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CASE REPORT

Anesthetic Management of a Sellar Mass Excision in a Patient of Multiple Endocrine Neoplasia Type 1 Syndrome: A Rare Case

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ABSTRACT

Multiple endocrine neoplasia (MEN) type 1 is an autosomal dominant disease, commonly characterized by neoplasms of parathyroid glands, pituitary gland and pancreas islet cells. The anesthesia management in such patients for surgical procedures is challenging due to multiple endocrine gland involvement and its resultant implications, making every case a unique entity. The anesthesia management for MEN type I patients pose unique challenges to the anesthesiologist requiring meticulous preoperative evaluation, intraoperative anticipation, prevention and management of potential complications along with postoperative monitoring. We present a case of successful management of excision of a sellar mass in a patient with MEN type I syndrome with full postoperative recovery with the use of propofol, dexmedetomidine and desflurane anesthesia.

Keywords: Dexmedetomidine, Hormones, Multiple endocrine neoplasia (MEN) type 1, Pituitary.

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INTRODUCTION

Multiple endocrine neoplasia (MEN) type 1 is an autosomal dominant disease, commonly characterized by neuroendocrine neoplasms of parathyroid glands, pituitary gland, and pancreas islet cells, which may be benign or malignant. Other endocrinal and nonendocrinal expressions of MEN type I include adrenocortical, gastric, thymic or bronchial tumors, foregut carcinoids, visceral and cutaneous lipomas, and meningiomas and facial angiofibromas.[1,2] It occurs in about 1 in 30,000 individuals, with an estimated prevalence of 2 to 3 per 100,000 population.[3] Anterior pituitary tumors are found in 30 to 40% cases of MEN type 1 syndrome patients.[1,2] They are commonly prolactinomas (60%), somatotropinomas (25%), < 5% corticotropinoma or nonfunctioning.[1,2] Treatment options are medical management, surgery, or radiotherapy. They are not completely curable due to the presence of multiple endocrine tumors which are large in size and aggressive with high prevalence of metastasis.

We present the anesthetic management of a case of 45-year-old male diagnosed with MEN type 1 syndrome with a sellar mass.

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CASE REPORT

A 45-year-old male patient, known case of familial MEN type I syndrome, presented with chief complaints of generalized bodyache. In 2011, he had undergone a transthoracic thymectomy for thymic carcinoma type C, followed by 30 cycles of radiotherapy. Postoperatively, he was detected to have right diaphragmatic eventration due to phrenic nerve palsy. He was under follow-up monitoring for MEN type I and was detected to have a large sellar mass, 2.6 × 2.3 cm, on MRI brain. He had no visual complaints. He was posted for transsphenoidal excision of the sellar mass.

Further endocrine evaluation, including investigations like whole-body positron emission tomography (PET) scan, and gastroenteroendoscopy, was done. It revealed presence of bilateral lower parathyroid adenoma with hyperparathyroidism, pituitary mass, a single neuroendocrine ulcer in gastric antrum, multiple nodules in 2nd part of duodenum, and a single nodule in head of pancreas along with nonsecreting right adrenal adenoma. At the time of preanesthesia checkup, he gave a history of diabetes mellitus since 2 years for which he was taking oral metformin 500 mg and glimiperide 1 mg twice a day.

He weighted 73 kg. He was afebrile with a pulse of 120 beats/minute, blood pressure of 140/90 mm Hg, and oxygen saturation of 99% on room air. His effort tolerance was more than 2 flight of stairs. Airway examination revealed no abnormalities, with a Mallampati grade I. His chest was clear with air entry reduced in right lower zone. Neurological examination was normal with no acromegalic or cushingoid features. All other systemic examination

was normal. His ophthalmological examination was normal with no papilledema. All his routine investigations were normal. His blood sugar levels were on higher side (fasting 137 mg/dL and postprandial 153 mg/dL) for which he was advised subcutaneous insulin injections according to sliding scale 1 day prior to surgery. His electrocardiogram showed tachycardia with left-axis deviation. Two-dimensional echocardiography showed ejection fraction 60% with good biventricular function. His chest X-ray showed right diaphragmatic eventration. Serum electrolytes were done – serum calcium 10.8 mg/dL and phosphorus 1.6 mg/dL with urinary calcium-to-creatinine ratio 0.31. His hormone profile is as follows: Prolactin – 167.79 ng/mL, IGF1 – 299, cortisol – 1.17 µg/dL, and serum aldosterone – 41.7 pg/mL. His thyroid function tests were normal. Serum parathyroid level was on higher side – 180 pg/mL.

