ANESTHESIA FOR NELSON’S SYNDROME

- Case Report -

MADHUR MEHTA*, GIRIJA P RATH**, AND GYANINDER PAL SINGH*

Introduction

Adrenalectomy in the setting of residual corticotrope adenoma tissue predisposes to the development of Nelson’s syndrome; 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 N₂O in 40% O₂ 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.

From Department of Neuroanaesthesiology, All India Institute of Medial Sciences, New Delhi, India.

* MD, Senior Resident.
* MD, Assist. Prof.

Correspondence: Dr. Girija Prasad Rath, Assist. Prof., Department of Neuroanaesthesiology, Neurosciences Centre, All India Institute of Medical Sciences, New Delhi-110029, India. Tel: 91-11-26588700-3474, Fax: 91-11-26586663.

E-mail: girijarath@yahoo.co.in
Discussion

The incidence of Nelson’s syndrome has been variably reported to be between 8-38%\(^2\). 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 (α-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.

References