Congenital craniopharyngioma - A rare case of congenital hypopituitarism.

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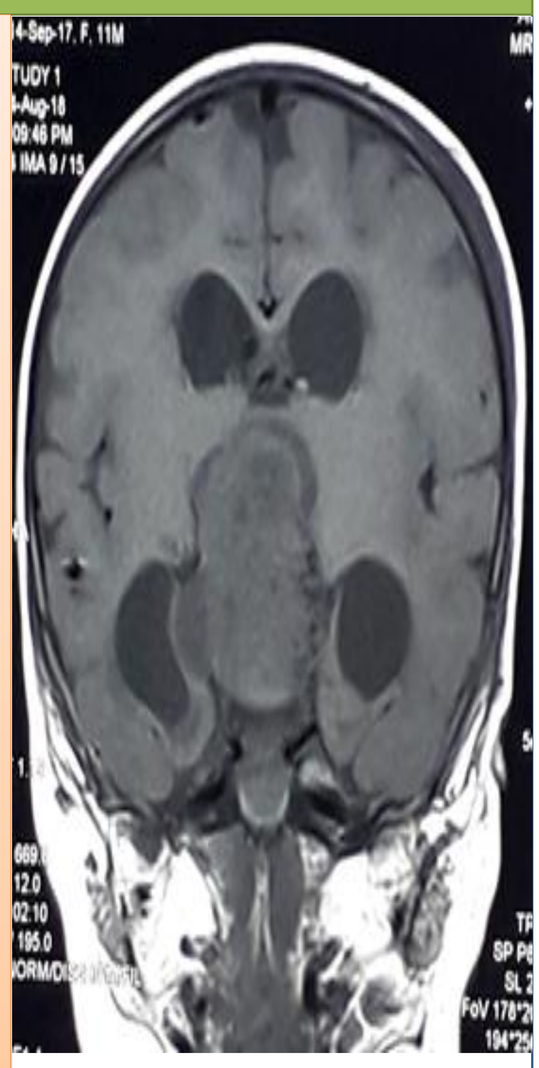
Introduction

- Hypopituitarism leads to one or more pituitary hormones deficiency.
- Hypopituitarism can be congenital or acquired.
- The incidence of congenital hypopituitarism is between 1 in 4000 and 1in 10 000 live births.
- Adamantinomatous type craniopharingioma is a relatively rare childhood tumor with the prevalence of 6%- 10% of all intracranial tumours.
- Congenital craniopharingioma is a very rare suprasella brain tumour which constitute around 0.5–1.5% of all congenital tumours.
- We report the first case of congenital craniopharingioma in Sri Lanka presenting with hypopituitarism.

Case report

- Term baby presented with micro phallus with bilateral hypoplastic scrotum and atrophic testis.
- Managed as hypopituitarism and planned MRI brain at one year
- Baby presented at the age of eleven months with bulging fontanel suggestive of increase intra cranial hypertension.
- MRI brain at that time revealed solid and cystic mass involving the sella and suprasella region with obstructive hydrocephalus compatible with adamantinomatous type craniopharingioma.



Investigations

		Initial Evaluation	Follow up
LH		<0.07IU/L	
FSH		0.07IU/L	
Basal cortisol		<22nmol/L	
SST	0min	<22	
	30min	78.7	
Free T4 (9-25)		10.10pmol/L	7.2
TSH (1.82-8.21)		3.22mIU/L	1.98

Management

- He underwent excision of the craniopharingioma.
- Post op needed multiple pituitary hormones replacement(hydrocortisone, thyroxine and desmopressin).
- Post operatively he had developmental deterioration and visual impairment .
- Radiotherapy was not given due small age.

Conclusion

- This case highlights the very rare congenital cause of hypopituitarism.
- Antenatal and early postnatal imaging with the ultra sound brain would help in the early diagnosis.
- Total resection is the treatment modality. Even with improving neurosurgical technology, prognosis is poor due to the age, size and location of the tumour.