Case Report

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Gigantic Solitary Dumbbell Spinal Neurofibroma Causing Gastric Outlet Obstruction: Anesthesia Management

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Abstract

Neurofibromatosis (NF) has been described greatly in literature and is broadly categorized into NF1 and NF2. The former NF1 is more common and is a heterogeneous condition and up to 38% of patients may present with spinal NF of which, 5% may have clinical problems. The latter is however, characterized by central nervous system tumors, particularly bilateral acoustic Schwannoma with or without other manifestations. Multiple spinal NF are very common in NF1. We report a solitary gigantic spinal NF of the dorso-lumbar region encroaching the abdominal cavity and causing gastric outlet obstruction and its anesthetic management, which to our knowledge has not been reported before in literature.

Key words: Dumbbell tumor, Extra spinal portion, Gastric outlet obstruction, Nerve sheath tumor, Neurofibromatosis Type 1

INTRODUCTION

Neurofibromatosis 1 (NF1) was first described by Freidrich von Recklinghausen in 1882 and there is a familial disposition for this condition with an incidence of 1:3300.[1] Clinical diagnosis is established by the presence of the following: Café au late spots (CLS), axillary and inguinal freckling, lisch nodule, optic glioma, cognitive disorders, scoliosis, bony skull defects, and spinal or paraspinal NF.[1,2] Anesthesia considerations are numerous and should be kept in mind before the practitioner attempts such cases as they are vital to patient safety.

CASE REPORT

An 18-year-old boy presented to neurosurgery outpatient department with vague abdominal discomfort and paresthesia of both the lower limbs, for the past 2 years. The patient had complaints of mild, dull pain abdomen predominantly in the left hypochondrium, flank and vomiting after having meals. On examination, inspection revealed distension on the left side, however no prominent veins or any visible peristalsis were present. Palpation revealed huge lump in the abdomen on the left side, well-rounded, well-defined, non- pulsatile, extending from left hypochondrium to left iliac crest. Rest of the physical examination was essentially normal. A magnetic resonance imaging (MRI) scan showed a large lobulated heterogeneous intra-dural extramedullary space occupying lesion (SOL) of the dorso-lumbar region, extending from D12-L4 vertebra without any intracranial lesions (such as optic or hypothalamic gliomas or meningiomas) (Figures 1-3). The dumbbell mass was encroaching the spinal canal through the enlarged intervertebral foramina and extending anteriorly into abdominal cavity and causing a gastric outlet obstruction (GOO) type phenomenon. A provisional diagnosis of NF1 was made and it was planned for complete surgical excision through lateral approach after thorough workup.

Pre-anesthetic checkup revealed that the boy did not have any systemic illness, allergies or prior hospitalization and work up for pheochromocytoma was negative. The

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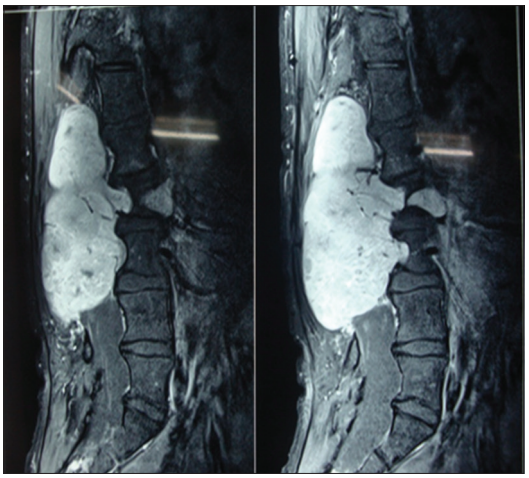

Figure 1: Magnetic resonance imaging scan showing a large lobulated heterogeneous intradural extramedullary mass noted in the dorso-lumbar region

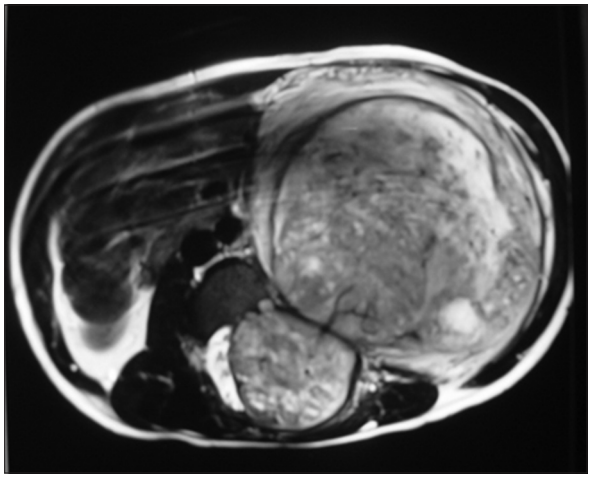

Figure 2: Magnetic resonance imaging scan showing a "dumbbell" spinal neurofibroma, with large with the extraspinal part being larger than the intraspinal portion

