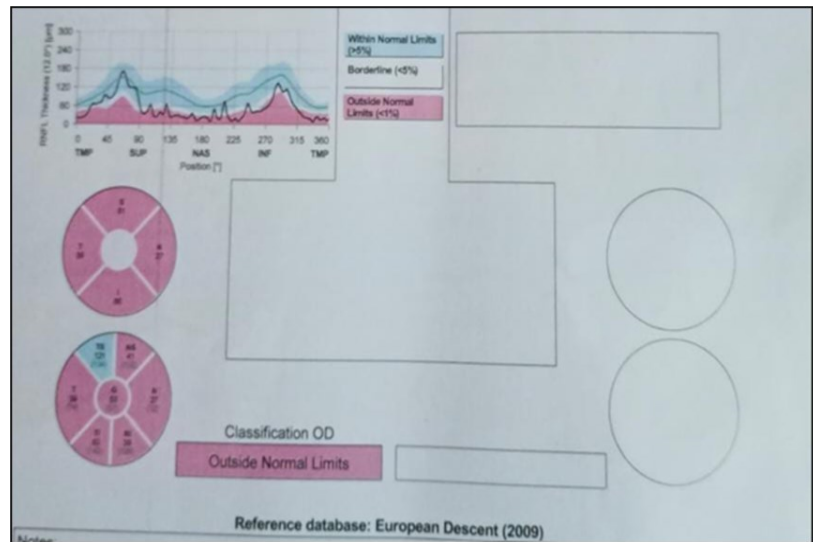
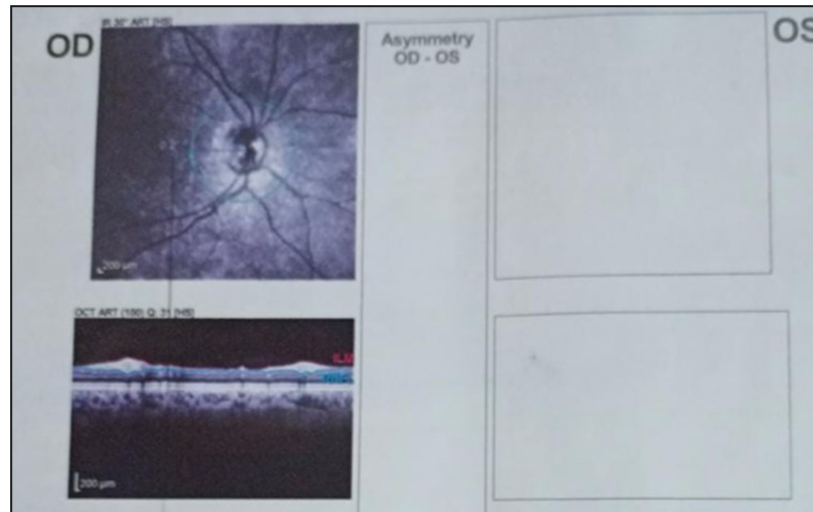
**Published:** December 18, 2023

## Academic Editor:

Jamal Uddin, Assistant Professor, Computer Science Department, Qurtuba University Of Science & Information Technology, Peshawar, KP, Pakistan.

## Citation:

Dr. Miny Susan Abraham, Dr. Nikhil Gladson (2023) A Treatable Cause of Progressive Visual Loss. International Journal of Neuroimaging - 1(1):1-3.



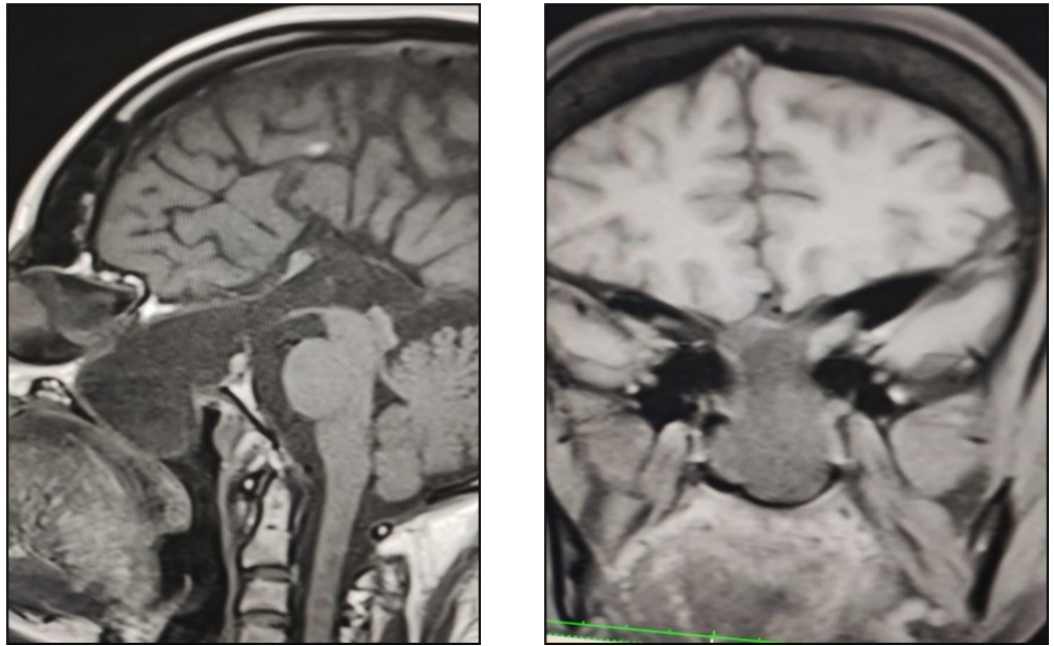


Figure 1. Fundus Images and OCT – right optic atrophy ; pale optic disc.

### Running Title

Sphenoethmoidal meningoencephalocele should be considered as one of the differential diagnosis of progressive vision loss in patients with midline craniofacial defects. Prompt diagnosis and surgical repair may prevent further visual deterioration.<sup>3</sup>

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