ANESTHESIA FOR NELSON'S SYNDROME

- Case Report -

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Introduction

Adrenalectomy in the setting of residual corticotrope adenoma tissue predisposes to the development of Nelson's syndrome[1]; a disorder characterized by rapid pituitary tumour enlargement and increased pigmentation secondary to high ACTH levels. We present the perioperative course of a child with Nelson's syndrome who underwent sublabial trans-sphenoidal hypophysectomy.

Case Report

A 12 yr-old, 30 kg female child was admitted with history of bilateral adrenalectomy for Cushing's syndrome 3 years back, following which she developed severe darkening of skin all over body and gradually progressing headache. Her endocrine profile revealed ACTH levels of 220 pg/mL (Normal = 6-76 pg/mL) with cortisol of 1.2 μg/dL (normal = 5-25 μg/dL) for which prednisolone 5 mg and fludrocortisone 50 μg daily were being supplemented. Growth hormone levels were increased (4 ng/mL; normal <2 ng/mL). Well defined hypointense mass (9 × 3 mm) arising from anterior pituitary was evident in CT scan. A diagnosis of Nelson's syndrome was made and sublabial transsphenoidal hypophysectomy was planned. Child was rendered euthyroid on eltroxin 50 μg daily. Airway examination revealed coarse facies with Mallampati II airway.

Anesthesia was induced with propofol and fentanyl. Tracheal intubation was facilitated with rocuronium. Maintainanence of anesthesia was done with isoflurane and N_2O in 40% O_2 and intermittent boluses of rocuronium and fentanyl. At end of procedure, neuromuscular blockade was reversed and trachea extubated. Postoperative course was uneventful. Perioperative steroid coverage comprised hydrocortisone 50 mg 8 hourly, started 1 day preoperatively till postoperative day 1, when oral prednisolone 25 mg daily in divided doses was started, subsequently tapered over next 2 days to scheduled preoperative dose. Normal skin colour was restored in 1-2 days, probably indicating successful removal of microadenoma.

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Discussion

The incidence of Nelson's syndrome has been variably reported to be between 8-38%[2-4]. Younger age and pregnancy are possible associated risk factors. Hyperpigmentation of skin, minimal sellar changes or raised ACTH though suggestive, need neuroradiological confirmation for definite diagnosis of Nelson's syndrome. The predominant cause of morbidity is from local tumor extension or invasion. Patients with this disorder become deeply pigmented because of excess alpha melanocyte stimulating hormone (a-MSH), a derivative of propiomelanocortin (POMC), the precursor peptide for ACTH. There may be loss of pituitary function because of compression or replacement of normal pituitary tissue or compression of structures adjacent to the pituitary fossa by the tumor. Lateral extension of the tumor may result in invasion of the cavernous sinuses and entrapment or compression of the cranial nerves (III, IV, V and VI). Superior extension of the tumor can lead to compression or invasion of optic apparatus and or the hypothalamus. Headaches are common, are probably due to stretching of the dura of the diaphragma sellae by the tumor. Pituitary adenomas exhibit widely variable endocrine profile. Panhypopituitarism rather than hypercortisolism is characteristic by virtue of compression of normal anterior pituitary by the tumour. Cortisol deficiency is likely particularly in setting of bilateral adrenalectomy. Perioperative steroid coverage is indicated in patients with low cortisol levels to prevent acute adrenal insufficiency. Though clear guidelines for perioperative glucocorticoid replacement in pituitary surgery exist[5], we found no such specific recommendations for Nelson's syndrome. We feel that patients with Nelson's syndrome may particularly need perioperative steroid replacement in view of their compromised adrenal functions, especially when cortisol levels are less than 3.6 μg/dL. Patients with Nelson's syndrome may be asymptomatic for long periods with only presentation being hyperpigmentation of skin. Hyperpigmentation in addition to thick, oily and hyperhydrotic skin may pose problems in securing intravenous line and tracheal tube. Though our patient had increased growth hormones and coarse facies, we did not encounter any acromegalic features of the airway. Recovery of normal skin colour may be an early indicator of successful surgery; however the results need to be confirmed by regular clinical and neuroradiological follow ups.

References

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