

Clinical course of primary empty sella in children: a ten-year single center experience

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INTRODUCTION

Empty sella is a neuroradiological and anatomical condition, characterized by intrasellar extension of the subarachnoid space with subsequent flattening of the pituitary gland (1).

PES is commonly not associated with any signs or symptoms; however, headache, obesity, menstrual irregularities, and galactorrhea can be present. Various studies, mostly conducted in adults, evaluated the hormonal axis in primary empty sella (PES), and reported different forms of pituitary deficiencies (1-4).

AIM

We report our 10-year experience of pediatric cases with PES, investigating the pituitary function, associated impairments, and responses to treatment.

METHOD

We reviewed 10,560 cranial and 325 pituitary magnetic resonance imagings (MRIs) performed in our university hospital between January 2010 and December 2020 and identified cases with PES.

Patients with additional abnormal MRI findings, history of cranial surgery or radiotherapy, autoimmunity, long-term use of chemotherapeutic or immune-suppressive agents or incomplete diagnostic evaluation were excluded.

Clinical, radiological and laboratory evaluations were recorded. The results are reported as median (interquartile range, minimum-maximum).

RESULTS

The study included 17 patients [9 girls, 8 boys; median age 12.4 years (7.25, 4.3-17)]. The median size of the pituitary was 2 mm (0.7, 1.2-3).

Based on age-dependent pituitary height measurements, fifteen (88%) subjects showed pituitary gland hypoplasia.

Five cases presented due to short stature, two had both pubertal delay and short stature, and one had pubertal delay. Nine cases had presented with neurological symptoms including headache, tinnitus, tics, and dizziness.

Five of the seven short subjects had growth hormone deficiency. The patients with pubertal delay showed normal pubertal responses to GnRH test. Six of the patients had a baseline cortisol level below 10 mcg/dL, but all showed normal cortisol responses to stimulation tests. None of the patients had hyper- or hypoprolactinemia, adrenal insufficiency, hypothyroidism, or diabetes insipidus.

There was no statistically significant association between the size of the pituitary gland and the severity of hypopituitarism ($p=0.42$).

CONCLUSIONS

PES may be associated with different neurological and endocrine conditions (mostly, short stature and transient delayed puberty), which may require specific treatments.

Size of the pituitary gland does not relate to the severity of the symptoms.

The high incidence of pituitary dysfunctions ascertain that this entity should not be considered as a normal variant but, instead, should be evaluated carefully with appropriate basal and dynamic hormone testing.

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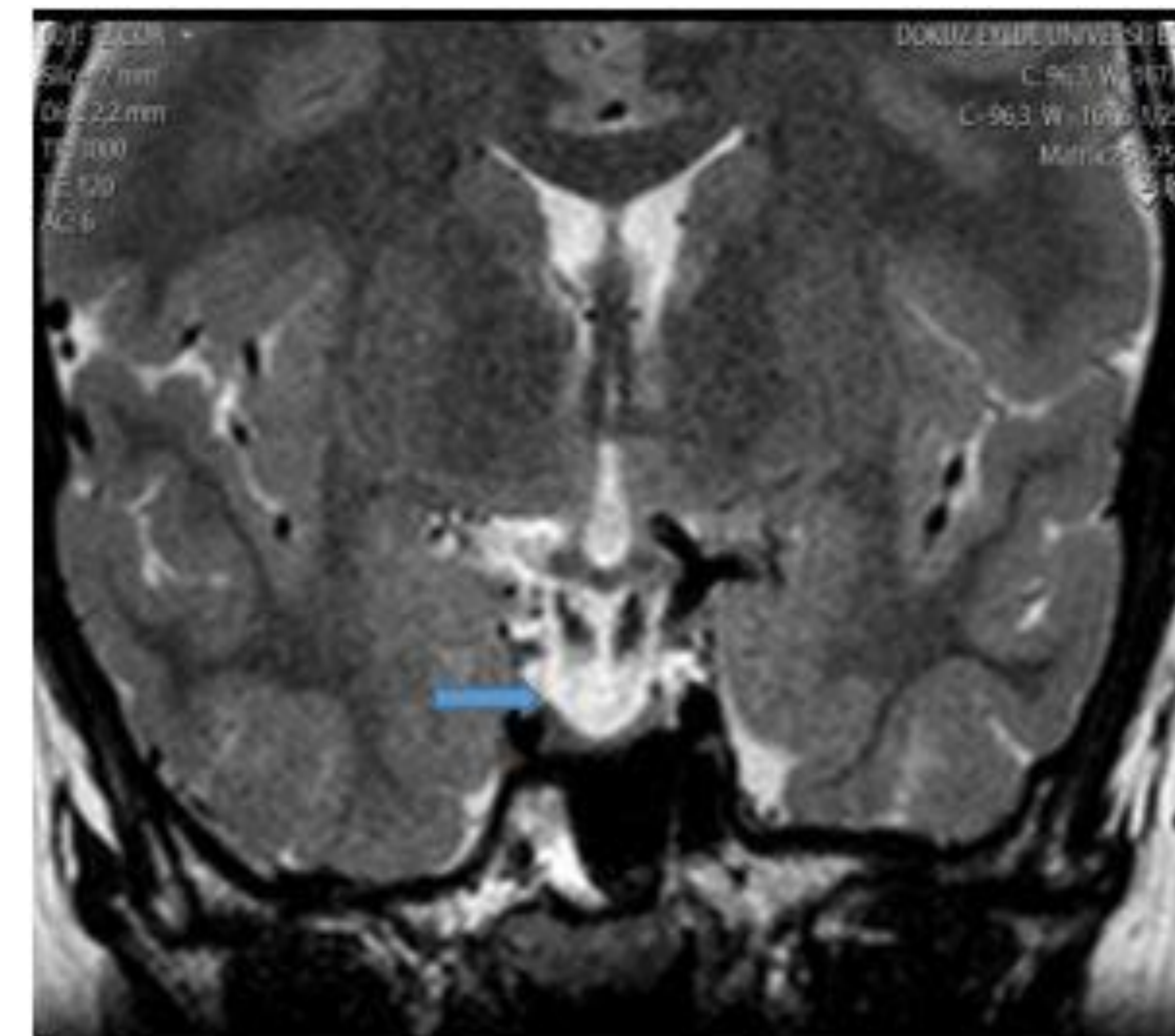
Approval was granted by the Ethics Committee of Dokuz Eylül University Faculty of Medicine (Ethics approval number: 2021/14-2906052021).

