

A CASE OF PARTIALLY EMPTY SELLA WITH HYPOPITUITARISM IN A CHILD

R PRATAP ^[1] L MASON ^[1] A ROMSAUEROVA ^[2] and S BASU ^[1]

1. Department of Paediatric Endocrinology, Oxford University Hospitals, Oxford.
2. Department of Paediatric Neuroradiology, Oxford University Hospitals, Oxford

INTRODUCTION

Empty Sella (ES) is a radiological finding which is due to herniation of subarachnoid space into sella turcica. It's partial, when less than 50% space is filled with cerebrospinal fluid (CSF) and complete, when CSF fills more than 50% with pituitary gland flattened to 2mm or less. Actual occurrence rate is debatable and often noted incidentally during magnetic resonance imaging (MRI).^{[1],[2]} However it can cause pituitary hormone (PH) dysfunction, most commonly growth hormone (GH) deficiency. We are reporting a case of partially empty sella with GH and cortisol deficiencies.

CASE SUMMARY

A 15year-old boy presented with delayed puberty and absence of secondary sexual characteristics (G1P1TV 2mL & 3mL). He had short stature. His height (153.2 cm) was below parental target(between 25th to 98th centile), on the 2nd centile and weight (58.05kg) on 50th centile. Mother felt he had low stamina during sports and took longer to recover from general illnesses. However, he did not have early morning tiredness. His birth and past medical history were insignificant. Parents were non-consanguineous and late developers. Mum needed oestrogen for pubertal induction.

His baseline tests were normal including thyroid function, coeliac, full blood count and IGF-1. His bone age was 2.66 years delayed. He had a normal male karyotype. In view of delayed puberty, he had a primed insulin tolerance test (ITT). It detected a poor GH peak of 1.7mcg/L and sub-optimum cortisol peak of 293nmol/L (acceptable at least 6.7mcg/L and 430nmol/L respectively). ACTH was within normal range (16.3ng/L, 0-46ng/L).

After diagnosing GH deficiency and secondary cortisol insufficiency, he was started on growth hormone and hydrocortisone (HC) replacement.

His T2 weighted MRI pituitary revealed partially empty sella (pituitary gland measuring 4mm cranio-caudally on right and 2mm on left). He responded well to GH and HC replacement. Eventually he progressed to puberty on his own (G2P2TV 5mL & 6mL)

DISCUSSION

ES is a rare entity in the paediatric population and probably less reported. In children it commonly present with hypothalamo-pituitary dysfunction, diabetes insipidus, optic atrophy, growth hormone deficiency, visual symptoms and cerebrospinal fluid (CSF) rhinorrhea. Although there is female preponderance in adults, in children radiological incidence of primary ES was reported as 1–48% with a male: female ratio of 1.4:1.0.^[3]

Primary ES can be idiopathic, developmental, due to increased intracranial pressure etc. Secondary ES is commonly due to injury to the gland from surgery, tumour and trauma. Presentation varies from incidental radiological findings to symptoms of hormone deficiencies. PH deficiency can be in up to 60% cases of ES with GH being most affected. Treatment consists of hormone replacement, close monitoring of evolving deficiencies and treatment of the secondary causes if present.^[4]

