Morning Glory Syndrome. Case Report and Literature Review

Síndrome de Morning Glory. Relato de Caso e Revisão de Literatura

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ABSTRACT
Morning Glory Syndrome is a very rare condition. It is characterized by an enlarged and funnel-shaped optic disc excavated and is associated with other craniofacial anomalies such as hypertelorism, cleft lip, cleft palate, and basal encephalocele. Case Report. A case of a 30 year-old man who came for neurosurgical evaluation after a magnetic resonance imaging (MRI) scan that showed sphenoidal encephalocele is reported. Conclusions. There is a high association between basal encephalocele, midface abnormalities and Morning Glory Syndrome. A neurosurgical, ophthalmological and endocrinological evaluation must always be done. The patients present with ophthalmic signs and/or strabismus or poor vision. An image study should always be performed to delineate the extent of the lesion and a complete hormone screening should be carried out to exclude pituitary deficiency.

Key words: Morning Glory Syndrome; Basal encephalocele; Hypertelorism; Midface Abnormalities.

RESUMO
A Síndrome de Morning Glory é uma entidade bastante rara. Caracteriza-se por uma papila óptica escava e em formato de funil e está associada com outras malformações craniofaciais como hipertelorismo, fenda labial, fenda palatal e encefalocele basal. Relato do Caso. Relatamos aqui o caso de um homem de 30 anos que veio para avaliação neurocirúrgica após a ressonância magnética (RM) de encéfalo evidenciar encefalocele basal. Conclusão. Existe uma alta associação entre encefalocele basal, malformação do andar médio da face e a síndrome de Morning Glory. Uma avaliação neurocirúrgica, oftalmológica e endocrinológica deve sempre ser realizada. Os pacientes com sinais ophthalmológicos, estrabismo e/ou baixa acuidade visual. Um exame de imagem deve ser realizado para delinear a extensão da lesão além se streaming hormonal completo para excluir panhipopituitarismo.

Palavras-chave: Síndrome de Morning Glory; Encefalocele Basal; Hipertelorismo; Anormalidades.

INTRODUCTION

The term ‘Morning Glory Syndrome’ (MGS) was first described by Kindler¹ because of the papillary region in the affected eye is reminiscent of (whitering) flower of ‘Morning Glory’. Clinical and histopathological reports gave rise to definition of MGS as a unilateral malformation involving the optic disc associated with a peripapillary scleral defect, absence of lamina cribosa and a recess, formed by an axial retrodisplacement of the optic nerve. Manschot had postulated that MGS is a mesodermic disorder². A number of ocular, craniofacial and renal abnormalities have been reported in association with MGS. The craniofacial abnormalities include hypertelorism, cleft lip and palate and basal encephalocele as described by Koenig et al.³ (1982), Caprioli & Lesser³ (1983), Hope-Ross & Johnston⁴ (1989), Itakura et al.⁵ (1992) and Eustis et al.⁶ (1984). This condition occurs when the bone at the base of the skull is deficient and causes the cranial contents to protrude beyond the skull. Basal encephalocele appears in 1:35,000 live births⁷. It is frequently occult and is associated with midline deficiencies such as cleft palate and corpus callosum agenesis⁸-¹¹. The proposed
hypertelorism, broad nasal root, cleft lip, and cleft palate. The condition has been classified into five anatomic types by several authors depending on the site of herniation:

- Spheno-ethmoidal - which describes the herniation of cranial contents through the sphenoid and ethmoid bone into the posterior nasal cavity;
- Trans-sphenoidal - herniation through the body of sphenoid bone into the sphenoid sinus or epipharynx;
- Spheno-orbital – herniation through the superior orbital fissure or osseous defect into the orbit;
- Transethmoid – herniation through the lamina cribosa into the anterior nasal cavity;
- Spheno-maxillary – herniation through the inferior orbital fissure into the pterygopalatine fossa;

Symptoms of basal encephalocele vary according to the site and size of lesion. They are often occult and are usually detected when complications caused by the encephalocele mass are arisen. Respiratory distress, nasal obstruction, apnoea episodes, difficulty feeding and failure to thrive are the result of the direct pressure of the encephalocele on the nasopharynx.

