

ent diffusion coefficient (ADC), from necrotic tumors, which show a low diffusion signal with high ADC.^{1,3}

The final diagnosis is usually histological after surgical drainage. Coagulase-negative *Staphylococcus* is the most commonly isolated microorganism.² According to the different series, the ratio between Gram stain and positive cultures ranges from 0% to 64%, which may first be explained by the low activity of bacteria in pituitary abscesses,⁵ and second, by preoperative antibiotic therapy.^{1,5}

The treatment of choice consists of transsphenoidal surgical drainage and antibiotic therapy for 3–6 weeks.^{1,2,4–7} However, conservative antibiotic therapy may be useful for early pituitary abscesses.¹ Visual deficiencies usually improve after treatment, but endocrine deficiencies may persist and require permanent replacement therapy. The recurrence rate is low, but MRI monitoring is advisable in order to detect recurrent abscess.

In conclusion, it can be said that preoperative diagnosis of pituitary abscess is difficult because of its insidious clinical signs and symptoms and poorly specific radiographic findings. Pituitary abscess should be suspected in a patient with a cystic sellar mass with ring-shaped enhancement,¹ particularly when associated with clinical signs of infection and/or diabetes insipidus.

Because of the low frequency of pituitary abscess, we would like to see a multicenter study with a larger patient sample being conducted, with the aim of furthering our understanding of this condition.

References

1. Liu F, Li G, Yao Y, Yang Y, Ma W, Li Y, et al. Diagnosis and management of pituitary abscess: experiences from 33 cases. *Clin Endocrinol*. 2011;74:79–88.
 2. Dalan R, Leow MK. Pituitary abscess: our experience with a case and a review of the literature. *Pituitary*. 2008;11:299–306.
 3. Shuster A, Gunnarsson T, Sommer D, Miller E. Pituitary abscess: an unexpected diagnosis. *Pediatr Radiol*. 2010;40:219–22.
 4. Vates GE, Berger MS, Wilson CB. Diagnosis and management of pituitary abscess: a review of twenty-four cases. *J Neurosurg*. 2001;95:233–41.
 5. Zhang X, Sun J, Shen M, Shou X, Qiu H, Qiao N, et al. Diagnosis and minimally invasive surgery for the pituitary abscess: a review of twenty nine cases. *Clin Neurol Neurosurg*. 2012;114:957–61.
 6. Kuge A, Sato S, Takemura S, Sakurada K, Kondo R, Kayama T. Abscess formation associated with pituitary adenoma: a case report: Changes in the MRI appearance of pituitary adenoma before and after abscess formation. *Surg Neurol Int*. 2011;2:3.
 7. Talluri SK, Talluri J, Besur S. Pituitary abscess. *Am J Med Sci*. 2011;341:484.
 8. Nattero L, Luque-Ramírez M, Azcárate A, Marazuela M. Hiponatreemia recurrente como característica presente en los abscesos de pituitaria: caso clínico. *J Endon*. 2010;12:3–125.
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Morning glory syndrome associated with transsphenoidal encephalocele and panhypopituitarism[☆]



Síndrome «morning glory» asociado a encefalocele transesfenoidal y panhipopituitarismo

So-called morning glory syndrome (MGS), although first described in 1929, owes its name to Kindler (1970).¹ In this syndrome, congenital optic nerve dysplasia exists. The incidence of MGS is very low, and it is caused by the failure of the closure of the choroidal embryonic fissure. MGS, which has an autosomal dominant inheritance, is usually unilateral,

although bilateral cases occur, predominates in females, and is characterized by a funnel-shaped excavation with central fibroglial tissue and radial retinal vessels mimicking the morning glory flower. MGS frequently occurs with isolated ophthalmological changes (decreased visual acuity associated with retinal detachment in 30–40% of patients), but systemic associations have been reported, including congenital anomalies in the forebrain and midline, with progressive endocrine, respiratory, or renal abnormalities.^{1,2}