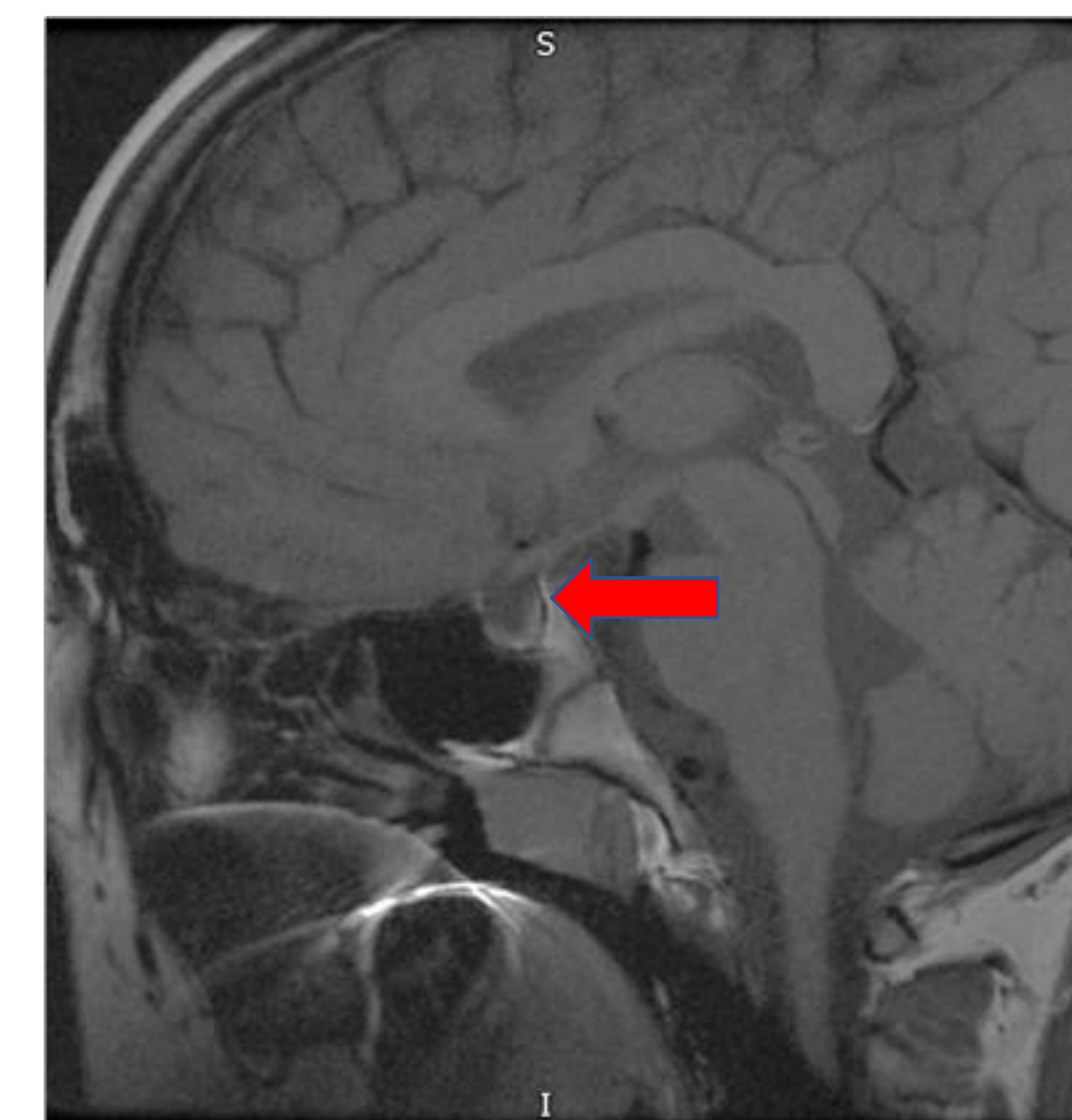


Fig 1

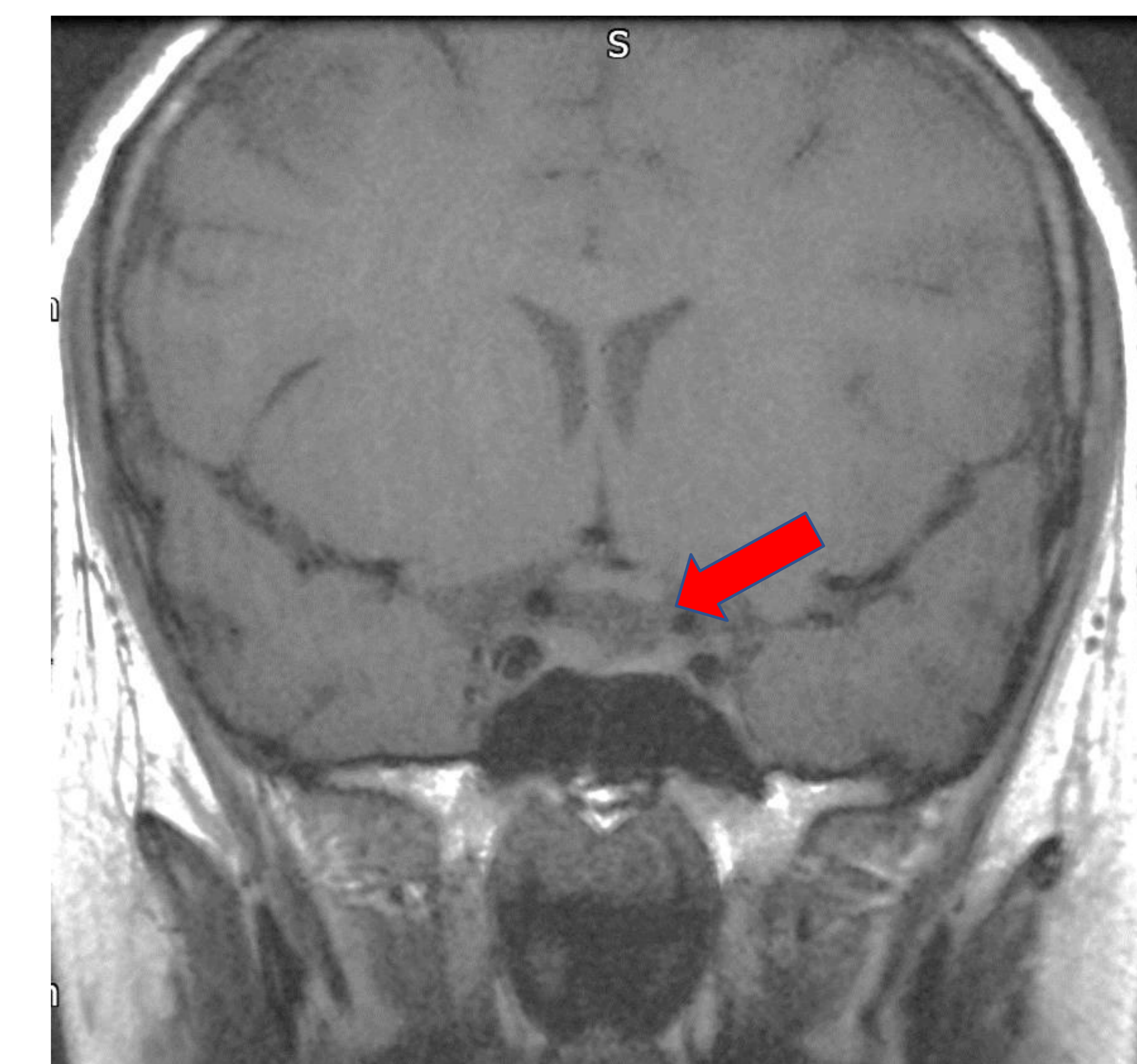


Fig 2

Fig 1 & 2 : There is a partially empty sella with asymmetric soft tissue, which is isointense on T1 and T2 lining the floor of the sella, predominantly within the right side. The infundibulum is not deviated, and the posterior pituitary bright spot is maintained. No parasellar abnormality, with normal appearances of the optic apparatus and no mass in the region of the cavernous sinuses. No hypothalamic mass is demonstrated.

TAKE HOME MESSAGE

Varying extent of hormonal abnormality may be associated with PES. Hence early assessment and management is needed. In cases with incidental finding, the need for close monitoring and follow up is of prime importance and should be practiced.

REFERENCES

1. De Marinis L, Bonadonna S, Bianchi A, Giulio M, Gustina A. Extensive clinical experience: primary empty sella. *Journal of Clinical Endocrinology and Metabolism*. 2005;5471–5477.
2. Chiloiro S, Giampietro A., Bianchi A, Tartaglione T, Capobianco A, Anile C, De Marinis L. Primary empty sella: a comprehensive review. *European Journal of Endocrinology*. 2017;177(6):R275–R285.
3. Debasmita Rath, Ranjan Kumar Sahoo,1 Jasashree Choudhury, Dillip Kumar Dash, and Anusandana Mohapatra. Empty sella syndrome in a male child with failure to thrive. *J Pediatr Neurosci*. 2015 Jan-Mar; 10(1): 45–47.
4. Miljic D, Pekic S, Popovic V. Empty Sella. [Updated 2018 Oct 1]. In: Feingold KR, Anawalt B, Boyce A, et al., editors. *Endotext* [Internet]. South Dartmouth (MA): MDText.com, Inc.; 2000

ACKNOWLEDGEMENT

Thanks to the department of Neuroradiology, John Radcliffe Hospital, Oxford for explaining and sharing MRI images for the poster.