CONTACT INFORMATION

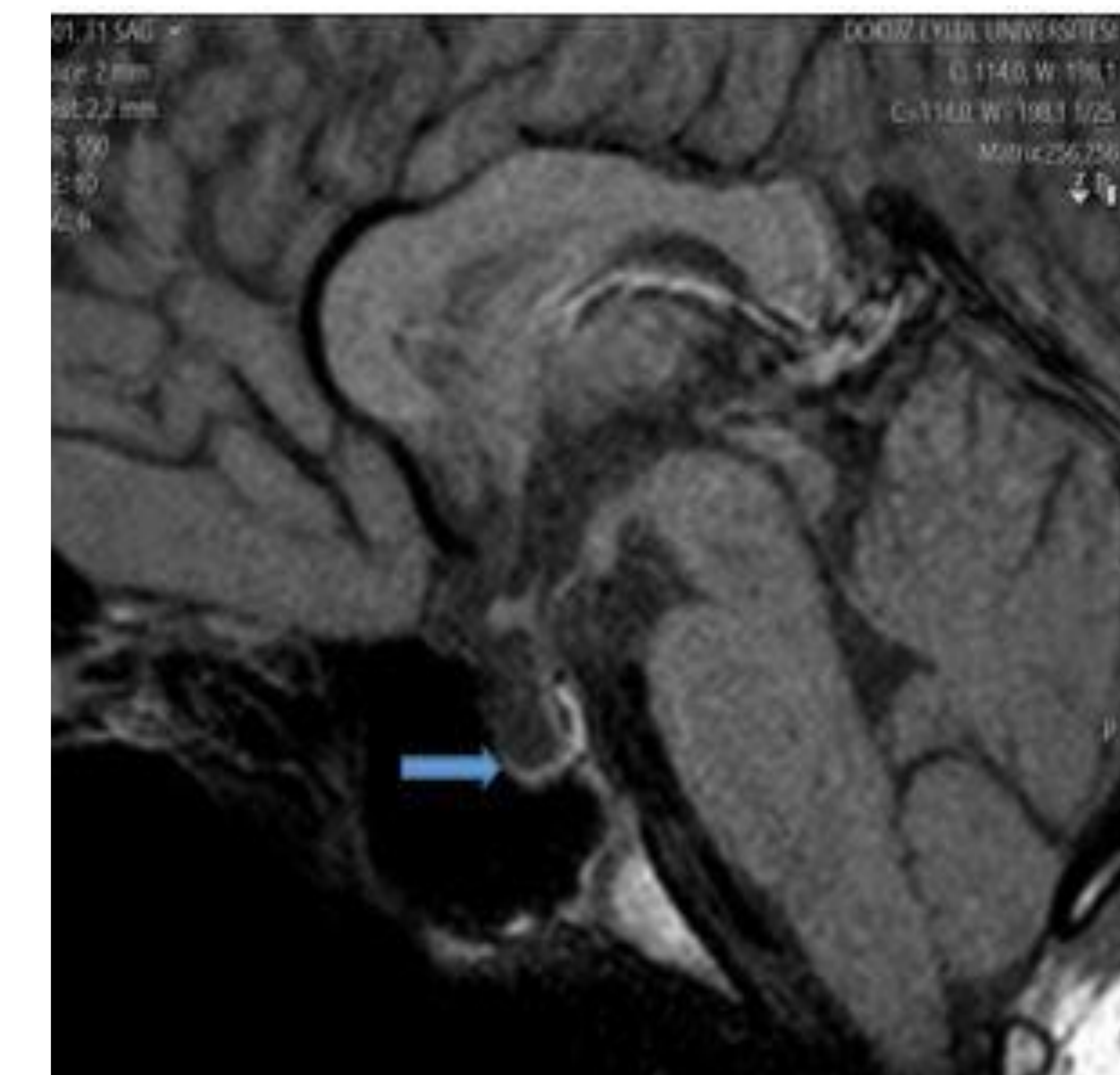
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Coronal T2-weighted (arrow: fluid attenuated) MR image shows the increased signal intensity of the cerebrospinal fluid occupying the sella turcica



Mid sagittal T1-weighted MR image that demonstrates a fluid-filled sella turcica with a flattened pituitary (arrow) at the base of the pituitary fossa.