Congenital basal encephalocele (EB) is due to a skull bone and dura mater defect with extracranial herniation of cranial structures. It is a rare anomaly, difficult to diagnose, with an estimated incidence of 1:35,000 newborns. There are four types of encephalocele, of which the transsphenoidal variety is the least common (accounting for 5% of BEs).³ This type of encephalocele is due to the persistence of the craniopharyngeal or transsphenoidal canal, with brain tissue herniation through it. It may result from a defect in the floor of the sella turcica, the sphenoid sinus, or the posterior ethmoid sinus. The transsphenoidal transsellar variant is the least common. It is often associated with midline defects, with hypothalamic-pituitary hormone and optic nerve changes, including MGS.⁴ MGS is associated with 67% of BEs.

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The incidence of hormone dysfunctions in patients with BE is 50–60%. GH deficiency, hypogonadotropic hypogonadism, hypothyroidism, and diabetes insipidus are the most common disorders. A review of 15 cases with transsphenoidal BE showed that GH and antidiuretic hormone deficiencies were the most common (66.7 and 60% respectively), followed by gonadotropin (33.3%), TSH (26.7%), and prolactin deficiency (13.3%).⁵

The natural course of hypothalamic-pituitary dysfunction is still uncertain, but progressive hormone dysfunction has been found in most patients with BE. Unfortunately, very few cases with endocrinological monitoring for 10 years or longer have been reported.³

We report a case of bilateral MGS associated with transsphenoidal encephalocele and panhypopituitarism in which imaging and hormonal studies were essential for diagnosis.

A 16-year-old female adolescent^{6/12} was referred for amenorrhea and delayed growth, which had been slow since infancy, but with no stagnation. Axillary hair growth and pubarche occurred at 11 years of age, but did not progress. Thelarche started at 16 years of age. No menarche occurred. The patient had no urinary frequency or polydipsia.

She had been born by normal delivery after a controlled 30-week pregnancy, and had a length of 49 cm (+0.05 SD) and a weight of 3240 g (+0.44 SD) at birth. At three months of age, the patient was diagnosed bilateral MGS, which caused at 11 years retinal detachment in the left eye and decreased vision in the right eye. She had no history of seizures or any other remarkable history.

A physical examination at 16^{6/12} years revealed the following: height 145 cm (−2.7 SD), BMI 23.16 kg/m² (+0.98 SD), target height 163 cm (−0.15 SD). Bone age of 14^{9/12} years. Microphthalmos, right eye nystagmus, very thin upper lip, gothic palate. There was no goiter or fat accumulation in the abdomen. Tanner 2 pubertal stage. There were no other significant findings.

Hormone test results included: TSH 1.67 μ IU/mL (NR, 0.350–4.950), FT4 1.08 ng/dL (NR, 0.700–1.600), basal cortisol: 21.20 μ g/dL (NR, 5.00–25.00), ACTH 50.3 pg/mL (NR, 5–46). Response to insulin-induced hypoglycemia was normal (peak cortisol level, 26.7 μ g/dL). PRL 61.6 ng/mL (NR,

<20). Basal GH <0.05 ng/mL with a peak of 0.06 ng/mL (NR, >7) in the clonidine test with no estrogen priming. IGF1 86 ng/mL (NR, 116–913). Basal FSH 2.77 mIU/mL, basal LH 1.21 mIU/mL. Estradiol 21 pg/mL (NR in prepubescents <12 pg/mL). A GnRH test showed a peak of FSH 7.46 mIU/mL and LH 9.2 mIU/mL. Bone densitometry showed osteoporosis of −3.2 SD (at L1–L4 levels). Urinary osmolality was not tested because there were no symptoms of diabetes insipidus.