Associated anomalies

Basal encephaloceles are associated with other congenital malformations. Diagnoses are often late and occur with the onset of symptoms of visual disturbance and pituitary dysfunction. Four main types of association that have been described are encephalocele-hypothalamic-pituitary dysplasia, encephalocele oculo-auriculo-vertebral spectrum, and encephalocele-frontonasal dysplasia.

Diagnosis

A CT (computed tomography) scan is very helpful to see bone defects. MRI scans are necessary to prove cerebral connections. MRI scans also have the advantage of highlighting ocular lesions. Diabetes insipidus is the most common finding in pituitary deficiency and is usually indicated by a variable thyroid hormone and cortisol response. Basal encephalocele must be remembered and suspected when nasal obstructions are found. Diagnostic CT and MRI delineate the anatomy of herniated mass. MGS is a congenital optic disc anomaly associated with basal encephalocele. MGS associations necessitate ophthalmological examination and endocrine review.
CASE REPORT

A 30-year-old man came for neurosurgery evaluation. He was born by a C-section delivery. He presented with cleft nasal, cleft lip, cleft palate and sphenoid-ethmoidal encephalocele. During his infancy he presented with episodes of mouth breathing and chronic mucous nasal discharge but no apnoea or respiratory distress. The pregnancy was uneventful and no significant congenital factors were discovered. He had never been sent for correction of the cleft palate. At this neurosurgical evaluation it could be seen hypertelorism, broad based root, cleft lip, cleft palate, and a sphenoid-ethmoidal encephalocele. In MRI scan we could see a sphenoid-ethmoidal encephalocele, and corpus callosum agenesis. In the palate, a pulsatile with a mucous membrane covering was seen.

Ophthalmologic examination showed a visual loss on the left side and it was seen a dysplastic and funnel-shaped optic disc, and dysplastic vessels arising the optic disc in radial disposition. Ultrasound showed a posterior sclera defect, that is the classic finding. A three-dimensional computed tomography (3D CT) revealed a midline bone hypoplasia, and in the anterior fossa a gap filled by an extensive meningcele that extended to the nasopharynx. MRI scan confirmed the presence of a large sphenoid-ethmoidal encephalocele that produced a large soft tissue mass within the posterior nasopharynx and communicating with the third ventricle.

Other findings included distortion of intracranial optic nerves, corpus callosum agenesis, ectopic pituitary gland at inferior margin of encephalocele, hypertelorism and low cribiform plate.
Figure 3. Cleft palate. Basal Encephalocele.

Figure 4. Midface Hypoplasia.

Figure 5. Anterior Fossa Gap.

Figure 6. Palate View - Palate Gap.

Figure 7. Basal Encephalocele.
In this syndrome we find a basal bone gap which results in protrusion of meninges and their contents. So, in these cases, it can be found brain tissue in nasal cavity, nasopharynx, the sphenoid and ethmoid sinuses, the orbit and pterygopataline fossa. That can be mistaken to nasal polyps or enlarged adenoids and may be associated with cleft palate. So, after operating on these cases, there can occur cerebral spinal fluid rhinorrhea, meningitis and even death.

Kindler\(^1\), in 1970, proposed that the optic nerve head was funnel-shaped, contained a central white dot of glial tissue, and was surrounded by a elevated annulus of chorioretinal pigment disturbance\(^2\). The retinal vessels radiate out from the edge of the disc as multiple narrow branches. According to Krause, the typical features of Morning Glory Syndrome is an excavated optic disc and an elevated circumpapillary ring of chorioretinal pigmentary disturbances\(^3\). The presence of midline facial anomalies such as broad based root, hypertelorism, cleft lip and cleft palate in association with anomalies of the optic nerve should alert the physician to the possible presence of encephalocele.

### References


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Não há conflito de interesses