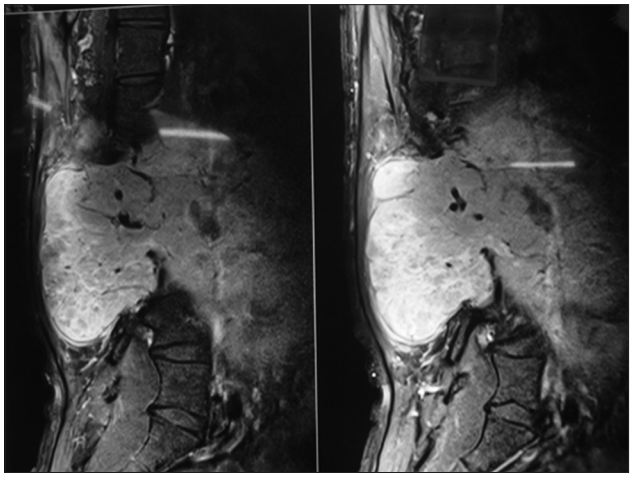

Figure 3: Magnetic resonance imaging scan again illustrating the same as in Figure 1

airway examination was routine and did not show any cause for concern. Blood grouping and cross matching were done and adequate blood products were arranged. His weight was 65 kg and pre-operative hemoglobin was 13.5 g%. The patient was pre-medicated with lorazepam 2 mg PO, on the night before surgery and an overnight fast was ordered. Aspiration prophylaxis was given with pantoprazole 40 mg PO and metoclopramide 10 mg PO, as well. Written informed consent was taken from the patient. On the day of surgery, patient was taken inside the operating room and monitors as per American Society of Anesthesiologists standards were applied (electrocardiogram, non-invasive blood pressure, SPO_2, TEMP after intubation, end-tidal CO_2 [$ETCO_2$]). A wide bore intravenous (IV) cannula (16 G) was inserted and a balanced IV solution was started. After pre-oxygenation with 100% oxygen for 5 min, anesthesia was co-induced with midazolam 1 mg IV, followed by fentanyl 150 μg and thiopental 550 mg IV. After the cessation of spontaneous breathing and loss of eyelash reflex, succinylcholine 100 mg was given and cricoid pressure was applied. Patient was intubated with an 8.5 ID armored tube after 45 s and cuff was immediately inflated. Tube position was confirmed with $ETCO_2$ and auscultation and it was secured snuggly with Durapore tape. Foley's catheter was inserted and urine output was monitored throughout the procedure. A triple lumen central venous catheter (7 French) was inserted in the right internal jugular vein followed by an "art" line in the left radial artery. This was based on discussion with surgeons, who had warned that the SOL was vascular and may lead to serious bleeding. After securing all invasive lines and endotracheal tube, patient was carefully position in the right lateral position (left side up). The patient was given 4 mg of vecuronium after he came out of the effect of succinylcholine and started breathing spontaneously. Anesthesia was maintained with oxygen and nitrous oxide and isoflurane titrated to depth of anesthesia. Muscle relaxation was maintained with vecuronium (intermittent dosing) monitored by neuromuscular monitoring.

Initial surgical approach from the surface was uneventful however when the superficial lobe was lifted up to gain access to the deeper one, substantial hemorrhage occurred. About 3 L of blood were lost, and this was replaced by crystalloids, colloids, and then packed red blood cells (PRBC). Four units of PRBC were transfused with eight units of fresh frozen plasma. Serial arterial blood gases were done and acid base balance and electrolyte disturbances were corrected, during the procedure. After a tough time, the tumor was removed in Toto and hemostasis was achieved. The size of the tumor was about 14" × 8" and it weighed about 4100 g (Figures 4 and 5). The patient did not require any inotropic support or any vasopressors, but as a result of massive blood transfusion and associated fluid therapy he had become hypothermic and hence, he was ventilated overnight in the neurosurgery Intensive

