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

1Anjana S Wajekar, 2Anita N Shetty, 3Shrikanta P Oak, 4Ruchi A Jain

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 endocrine and nonendocrine expressions of MEN type I include adrenocortical, gastric, thymic or bronchial tumors, foregut carcinoids, visceral and cutaneous lipomas, and meningiomas and facial angiofibromas. It occurs in about 1 in 30,000 individuals, with an estimated prevalence of 2 to 3 per 100,000 population. Anterior pituitary tumors are found in 30 to 40% cases of MEN type 1 syndrome patients. They are commonly prolactinomas (60%), somatotropinomas (25%), <5% corticotropinoma or nonfunctioning. 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.

CASE REPORT
A 45-year-old male patient, known case of familial MEN type I syndrome, presented with chief complaints of generalized body ache. 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 glinipiride 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...
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The patient with propofol 150 mg and succinylcholine was premedicated with ranitidine 50 mg, midazolam 1 mg, and fentanyl 100 µg intravenously. We induced dexmedetomidine 1 µg/kg over 10 minutes via infusion. 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. We extubated 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. Pneumatic pressure points were 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. We extubated the patient 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. Incidence is found to be similar across all ages and both sexes. 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. 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 geneticist.

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. Since it was a large nonfunctioning mass, surgical resection was the only option. Also, the tumors associated with MEN type I tend to be large macroadenomas with 45% being resistant to all medical, surgical, and radiotherapeutic management. Growth hormone producing pituitary tumors causing acromegaly can present a major anesthetic airway concern due to macroglossia, proptosis, and distorted glottis leading to difficult ventilation and intubation in these
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.5 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 anesthetic 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.

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