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Case report

Total gastrectomy for rare refractory gastroparesis in patient with syringomyelia: A good impact on quality of life

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HIGHLIGHTS

- Syringomyelia is a spinal disorder rarely associated with gastrointestinal dysfunction.
- We present a case of refractory gastroparesis in a female patient with syringomyelia.
- Surgery may be indicated if dietary changes and medical management fails.
- We performed a total gastrectomy with improvement in the patient's quality of life.

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ABSTRACT

Syringomyelia is a chronic progressive disease of the spinal cord. In symptomatic patients, bilateral sensory motor signs and symptoms prevail, moreover they can develop gastrointestinal disorders, although few studies have succeeded in explaining this correlation so far.

We report a case of a 67-year-old woman with a history of pain in the back-lumbar spine and lower limbs, paresthesia and urinary incontinence. MRI revealed syringomyelia, extended from T3 to the medullary cone. Neurological picture was worsened by progressive and increasingly debilitating gastrointestinal symptoms refractory to dietary changes and medical treatment. Blood tests, gastrointestinal investigations and imaging were all normal apart from scintigraphy which confirmed delayed gastric emptying. The neurological symptoms disappeared after removal of an hemangioblastoma of the medullary cone. The persistent gastroparesis was treated by total gastrectomy with complete resolution of the patient's gastrointestinal symptoms.

1. Introduction

Syringomyelia literally means “cavity within the spinal cord” and it is usually a chronic progressive disease. The syrinx may look like a fluid-filled, gliosis-lined cavity within the spinal cord parenchyma or a focal expansion of the center canal; in this case, we call it hydromyelia. Most injuries are located between C2 and T9, but they may go down to the medullary cone or extend upwards to the brainstem (syringobulbia).

In Western countries, the prevalence rate has been estimated to be 8.4 per 100,000. In children, syringomyelia usually develops in the context of congenital abnormalities, mostly Chiari I malformation and tethered cord, but it can also develop years later, as a result of meningitis, spinal trauma or extramedullary/intramedullary tumors. In symptomatic syringomyelia patients, bilateral sensory motor signs and symptoms prevail [1].

Moreover, syringomyelia patients can develop gastrointestinal disorders, although few studies have succeeded in explaining this correlation so far [2–5]. This report describes the case of a female patient with syringomyelia and a highly disabling gastroparesis

Abbreviations: MRI, Magnetic Resonance Imaging; GERD, Gastro-Esophageal Reflux Disease; CT, Computed Tomography; MVGS, Modified Visick Grading System; FDA, Food and Drug Administration.

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which was resistant to medical therapy and was successfully treated with total gastrectomy.

2. Case report

We present the case of a woman of 67-years-old with a long history of pain in the back-lumbar spine and lower limbs, paresthesia of the right lower and urinary incontinence, previously operated for herniated disc L5-S1 (1979) and lumbar canal stenosis (1983). Following MRI of the lumbar spine in 2007 she was diagnosed with syringomyelia, extended from T3 to the medullary cone.

Three years later, neurological picture was worsened by progressive and increasingly debilitating gastrointestinal symptoms: nausea, upper abdominal pain, early satiety, postprandial fullness, anorexia, GERD-like symptoms, dysgeusia with persistent feeling of "salt in the mouth" and rare episodes of vomiting: initial treatment envisaged dietary modifications, proton pump inhibitors and H2 antagonists, without any success.

Since then, she has been experiencing frequent hospitalizations, marked by several diagnostic tests: blood tests had normal values (including immunological-allergy tests and viral serology tests); two esofagogastroduodenoscopy turned out negative for esophageal-gastric organic diseases; Urea Breath test was negative for *Helicobacter pylori*; pH 24 h impedenziometry was negative for acid/no acids refluxs; esophageal manometry was negative for esophageal motility disorders; upper gastrointestinal tract radiography and entero-MRI were normal, abdomen CT was negative for organic diseases. At a first gastric 99^{m} Tc-scintigraphy there was evidence of marked slowing of gastric emptying: 65% gastric contents at 60 min (35% ±5%) and 52% at 120 min (9% ±3%).

Following a diagnosis of gastroparesis in 2013, an initial adequate prokinetic therapy based domperidone was chosen (she had history of intolerance to metoclopramide) in addition to antiemetic agents; since it turned out to be ineffective, it was replaced by erythromycin, in addition to antiemetic agents and selective serotonin reuptake inhibitors, with poor results.

Spine control MRI highlighted a significant increase of syringomyelic cavitation and a hypervascular oval lesion (hemangioblastoma) located in the medullary cone that was identified as the cause of syringomyelic degeneration and then successfully removed by neurosurgery. Despite a reduction of neurological disorders, gastrointestinal symptoms did not get benefit from marked weight loss (−13 kg/10 months) which was related to a reduced food intake. A 2014 gastric scintigraphy showed a further slowing of gastric emptying (76% at 60 min, and 66% at 120 min).