After confirming nil by mouth status and consent, standard monitoring applied included 5 lead electrocardiogram, noninvasive blood pressure, pulse oximetry, end tidal carbon dioxide, and temperature. The patient was explained that due to nasal packing in the postoperative period, he would have to breathe through the mouth. After securing a wide bore intravenous line in left hand, a Ringer's lactate at 4 mL/kg/hour was given intraoperatively.

We gave hydrocortisone 100 mg. The patient was premedicated with ranitidine 50 mg, midazolam 1 mg, dexmedetomidine 1 µg/kg over 10 minutes via infusion and fentanyl 100 µg intravenously. We induced the patient with propofol 150 mg and succinylcholine 100 mg. The patient was intubated with an 8.5 no. disposable cuffed endotracheal tube, positioned to left angle of mouth followed by pharyngeal packing to prevent aspiration. The patient was positioned with a slight head elevation to reduce venous congestion. Padding of pressure points was done. The patient was maintained on 1.2 MAC desflurane in an oxygen:nitrous mixture of 50:50%, along with dexmedetomidine infusion at a rate of 0.5 µg/kg. The intraoperative period was uneventful. Before extubation, intravenous paracetamol 1 gm was given for analgesia and ondansetron 4 mg for prevention of nausea and vomiting. The oropharynx was thoroughly suctioned after the pharyngeal pack removal. The patient was extubated on table within 8 minutes of stopping desflurane and dexmedetomidine, with full neurological recovery. In the immediate postextubation period, patient was maintaining a saturation of 89 to 92% on room air and 99% with oxygen by simple mask at 5 L/minute flow. He was shifted to intensive care unit (ICU) for observation. He was given propped up position and oxygen was continued for next 4 hours. He was monitored for diabetes insipidus postoperatively. Strict input output measurement was done. He maintained a saturation of 100% on room air overnight, pain was controlled with round the clock paracetamol injection on day 1 and oral paracetamol 500 mg as required from day 2 onward. Sugars were controlled with insulin injections on day 1 and later shifted to oral hypoglycemic drugs. He was given steroid hormone replacement with intravenous hydrocortisone 100 mg 8 hourly on day 1 and later shifted to tab prednisolone 2.5 mg daily from day 2. Daily monitoring of serum calcium levels revealed 13.3 mg% on day 9 for which he was started on tablet calcitonin 50 mg 8 hourly. One week after the patient was operated for parathyroidectomy which was also uneventful and patient was discharged from hospital on day 34. Excisional biopsy of the sellar mass revealed a thymic metastatic nodule.

DISCUSSION

Multiple endocrine neoplasia type 1 is a rare disorder, occurring in about 1 of 30,000 individuals, with an estimated prevalence of 2 to 3 per 100,000 population.[3] Incidence is found to be similar across all ages and both sexes.[1] It is an autosomal dominant disease characterized by mutations in tumor suppressor gene MEN1 (11q13 chromosome location) with a high penetrance, getting detected in >98% by the fifth decade.[1,4] It is diagnosed by presence of tumors in any two of the three endocrine glands – parathyroid, pancreas, or pituitary. There is also a significantly increased morbidity associated with MEN type I malignancies due to its manifestations in various organs with many of them not amenable to surgery. It affects multiple systems requiring a multidisciplinary team for management, which includes endocrinologist, gastroenterologist, oncologist, surgeon, anesthetist, histopathologist, radiologist, and genetecist.[1,5]

