**CASE REPORT**

**A 7-Month-Old Infant with Cushing’s Disease**

Rabia Abbas, Mohammad Haroon Hamid, Mohammad Sarwar, Taeeed A. Butt, Abid Qureshi and Nadeem Malik

**ABSTRACT**

Cushing's disease in children is not rare but in infants it is quite rare and an important medical condition needing proper line of investigations and management options. Craniopharyngioma as a cause of Cushing's disease is well reported and practical inference of the condition is of clinical importance. Craniopharyngioma generally affects children at 5 – 10 years of age and is rarely seen in infancy. It usually manifests as endocrinological deficits such as short stature, delayed puberty, and obesity. We report the case of a 7 months old infant who presented with obesity and Cushing’s disease associated with craniopharyngioma.

**Key Words:** Craniopharyngioma. Cushing’s disease. Obesity. Infancy.

**INTRODUCTION**

Cushing's syndrome in paediatric patients is usually caused by adrenal tumours. Pituitary tumours causing Cushing's disease are extremely rare under the age of 7 years and more so in infancy. Craniopharyngioma are benign, partly cystic neuro-epithelial tumours of the central nervous system that are relatively rare, representing 6-10% of intracranial malignancies in children and adolescents (approximately 2 – 3 cases per 1,000,000 children). Craniopharyngiomas are often located near the optic nerves and pituitary gland. Symptoms include vision changes, headache, weight gain and endocrine changes. Overall prognosis is good due to benign histopathology, but high recurrence rate after treatment and complications of curative therapy can cause significant morbidity in survivors. An improvement in its prognosis will require earlier diagnosis and the development of improved neurosurgical and radio therapeutic treatment strategies.

The report describes an uncommon case of an infant with Cushing’s disease due to craniopharyngioma.

**CASE REPORT**

A 7-month-old female infant presented with generalized obesity since one month of age. She had no symptoms of visual attention, vomiting, seizures or enlargement of head. On examination, the child was 77 cms long (97th centile standardized growth charts) and weighed 10.5 kg (above 97th centile).

On examination, she had generalized obesity, blood pressure was 121/101 mm of Hg, moon like facies and gynaeacomastia (Figure 1). The fundoscopic examination was normal. Investigations showed a raised midnight adrenocorticotropic hormone (ACTH, 196 pg/ml, normal = less than 11 pmol/L and plasma cortisol; the latter's values being 39.9 mcg/dL (normal = 5 – 23 mcg/dL) and 46.1 mcg/dL (normal = 3 – 16 mcg/dL) taken at 8.00 a.m. and 4.00 p.m. respectively. Her thyroid function tests, basal prolactin concentration, LH and FSH were normal. Postprandial glucose level was 79 mg%. Serum sodium and potassium were 141 meq/L and 6.5 meq/L respectively. Ultrasound scan of abdomen was normal. MRI scan of the brain showed a complex partly solid and cystic calcified mass in supra and parasellar location measuring 4.2 x 2 cm adjacent to the antero-inferior wall of third ventricle, compressing the optic chiasma and circle of willis. Inferiorly the lesion buldges into the sella and is in close approximation to cavernous sinus to which it was inseparable elevating the optic chiasma. (Figure 2). The report describes an uncommon case of an infant with Cushing’s disease due to craniopharyngioma.

Figure 1: Cushingoid facies of seven months old infant.

Figure 2: An MRI of the brain showing calcified mass in supra and parasellar location measuring 4.2 x 2 cm adjacent to the antero-inferior wall of third ventricle, compressing the optic chiasma and circle of willis.
She was managed with careful monitoring in Paediatric ICU care but succumbed to septicemia and multi-organ failure.

**DISCUSSION**

Cushing syndrome in childhood is most often iatrogenic caused by excessive glucocorticoid hormone administration. Endogenous Cushing syndrome in children is rare in infancy and less than 100 cases have been described worldwide. Cushing disease is caused by excessive secretion of ACTH which occurs due to abnormal pituitary function, ectopic corticotrophin-secreting tumours, or by autonomous secretion of cortisol by an adrenal tumour or bilateral micro-nodular adrenal hyperplasia. It usually presents with delayed growth and development. Cushing disease is exceedingly rare in children, especially in those under the age of 2 years of age but accounts for approximately 75% of all Cushing syndromes in children older than 7 years of age.

Craniopharyngiomas commonly present with symptoms of raised intracranial pressure due to compression of adjacent neural structures, obstructing cerebral spinal fluid causing hydrocephalus, headaches, and projectile vomiting. In more than half of patients, there is endocrinological dysfunction and patients usually present with short stature, delayed or arrested puberty, obesity, or diabetes insipidus. Other manifestations include visual impairment (62 – 84%), endocrine deficits affecting the secretion of growth hormone (75%), gonadotropins (40%), ACTH (25%), and thyroid-stimulating hormone (25%). Central diabetes insipidus is found in 17%.

On neuroimaging, a craniopharyngioma usually appears as a cystic tumour with calcification which is found in approximately 90% of tumours. CT may be better at revealing calcification; the most common site is suprasellar location (80%).

Trans-sphenoidal surgery remains the treatment of choice. Pituitary irradiation should be considered for those patients whose tumours cannot be totally removed. For unfavourably localized tumours, there is controversy over total resection or a limited resection (biopsy, partial/sub-total resection).

In radical resection, there is risk of surgically induced deficits (mainly hypothalamic) and the high rate of recurrence (23%) in infants and small children despite complete resection. After subtotal resection, the tumour remnant continues to grow in 71% of patients. The rate of progression after sub-total resection followed by radiotherapy is 21%.

The prognosis for patients with craniopharyngioma is generally favourable, with an 80 – 90% chance of permanent cure if the tumour can be completely removed with surgery or treated with high doses of radiation. Careful follow-up of these patients with awareness of the possibilities of relapse and the complications of hypopituitarism is indicated. Most recurrences occur within the first 2 years after surgery. A significant percentage of patients have long-term hormonal, visual, and neurological problems following the treatment of craniopharyngioma.

This patient had an aggressive course with unfavourable outcome likely to be due to the occurrence in infancy and the extensions of the lesion which precluded removal and control.

**REFERENCES**