Letter

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A rare challenge in general surgery: double surgical procedure for large and small bowel obstruction in a patient with Gerstmann-Sträussler-Scheinker syndrome

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SUMMARY Gerstmann-Sträussler-Scheinker syndrome (GSS) is a rare, infectious syndrome related to a mutation in the prion protein gene. Described here are the challenges posed by surgery for a patient with GSS. A 61-yr-old woman with GSS was admitted to this department and underwent surgery twice for large and small bowel obstruction. This is the first report of two major surgical procedures in a patient with GSS. Experiences with this case and precautions when using a disposable device during endotracheal intubation and a surgical procedure to manage a patient with GSS are described.

Keywords Gerstmann-Sträussler-Scheinker syndrome, general surgery, bowel obstruction

Gerstmann-Sträussler-Scheinker Syndrome (GSS) is a rare prion disease (PD). This familial (autosomal dominant), fatal neurodegenerative disease affects patients from 20 to 60 years of age and is related to a prion protein (PrP) mutation in the group of subacute spongiform encephalopathies (SEs) (*1*). Human SEs include Creutzfeldt-Jakob disease, GSS, Kuru, and fatal familial insomnia. GSS is characterized by amyloid deposition in the cerebral parenchyma or blood vessels (*2*). Symptoms include dysarthria, progressive cerebellar truncal ataxia, pyramidal signs, and adult-onset dementia. Some studies have suggested that a PD might be transmitted by blood or plasma-derived products from patients during the prodromal stage. In animal studies, intracerebral inoculation of infected cells has been associated with development of disease, and infectivity was also detected in the blood (*3*). Given the lack of information, the resistance of PrP to conventional sterilizing measures is a major problem. Current recommendations are to identify at-risk patients and to use disposable devices during endotracheal intubation, spinal anesthesia, and surgery (*4*).

A 61-yr-old woman was admitted to this Hospital for abdominal pain. At the age of 53, she complained of dysarthria, spastic paraparesis, cognitive decline, amyotrophy, depression, cerebellar ataxia, pseudo-bulbar palsy, and absent tendon reflexes in the lower limbs. Her father and sister had the same symptoms. A typical mutation of the 102nd amino acid (PRNP-p.D202N) and a drastic change were found in the normal prion gene. She was then diagnosed with classical GSS. Her Unified Parkinson's Disease Rating Scale III (UPDRSIII) score was 26 and her Hoehn and Yahr scale score was 3 (*5*). The patient was admitted to the emergency department for abdominal pain. Laboratory results revealed an increased white blood cell count (WBC) [14×10^9/L (NV 4–10×10^9] and elevated C-reactive protein (CRP) level [33.3 mg/L (NV < 1.0)]. A CT scan of the abdomen and thorax revealed sigmoid volvulus. The endoscopic evaluation confirmed sigmoid stenosis and mucosal necrosis. After an interview with the patient's caregivers who are actively present in the patient's life and fully dedicated to her medical and physical care and a consultation with anesthesiologists and physiotherapists, a plan was formulated to use general anesthesia with tracheal intubation in order to perform an explorative laparotomy.

A volvulus of the dolichosigmoid colon with bowel obstruction and ischemia was detected intraoperatively. In order to avoid anastomotic complications, the patient underwent a Hartmann's resection with end-colostomy. The highest level of protection was used during surgery. In addition to disposable masks and caps, all medical staff wore gowns and gloves. Disposable surgical instruments and anesthetic equipment were used. The scrub team was equipped with full personal protective equipment. The number of surgical, nursing, and anesthesia team members was limited to

Table 1. Patient characteristics

	Age (yrs)	Neurological Symptoms	Abdominal Symptoms	WBC ($\times 10^9$/L)	CRP (mg/L)	Radiological Findings	Surgical Procedure	Anesthetic Outcome	Surgical Outcome
1st surgery	61	Dysarthria; Spastic Paraparesis; Dementia; Cerebellar Ataxia; Pseudo-bulbar Palsy; Tendon Reflexes Absent	Abdominal Pain; Nausea and Vomiting; Bowel Obstruction	14	33	Colonic Distension; Bowel Obstruction; Sigmoid Volvulus; No Signs of Perforation	Laparoscopic Hartmann's Resection with end Colostomy	No Airway Obstruction; Extubated on Day 1 Postop Percutaneous mini-tracheostomy	Uneventful; Discharged on Day 6 Postop
2nd surgery	62	Stable	Abdominal Pain; Nausea and Vomiting; Small Bowel Obstruction	12	9	Bowel Distention; Closed-loop Obstruction	Laparotomic Small Bowel Resection; PEG Tube Placement	Uneventful	Uneventful; Discharged on Day 9 Postop

yrs: years; WBC: white blood cell count; CRP: C-reactive protein.

the minimum required to perform the surgery. After confirming full recovery of muscle strength, double-burst stimulation, and spontaneous eye opening, the tracheal tube was removed with no incidence of airway obstruction. She was admitted to the intensive care unit on day 1 postoperatively, where her vital signs were stable. She was extubated on day 1 postoperatively and a percutaneous mini-tracheostomy for pulmonary aspiration was performed. The postoperative course was uneventful. The patient underwent physiotherapy sessions and was discharged on day 6 postoperatively without complications.

One year later, the patient was seen by the emergency department for small bowel occlusion. Blood tests revealed increased CRP [9 mg/L (NV < 1.0), WBC [12 10^9/L (NV 4.0-9.0)] and plasma lactate (3.9 mmol/L). Neurological symptoms were stable and the level of home care was optimal. A second emergency laparotomy was performed, and small bowel ischemia due to ileal obstruction was detected intraoperatively. A small bowel resection was performed and an endoscopic gastrostomy (PEG) tube was placed for enteral nutrition. The same level of protection as in the first surgical procedure was used. The postoperative course was uneventful, and the patient was discharged on day 9 postoperatively with a permanent urinary catheter and PEG tube for enteral nutrition (Table 1).

There are no cases of a patient with GSS undergoing multiple abdominal surgeries under general anesthesia in the literature (according to a search for relevant articles on PubMed and Embase using the terms "GSS" OR "PD" AND "Surgery" (*4*)).

The challenge in the surgical treatment of patients with GSS involves intra- and post-operative anesthesiological risks (*e.g.* airway obstruction, bronchospasm, or pneumonia), surgical risks (*e.g.* immobility, dysphagia, or impaired canalization), and risks of infection. PrP is present in the central nervous system, appendix, and lymphatic tissues and is resistant to inactivation by radiation, heat, or aggressive chemical treatments. Patients with GSS must be managed with specific precautions to prevent infections.

The feasibility of abdominal surgical procedures in patients with GSS cannot be determined based on this single case, despite its favorable outcome. Nonetheless, it indicates that surgery, with adequate caution, can be used to treat this complex condition. In conclusion: *i*) the patient must have family members or caregivers actively participating in post-operative management in the hospital and at home; *ii*) use of disposable equipment is mandatory to avoid the transmission of infection to medical staff; and *iii*) the increased risk of post-operative complications must be taken into account.

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