Since multiple hormonal derangements along with the accompanying disorders are common in MEN type 1 patients, a meticulous preanesthesia evaluation and preparation are essential. The hormone levels should be thoroughly investigated preoperatively. In our patient, prolactin and growth hormone levels were mildly raised, but cortisol levels were significantly decreased. This was attributed to pituitary "stalk effect" secondary to enlarged pituitary mass. Mortality related to pituitary tumor in MEN type I is not high.[6] Since it was a large nonfunctioning mass, surgical resection was the only option. Also, the tumors associated with MEN type 1 tend to be large macroadenomas with 45% being resistant to all medical, surgical, and radiotherapic management.[1] Growth hormone producing pituitary tumors causing acromegaly can present a major anesthetic airway concern due to macroglossia, prognathism, and distorted glottis leading to difficult ventilation and intubation in these

patients.[7,8] Furthermore, cardiac myopathy secondary to acromegaly can significantly increase the morbidity and mortality of these patients.[9] Hypercortisolemia secondary to pituitary or adrenal tumor may create a different set of problems including fluid electrolyte dysfunction, hypertension, atherosclerosis, glucose intolerance, increased risk of thromboembolism, osteoporosis, presence of obstructive sleep apnea, and difficult airway management.[1,9,10] Primary hyperparathyroidism occurs in 90% of the patients with MEN type I, leading to hypercalcemia, renal stones, osteoporosis, and rarely electrocardiographic changes secondary to hypercalcemia.[1] Although serum parathyroid hormone was on the higher side, since the serum calcium levels were near normal, we decided to go ahead with the surgery. Adequate hydration was ensured and careful positioning and proper padding of pressure points done. Treatment with calcitonin or other agents is required only after serum calcium exceeds 13 mg/dL.[11] Pancreatic insulinomas are quite common too, occurring in 10 to 30% cases. Gastrinomas or Zollinger Ellison syndrome is also very common in MEN type I, making ulcer prophylaxis imperative.[1] Management of these patients for any surgical procedure presents major anesthetic concerns due to the above endocrine dysfunction.

Our anesthetic considerations were airway and respiratory concerns due to preoperative right diaphragmatic palsy, past history of chest radiotherapy and postoperative nasal packing, steroid replacement due to low serum cortisol levels, hydration and positioning due to hyperparathyroidism, high sugars, gastroduodenal ulcers, and early postoperative recovery for neurological examination.

Invasive arterial and central venous cannulations are not indicated routinely for transsphenoidal surgeries in absence of cardiovascular complications[9,12] and hence not done by us. Also, there is only a 10% risk of venous air embolism (VAE) due to head-up position, there is no supporting evidence in literature of severe VAE during this surgery.

Anesthesia technique and choice of anesthesia agents depend on the patient's comorbidities, allergies, and need for rapid emergence. We used dexmedetomidine due to its early recovery with minimal postoperative sedation and no respiratory depression, as also due to its central sympatholytic action.[13] We used a standard cuffed endotracheal tube inserted orally and shifted to left angle of the mouth. We decided to use succinylcholine for intubation since the patient had no signs of raised intracranial pressure and normal serum potassium levels. We also had airway concerns due to past history of chest radiotherapy and right phrenic nerve palsy. Moreover, we wanted to avoid nondepolarising muscle relaxant to facilitate early postoperative neurological recovery with surgical duration being short (25 minutes). Hence, we used succinylcholine after adequately premedicating the patient.[14] Short acting agents like propofol and desflurane were used to help with rapid recovery.

The postoperative care after pituitary surgery includes airway management, analgesia, prevention of nausea and vomiting, neurological, and visual and endocrinal dysfunction assessments. Common postoperative problems are epistaxis, cerebrospinal fluid (CSF) leak, disorders of water balance like diabetes insipidus and syndrome of inappropriate antidiuretic hormone secretion (SIADH), cranial nerve injury, vascular injury, septal perforations, visual loss, etc.[5,9,12] Epistaxis may be treated with nasal packing for 2 and 3 days postoperatively. Very rarely, reoperation or endovascular embolization of feeding vessel may be required. Cerebrospinal fluid leak was checked by Valsalva maneuver intraoperatively and thigh fat insertion for dural closure was done. Diabetes insipidus and SIADH is found in up to 25% cases after pituitary surgeries.[9] Strict fluid intake and output charts were maintained along with monitoring of serum sodium and urinary specific gravity. Also, monitoring of serum calcium along with parathyroid hormone was done, followed by parathyroidectomy subsequently.

