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Rathke's cleft cyst associated with pituitary granulomatosis with polyangiitis: An unusual combination of hypothalamus-pituitary region pathologies

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ABSTRACT

The authors present an unusual case of a patient suffering from visual deficit due to pituitary granulomatosis with polyangiitis (GPA) associated with Rathke's cleft cyst (RCC). The patient was referred to our Neurosurgery Department presenting right eye amaurosis, third cranial nerve palsy, and left temporal hemianopsia. Magnetic resonance imaging documented a sellar or suprasellar lesion with solid and cystic components. The dura mater of the skull base was also strongly enhanced. The patient underwent surgery. Histologic examination revealed RCC associated with pituitary GPA. To our knowledge, this is the first reported case of concomitant pituitary GPA and RCC. Pituitary involvement in GPA is rare, usually diagnosed in hormonal dysfunctions. The patient in case first presented optic chiasm compression, probably due to inflammation of both the pituitary gland and the previously asymptomatic RCC. We focus on the symptoms that led us to diagnose GPA pituitary involvement and on the peculiar and unusual Magnetic resonance imaging of the case presented.

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Case report

In September 2012, a 48-year-old woman started complaining of headache associated with central scotoma in her right eye. Ophthalmological assessment documented right papilledema. Her general physician ordered a magnetic resonance imaging (MRI) of brain and corticosteroid therapy. Her vision recovered after a few days.

Brain MRI documented a sellar or suprasellar lesion causing compression of the optic chiasm. The lesion was round and showed solid and cystic components. Its solid part (inferior and anterior sellar contents) was enhanced after gadolinium injection, whereas its posterior part contained fluid (isointense). The medial dura lining of the right cavernous sinus was thickened and clearly enhanced. The pituitary stalk was swollen and enhanced, whereas gadolinium staining of the gland was irregular and faint (Fig. 2A, *arrow*). The sphenoid sinus showed no pneumatization (conchal variant according to Hamberger's classification [1]). The dura mater of the spheno-ethmoidal plate, diaphragma sellae, clivus, and mesial part of both middle cranial fossas was strongly enhanced after gadolinium injection. Bilateral upper pharynx mucosa was also enhanced (Figs. 1 and 2). Panhypopituitarism was established after basal hormonal evaluation.

A few weeks later, the patient developed right eye amaurosis, diplopia, right eyelid ptosis, and visual field loss, and was admitted to the Neurosurgical Unit. Neurologic examination revealed right third nerve palsy, right eye amaurosis, and left temporal hemianopia. The patient underwent surgery using an

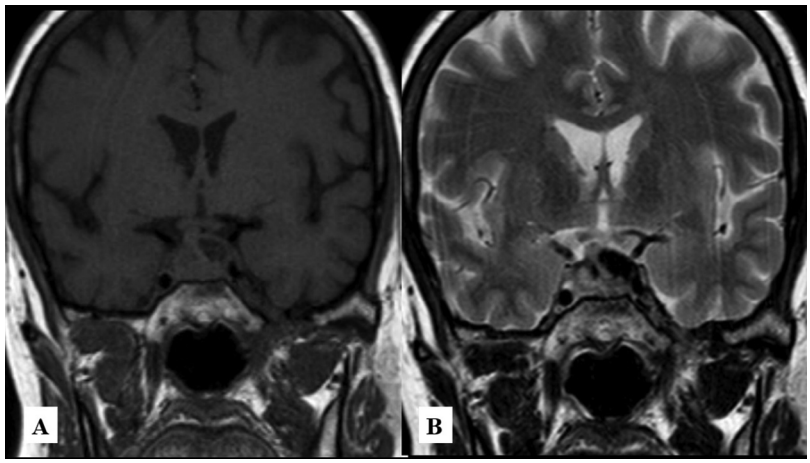

Fig. 1 – (A) Magnetic resonance imaging (MRI) T1 coronal section documenting the sellar or suprasellar lesion. The cystic fluid component was hypointense, whereas the solid part, the pituitary stalk, and gland were isointense to gray matter. (B) MRI T2 coronal section revealing the strongly hypointense fluid contents of the lesion.

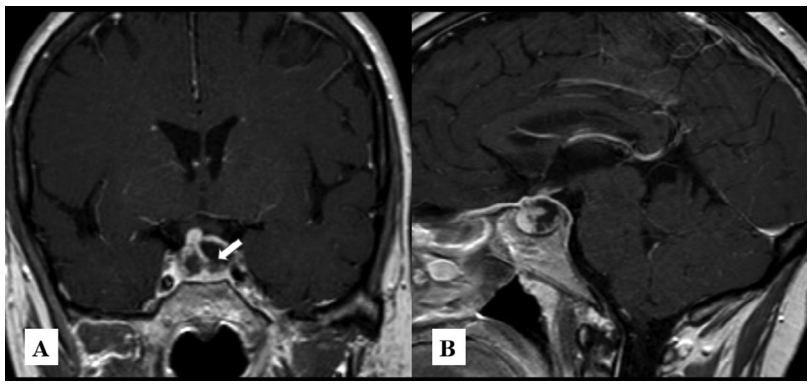

Fig. 2 – (A) Magnetic resonance imaging (MRI) T1 coronal section after gadolinium injection showing a sellar or suprasellar lesion with solid and cystic components. The pituitary stalk was swollen and enhanced, whereas the gland was not enlarged; gland staining after gadolinium injection was faint and inhomogeneous (*arrow*), both atypical features of hypophysitis. (B) MRI T1 sagittal section after gadolinium injection. The sphenoid sinus showed no pneumatization (conchal variant). The solid part of the lesion was clearly enhanced (inferior and anterior sellar contents) whereas its posterior part confirmed its fluid content (isointense). Intense enhancement involved the dura mater of planum sphenoidalis, diaphragma sellae, clivus and the mucosa of upper pharynx too. The lesion did not present typical MRI features of RCC (hyperintensity in T2 sequences, intracystic enhanced nodule and "claw-sign") or of hypophysitis—except for enhancement of the stalk—since the gland was not enlarged and its contrast enhancement was slight/faint.