A CT scan with 3D reconstruction and MRI of the brain (Fig. 1) showed transsphenoidal meningoencephalocystocele, containing dysplastic hypothalamic-pituitary tissue in its lower part, agenesis of the rostrum of the corpus callosum, and a right retinal coloboma. Estrogen therapy was started with transdermal estrogens.^{6,7} GH was not administered because of bone age, as this treatment is clearly effective only if administered at an early age.⁸

Due to osteoporosis, calcium and vitamin D were started in addition to estrogen. Mild hyperprolactinemia could be attributed to elongation of the pituitary stalk. Surgical repair of encephalocele, as advocated by many authors, was not performed because it is often not beneficial due to the risk of damaging functioning tissue.⁹

Our patient had bilateral MGS associated with transsphenoidal encephalocele with endocrine changes, including GH and gonadotropin deficiency with a late diagnosis of hypothalamic-pituitary involvement, despite the fact that the history of the syndrome was known. We therefore emphasize the need for early endocrinological work-up and long-term follow-up, because hormone deficiencies may appear years after the initial diagnosis.^{3,10} A CT or MRI scan makes it possible to outline the anatomy of the herniated mass.

To sum up, MGS requires an imaging study to rule out encephalocele, and an assessment of hypothalamic-pituitary function for the early diagnosis and treatment of hormone deficiencies. The hormones mainly affected include GH, with an impact on final height, and gonadotropins with the resultant lack of estrogenization, which leads to early osteoporosis, in addition to a lack of pubertal development. Hormone replacement therapy is highly effective.^{6–8} It should be borne in mind that hormone deficiencies may be progressive and occur years after

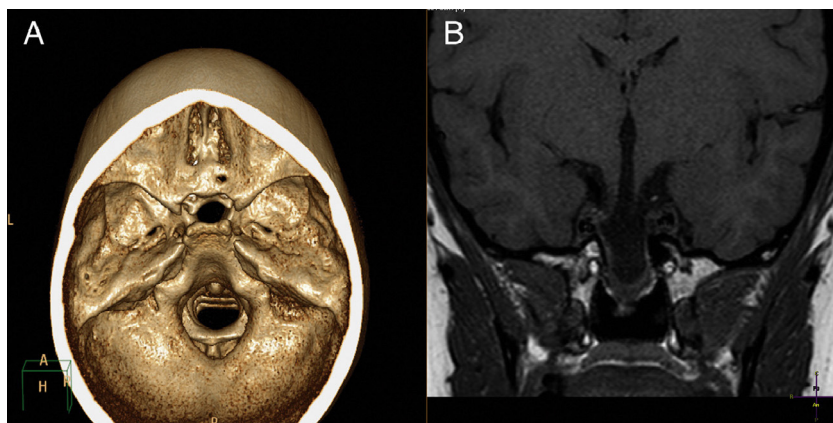


Figure 1 (A) Three-dimensional computed tomography showing bone defect at the base. (B) Magnetic resonance imaging. T1-weighted coronal section showing cystic mass extending to the nasopharynx through bone defect.

the initial diagnosis. Adequate monitoring of hypothalamic-pituitary axis function is therefore required.