The excised sellar mass turned out to be neuroectodermal thymic carcinoma metastasis on biopsy. Occult metastasis are more common in patients with MEN type I with neuroectodermal tumors.[1]

CONCLUSION

The anesthesia management for MEN type I patients pose unique challenges to the anesthesiologist due to involvement of multiple endocrine glands which necessitates a multidisciplinary approach. Meticulous preoperative evaluation, intraoperative anticipation, and prevention and management of potential complications along with postoperative monitoring form the cornerstone of successful perioperative management of these patients.

REFERENCES

1. Thakker RV, Newey PJ, Walls GV, Bilezikian J, Dralle H, Ebeling PR, Melmed S, Sakurai A, Tonelli F, Brandi ML, et al. Clinical practice guidelines for multiple endocrine neoplasia type 1 (MEN1). J Clin Endocrinol Metab 2012 Sep;97(9):2990-3011.
2. Brandi ML, Gagel RF, Angeli A, Bilezikian JP, Beck-Peccoz P, Bordi C, Conte-Devolx B, Falchetti A, Gheri RG, Libroia A, et al. Guidelines for diagnosis and therapy of MEN type 1 and type 2. J Clin Endocrinol Metab 2001 Dec;86(12):5658-5671.
3. Romei C, Pardi E, Cetani F, Elisei R. Genetic and clinical features of multiple endocrine neoplasia types 1 and 2. J Oncol 2012 Jan;2012:705036.
4. Niina Y, Fujimori N, Nakamura T, Igarashi H, Oono T, Nakamura K, Kato M, Jensen RT, Ito T, Takayanagi R. The

current strategy for managing pancreatic neuroendocrine tumors in multiple endocrine neoplasia type 1. Gut Liver 2012 Jul;6(3):287-294.
5. Zada G, Woodmansee WW, Iuliano S, Laws ER. Perioperative management of patients undergoing transsphenoidal pituitary surgery. Asian J Neurosurg 2010 Jan;5(1):1-6.
6. O'Brien T, O'Riordan DS, Gharib H, Scheithauer BW, Ebersold MJ, van Heerden JA. Results of treatment of pituitary disease in multiple endocrine neoplasia, type I. Neurosurgery 1996 Aug;39(2):273-278; discussion 278-279.
7. Nemergut EC, Zuo Z. Airway management in patients with pituitary disease: a review of 746 patients. J Neurosurg Anesthesiol 2006 Jan;18(1):73-77.
8. Friedel ME, Johnston DR, Singhal S, Al Khalili K, Farrell CJ, Evans JJ, Nyquist GG, Rosen MR. Airway management and perioperative concerns in acromegaly patients undergoing endoscopic transsphenoidal surgery for pituitary tumors. Otolaryngol Head Neck Surg 2013 Dec;149(6):840-844.
9. Dunn LK, Nemergut EC. Anesthesia for transsphenoidal pituitary surgery. Curr Opin Anaesthesiol 2013 Oct;26(5):549-554.
10. Simonds WF, Varghese S, Marx SJ, Nieman LK. Cushing's syndrome in multiple endocrine neoplasia type 1. Clin Endocrinol (Oxf) 2012 Mar;76(3):379-386.
11. Sjöberg HE, Hjern B. Acute treatment with calcitonin in primary hyperparathyroidism and severe hypercalcaemia of other origin. Acta Chir Scand 1975 Jan;141(2):90-95.
12. Horvat A, Kolak J, Gopcević A, Ilej M, Zivko G. Anesthetic management of patients undergoing pituitary surgery. Acta Clin Croat 2011 Jun;50(2):209-216.
13. Brady T. Anesthetic management of a pituitary tumor resection with dexmedetomidine. AANA J 2010 Apr;78(2):125-128.
14. Maurtua MA, Deogaonkar A, Bakri MH, Mascha E, Na J, Foss J, Sessler DI, Lotto M, Ebrahim Z, Schubert A. Dosing of remifentanil to prevent movement during craniotomy in the absence of neuromuscular blockade. J Neurosurg Anesthesiol 2008 Oct;20(4):221-225.