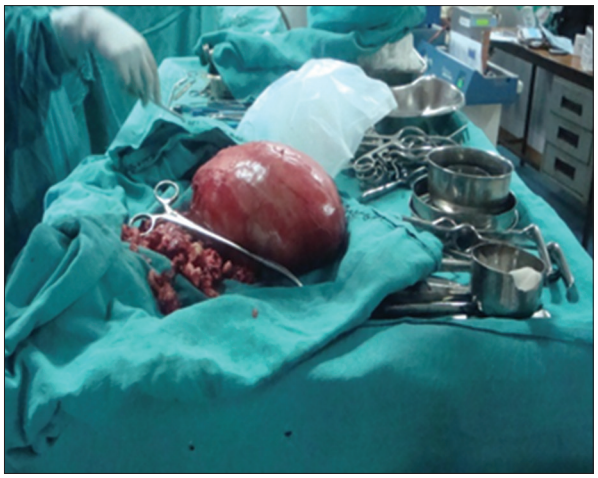

Figure 4: The intraoperative image of the mass upon removal, along with sponge holding forceps to get an idea of the size

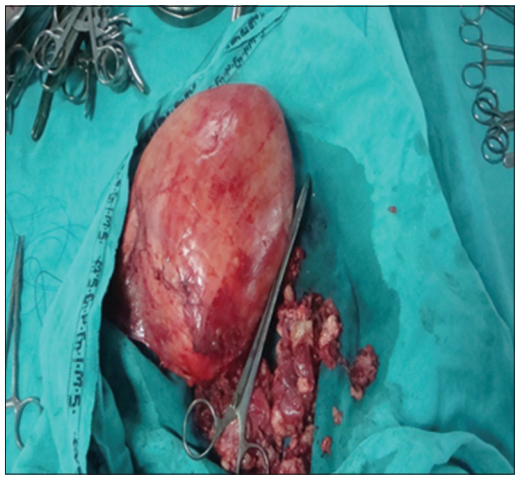

Figure 5: The mass having dimensions of 14" × 8" and weight 4.1 kg

Care Unit. On the following morning, he was extubated uneventfully, after he was assessed to be awake and breathing spontaneously and adequately. Post-operative course was uneventful and he was discharged from the hospital after 9 days, without any neurological deficit.

DISCUSSION

NF1 is a multisystem disorder that may virtually involve any organ in the body and the clinical presentation depends on the organ involved.[1,2] Otherwise known as "von Recklinghausen disease" or "peripheral NF," it comprises 90% of all NF cases. It is a type of phakomatosis which is inherited in an autosomal dominant fashion.[1,2]

The anesthesiologist should know that there may be airway involvement (NF of larynx, supraglottic area or aryepiglottic fold) with potential for loss of control over the airway, cervical NF, hypertension secondary to pheochromocytoma or renal artery stenosis and vocal cord dysfunction.[3-8]

This case report, illustrates a solitary, dorso-lumbar spinal NF without the classical signs of NF1 such as CLS, iris Lisch nodules and cutaneous NF, causing GOO phenomena.[9]

NF in the spinal canal frequently invade the peripheral segment of the nerve through the intervertebral foramen and develop a dumbbell shape.[1] The extraspinal portion of the tumor is larger than the intraspinal portion in such cases, and it is this extraspinal portion that gets attached to surrounding structures and may leading to extensive bleeding as in our case.[1] There may be areas of degeneration in the extraspinal portion, as seen in the MRI of our patient.

The dumbbell tumor by virtue of mass effect on the thoracic or abdominal cavities may cause respiratory embarrassment or gastrointestinal dysfunction, respectively.[1] This was seen in our case as after the surgery, the patient had relief from features of GOO and lower limb paresthesia. Other cases of NF have been described that caused GOO, however they were arising from the stomach or other parts of the gastrointestinal tract.[10-12] This is the first case to describe GOO from an extraspinal portion of a dumbbell NF.

The extraspinal portion was occupying a large part of the abdominal cavity, retroperitoneally, leading to the theoretical risk of aspiration secondary to impaired gastric emptying due to mass effect. Hence, aspiration prophylaxis was given with pantoprazole 40 mg PO and metoclopramide 10 mg PO, (on night before surgery and 2 h prior to surgery) and modified rapid sequence intubation with cricoid pressure was done in this case to circumvent this issue.

CONCLUSION

We successfully managed a huge dumbbell, spinal NF in the dorso-lumbar region causing GOO. Going into the literature, we understand that the anesthesia considerations are mainly as follows: (1) Bleeding from the extraspinal portion of the dumbbell tumor due to its adhesions, and (2) problems from the "mass" i.e., "mass effect" (in our case GOO, in others respiratory embarrassment). Therefore, we were prepared to manage the hemorrhage (blood products, invasive monitoring, and vascular access) and modified rapid sequence intubation for general anesthesia. To our knowledge, there has been no report of a similar case in literature.

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