Nelson Syndrome

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Risk

- Incidence: Reported in 8–44% of pts following bilateral adrenalectomy for Cushing disease
- More likely in pts with younger age and pregnancy

Perioperative Risks

- Lyte imbalances
- DI
- Vision loss
- Challenges specific to type of surgery

Worry About

- Panhypopituitarism
- Volume status imbalance
- Steroid supplementation

Overview

- The first case of Nelson syndrome was reported in 1958 by Dr. Del Nelson, who named this condition.
- It is also known as post-adrenalectomy syndrome and occurs as result of bilateral adrenalectomy performed for treatment of Cushing disease.
- It can develop as long as 24 y after a bilateral adrenalectomy, but the mean age of presentation is 15 y after the adrenalectomy.
- Nelson syndrome differs from Cushing disease in that the hypercortisolism cannot occur because of the adrenalectomy, and a pituitary tumor is known to be present.
- The pathophysiology of Nelson syndrome is poorly understood. It possibly occurs due to release of the negative feedback that would otherwise suppress high cortisol levels, in turn leading to restoration of CRH production by the hypothalamus going on to stimulate corticotroph neoplasia.
- The signs and symptoms of Nelson syndrome are due to the effects of raised ACTH (more than 154 pmol/L) and the pressure of the tumor on surrounding structures, inhibiting release of other pituitary hormones, and thereby leading to panhypopituitarism. The symptoms include hyperpigmentation, headache, and visual disturbances. Increased urine output may suggest development of DI.
- ACTH levels are markedly elevated in Nelson syndrome and because of an exaggerated ACTH response to CRH.
- Other tests for hormones to assess panhypopituitarism may be done. Thyroid-function tests, prolactin levels, and IGF-1 IGF-BP3 measurement; measure gonadotropin levels in adolescents showing pubertal arrest and urine osmolality and specific gravity to rule out DI.
- No clear guideline is provided for periop glucocorticoid replacement, although serum cortisol values less than 3.6 µg/dL should be treated with supplementation.

Etiology

- Exact pathogenesis remains unclear.
- After a bilateral adrenalectomy is performed, cortisol levels are no longer normal, and it increases CRH production.
- The loss of partial cortisol inhibition because of the adrenalectomy allows the pituitary tumor to secrete tremendous amounts of ACTH and may promote growth of the adenoma.

Usual Treatment

- Radiotherapy
- Surgical resection of pituitary tumor, transsphenoidal or transcranial
- Pharmacologic agents along with surgery: pasireotide, temozolomide, and octreotide

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Eyes	Reduced visual acuity	Ophthalmoscopy	Visual field testing
CV	Hypovolemia, hypotension, and tachycardia		Orthostatic hypotension Volume status and BP	Steroid supplementation
GI	Hyperaldosteronism	Anorexia, N/V		Hypokalemia, hyponatremia, hypovolemia
ENDO	Increased ACTH Decreased LH and FSH Decreased TSH Decreased GH	Increased pigmentation Decreased sexual function and amenorrhea Increasing weight, cold intolerance, depression, constipation, and sleep disturbances Fatigue	Diminishing secondary sexual features Myxedema	FSH, LH levels TSH, free T_3, T_4, IGF-1
CNS	Headache			MRI (brain)
RENAL	Decreased vasopressin	Increased UO	Decreased BP and hypovolemia	Hypernatremia, decreased urine specific gravity

Key References: Barber TM, Adams E, Ansorge O, et al.: Nelson's syndrome. *Eur J Endocrinol* 163(4):495–507, 2010; Mehta M, Rath GP, Singh GP: Anaesthesia for Nelson's syndrome, *Middle East J Anaesthesiol* 20(2):313–314, 2009.

Perioperative Implications

Preoperative Preparation

- Serum lyte correction
- Fluid replacement and volume status management
- Hormonal replacement to treat panhypopituitarism
- Periop steroid supplementation
- DI management, if present

Monitoring

- Invasive arterial catheter
- Frequent ABGs and lytes

Airway

- Avoid succinylcholine and hyperkalemia.

Maintenance

- Maintenance of normocarbia for pituitary surgery
- Management of hemodynamic fluctuations during transsphenoidal surgery
- Titration of anesthetic agents to prepare for early extubation

Extubation

- Before extubation, ensure the pt is fully awake and able to maintain airway reflexes.
- Blood may be present in the stomach despite pharyngeal packing.

Adjuvants

- Esmolol and labetalol during epinephrine use during nasal packing

Anticipated Problems/Concerns

- Unstable hemodynamics
- Possible lyte changes and DI
- The possibility of difficult extubation with bleeding through the nose packing

Neurofibromatosis

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Risk

- NF-1 birth incidence: 1:3000
- NF-2 birth incidence: 1:25,000
- Schwannomatosis incidence: 1:40,000

Perioperative Risks

- Depend on tumor extent and location

Worry About

- Difficult intubation
- Intraop Htn
- Intraop tachycardia

Overview

- NF-1, formerly known as von Recklinghausen disease, is a relatively common, neurologic genetic disorder with variable clinical presentation. It involves multiple organs, such as the skin and peripheral nervous system, which serve as sites for tumors and hamartomas.
- Hallmarks include café-au-lait spots (more than 6 that are >1.5 cm in diameter), Lisch nodules (benign iris hamartomas), axillary and groin freckling, and multiple neurofibromas.
- Laryngeal and tracheal compression may occur secondary to associated tumors.