References

1. Pierre-Filho Pde T, Limeira-Soares PH, Marcondes AM. Morning glory syndrome associated with posterior pituitary ectopia and hypopituitarism. *Acta Ophthalmol Scand.* 2004;82:89–92.
2. López-Lizárraga EP, Bolaños-Jiménez R, Treviño-Alanís MG, Rivera-Silva G. Morning glory syndrome. *Gac Med Mex.* 2011;147:70–1.
3. Tanimoto K, Onda S, Sawaki H, Hiraiwa T, Sano H, Ohnishi M, et al. Hypopituitarism in a patient with transsphenoidal cephalocele: longitudinal changes in endocrinological abnormalities. *Endocr J.* 2011;58:193–8.
4. Minotto I, Abdala N, Miachon AA, Spinola e Castro AM, Ima-mura P, Nogueira RG. Basal encephalocele associated with morning glory syndrome: case report. *Arq Neuropsiquiatr.* 2007;65:988–91.
5. Morioka M, Marubayashi T, Masumitsu T, Miura M, Ushio Y. Basal encephaloceles with morning glory syndrome, and progressive hormonal and visual disturbances: case report and review of the literature. *Brain Dev.* 1995;17:196–201.
6. Mauras N, Torres-Santiago L, Taboada M, Santen R. Estrogen therapy in Turner syndrome: does the type, dose and mode of delivery matter? *Pediatr Endocrinol Rev.* 2012;2:718–22.
7. Nabhan ZM, Dimeglio LA, Qi R, Perkins SM, Eugster EA. Conjugated oral versus transdermal estrogen replacement in girls with Turner syndrome: a pilot comparative study. *J Clin Endocrinol Metab.* 2009;94:2009–14. <http://dx.doi.org/10.1210/jc.2008-2123>.
8. Reiter EO, Price DA, Wilton P, Albertsson-Wikland K, Ranke MB. Effect of growth hormone (GH) treatment on the near-final height of 1258 patients with idiopathic GH deficiency: analysis of a large international database. *J Clin Endocrinol Metab.* 2006;91:2047.
9. Sanjari R, Mortazavi SA, Amiri RS, Ardestani SH, Amirjamshidi A. Intrasphenoidal meningo-encephalocele: report of two rare cases and review of literature. *Surg Neurol Int.* 2013;4:5. <http://dx.doi.org/10.4103/2152-7806.106260>.
10. Chen CS, David D, Hanieh A. Morning glory syndrome and basal encephalocele. *Childs Nerv Syst.* 2004;20:87–90.

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Spinal epidural abscess in a diabetic patient[☆]



Absceso epidural espinal en un paciente diabético

Spinal epidural abscess (SEA) is a rare infection of the central nervous system with an incidence of 2–25 per 100,000 patients admitted to hospital.⁸ However, there has been an increase in its frequency in the past 30 years due to the increasing number of patients undergoing spinal invasive procedures, either as anesthetic procedures or for pain control, and also because of the improved resolution and/or increased number of imaging procedures. SEA poses a diagnostic challenge because its manifestations are nonspecific and a delay in diagnosis may lead to irreversible neurological sequelae and even death. Diabetes is considered as a risk factor for the occurrence of SEA, and should therefore be included in differential diagnosis of any diabetic patient with lumbar pain.

We report the case of a 70-year-old male Caucasian patient who attended the emergency department for a second consecutive time reporting mechanic pain refractory to standard analgesics in the left back region, irradiating to the lumbar spine, for approximately 15 days. Twenty

days after the pain started, the patient experienced functional impotence with walking difficulty due to lower limb weakness. He had not experienced fever or sphincter changes, and did not report any other associated symptoms. There was no history of recent trauma, anesthetic block, or peripheral infection. The only remarkable history was amputation of the fourth and fifth metatarsal bones of the right foot for osteomyelitis with a microbiological report of the soft tissue culture positive for *Pseudomonas* sp.

The personal history of the patient included longstanding type 2 diabetes with fair chronic metabolic control, with an HbA1c value of 7.7% one month before admission, a slight worsening in metabolic control in laboratory tests at admission (HbA1c 7.9%), and multiple microvascular (non-proliferative retinopathy, nephropathy with stage 3B chronic renal disease, sensorimotor polyneuropathy, and Charcot arthropathy) and macrovascular (chronic ischemic cardiopathy, stroke some 25 years before with no sequelae, and Fontaine stage II intermittent claudication) complications. He was being treated with multiple insulin doses. The patient also had high blood pressure, adequately controlled with lercanidipine and irbesartan, and dyslipidemia treated with atorvastatin.

A physical examination revealed a good general condition, and the patient was afebrile and hemodynamically stable. There were no respiratory, cardiovascular, or abdominal pathological findings. A neurological examination found no motor or sensory deficits in the upper limbs or trunk. However, a lower limb examination revealed decreased patellar and ankle jerk reflexes in both limbs, as well as motor deficit

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