

## Case Report

# Secondary narcolepsy in a child after resection of a craniopharyngioma

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### ABSTRACT

Narcolepsy is a neurological disorder of sleep characterised by excessive daytime sleepiness and has been reported following the removal of a craniopharyngioma due to a host of neurological and endocrinological causative factors. We present here a case of a child that developed secondary narcolepsy following resection of a craniopharyngioma.

**Keywords:** Narcolepsy, Sleep, Craniopharyngioma

### INTRODUCTION

Narcolepsy is a sleep disorder seen in both adults and children and characterised by excessive daytime sleepiness, cataplexy, hypnagogic hallucinations and sleep paralysis.<sup>1</sup> It is now proven that narcolepsy is caused by pathology in the orexin (hypocretin) system which is a peptide system in the brain known to maintain the sleep wake cycle.<sup>2</sup> Craniopharyngiomas are tumors of embryonic malformations that are low grade histological malignancies arising out of ectoblastic remnants of the Rathke's pouch and is known to occur along the hypothalamic and pituitary tissue.<sup>3</sup> They make up 2-10% of pediatric space occupying lesions and 35-55% cases are seen in children between the age of 4-12 years.<sup>4</sup> The effects of craniopharyngioma removal on sleep has been studied and attributed to a host of factors like its effects on neurobiology via resection as well hormonal changes caused by the removal of the tumor.<sup>5,6</sup> The surgery for craniopharyngioma may lead to some potential damage to neighbouring hypothalamic structures as nuclei responsible for sleep control.<sup>7</sup> These cases have been labelled as secondary narcolepsy as narcolepsy comes up after the procedure and is not present prior to the surgery.<sup>8</sup>

We hereby present a case of narcolepsy that developed in a 10 year child after removal of the craniopharyngioma and discuss the pathophysiology involved in such cases.

### CASE REPORT

A 10 year old boy studying in 5<sup>th</sup> standard was referred to the psychiatry department for an evaluation, in view of altered behavior in the form of episodic sleepiness during the day time and not studying well since last 1 month prior to presentation. The child while sitting would experience attacks of sleepiness which would be followed by loss of tone and falls. The sleepiness episodes would last for 10 to 15 minutes at a time. This would happen multiple times in a day and would be precipitated by intense emotions or sometimes had no precipitating factor at all. Gradually his symptoms had worsened and this affected his performance at school. His teachers who were unaware of the problem often pulled him up in class for sleeping during class hours and a complaint had reached the parents as well. He suffered from multiple bruises owing to the falls and his parents would fear that he may get seriously injured if this continued. The child was unaware of these falls and would cry after being injured following the fall but was unable to explain how he sustained the injury.

On history the parents revealed that about a month back, the child had experienced severe headache with repeated bouts of forceful vomiting. The picture got complicated when he developed seizures which were of a generalized tonic clonic type. The child was admitted to the pediatric department where neuro-imaging (CT) revealed presence of a space occupying lesion in the pituitary region which was suspected to be a craniopharyngioma. During the procedure of transnasal resection, the child was found to have a tumour in the suprasellar region which was diagnosed to be craniopharyngioma after histopathological examination.

Post operatively, the child was alright around till a month prior to presentation when he started experiencing aforementioned symptoms with additional symptoms suggestive of pan hypopituitarism. The child was not cooperative for a polysomnography evaluation which hence could not be performed. The only symptom in his case was excessive daytime sleepiness with multiple short naps. No hallucinations or sleep paralysis was reported on history. No family history of narcolepsy existed. Based on the symptom profile he was diagnosed to have secondary narcolepsy clinically and was started on Tab. Modafinil 100 mg in the morning time and showed improvement on subsequent follow up. This was then increased to 200 mg per day in daytime divided doses. The child was also on steroid supplements and thyroid hormones in view of the pan hypopituitary symptoms. The patient was also on carbamazepine in a total dose of 400 mg per day in view of the seizure episode in the past.

## DISCUSSION

Judging from the clinical course of the patient, a diagnosis of secondary narcolepsy caused by craniopharyngioma resection fitted as a diagnosis. In a huge case series of such patients author have reported that patients may have narcolepsy symptoms even before surgery due to the tumor itself while there are also reports that attribute tumor removal to be a secondary cause of the narcolepsy.<sup>9,10</sup> Facilities for HLA-DRB1 or HLA-DQB1 testing or CSF orexin levels are not available in our center hence was not done. Another reason for avoiding excessive sleep based and genetic investigation in this case was non-affordability on the part of the patient. We diagnosed the case clinically and with neuroimaging (Figure 1 and 2). Wakefulness generating neurons include orexinergic and histaminergic cells as well as doapmine and serotonergic neurons. These project from the hypothalamic nuclei to the frontal lobes of the brain and this connections are often disrupted during the resection of a craniopharyngioma.<sup>11</sup> This coupled with disruptions in melatonin secretion via the pituitary have been implicated in the pathogenesis of secondary narcolepsy in these patients.<sup>12</sup> Craniopharyngiomas are commonly seen in childhood and neurosurgeons involved in removal of these tumors must be aware of the complications of resection so that parents are forewarned

in the beginning and so that these symptoms are examined on follow up.

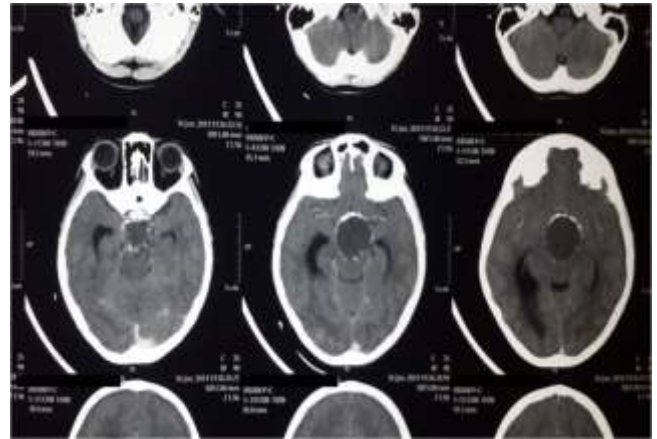


Figure 1: CT brain preoperative.

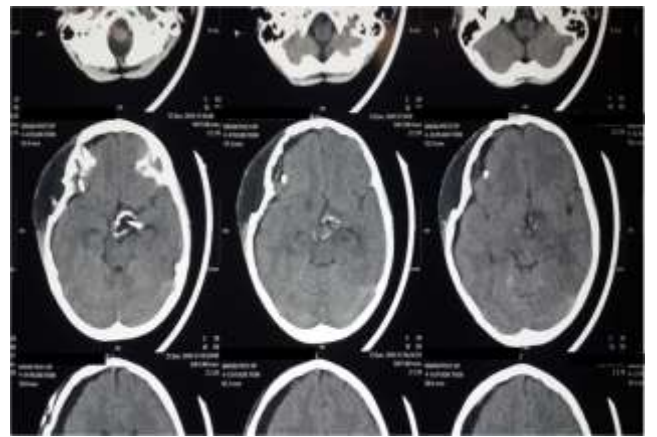


Figure 2: CT Brain postoperative.

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