Therefore, after multidisciplinary clinical case reassessment, a Roux-en-Y total gastrectomy was performed, with an end-to-side circular stapled esophagojejunostomy and a retro-colic alimentary limb of the length of 60 cm.

The clinical course was uneventful, without any complications and complete resolution of gastrointestinal symptoms, shortly in the close postoperative period. She was discharged on the tenth day and six months after surgery a marked improvement in the quality of life was recorded (from Grade 4 to Grade 1 of MVGS). Follow-up X-ray of gastrointestinal tract showed regular progression of barium and regular bowel emptying, with absence of significant reflux (Fig. 1).

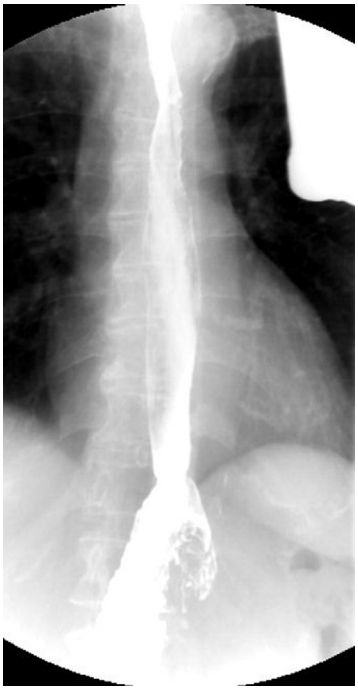

Fig. 1. X-ray of gastrointestinal tract showed regular progression of barium in esophagus, regular trans-anastomotic outflow in the bend of the same downstream and regular bowel emptying, with absence of significant esophageal reflux of barium.

3. Discussion

Syringomyelia is a rare chronic degenerative disease of the spinal cord and brainstem marked by the infiltration of cerebrospinal fluid and the formation of a cystic cavity (syrinx), which can generate compressions and/or injuries to the nerve fibers of the spinal cord itself. The first syringomyelia symptoms (loss of pain and temperature sensitivity) come from the involvement of the spinothalamic fibers crossing anterior commissure's white matter, which mainly affect the upper part of neck, shoulders, arms, hands. The enlargement of the syrinx can even damage anterior horn cells and corticospinal tract, with following loss of motor functions of upper and/or lower limbs. Other syringomyelia signs and symptoms may include: muscle weakness and wasting (atrophy), loss of reflexes, headache, diplopia, stiffness, ataxia, hiccups, neuropathic pain, gastrointestinal dysfunction and bladder, curvature of the spine (scoliosis) [2,6].

Syringomyelia patients may show gastrointestinal disorders, although few studies have described the link between syringomyelia and gastrointestinal dysmotility (oropharyngeal, cricofaryngeal, esophageal-gastric and anorectal abnormalities) so far [2–5]. In our case report, the patient showed symptomatic syringomyelia for hemangioblastoma and gastroparesis, marked by the association of cardinal symptoms (early satiety, postprandial fullness, nausea, vomiting, bloating and upper abdominal pain) and objectively delayed gastric emptying in absence of mechanical obstruction [7]. In literature the most common causes of gastroparesis are represented by idiopathic (36%), diabetes (29%), postgastric surgery (13%), Parkinson's disease (7.5%), collagen vascular disease (4.8%), intestinal pseudo-obstruction (4.1%) and miscellaneous (6%). Other possible causes are: neurological disorders, eating disorders, different metabolic or endocrine disorders (hypothyroidism) [8].

In the absence of any other obvious cause of gastrointestinal dysmotility, we attributed it to her syringomyelia as her symptoms developed and worsened as the syringomyelia cavity appeared and enlarged.

The etiology of motor related disorders in syringomyelia patients is unknown [2]. Although an accurate identification of the area of neurological dysfunction responsible for gastric dysmotility has not been made yet, in our case it could be the result of the progressive and marked syrinx widening in spinal cord and

consequent damage to the afferent and/or efferent fibers of autonomic nervous system, which are responsible for controlling gastric motor activity. Histologic examination of surgical specimen showed neither alteration of smooth muscle cells, morphology of enteric neurons and interstitial cell of Cajal, not an increase in inflammatory cells or alterations in microvascular tissue acid.

For gastroparesis patients of any etiology, therapy starts by improving diet and lifestyle, usually in combination with a medical therapy aimed to achieve better management of chronic symptoms; all this in order to allow those patients to enjoy a reasonably good quality of life. However, for those patients who suffer from debilitating and refractory disease, although all previous therapies failed, there are several available surgical options: they include gastrostomy or jejunostomy feeding tubes, pyloroplasty, gastric electrical stimulation (GES) and gastrectomy. GES was approved by the FDA in 2000 as a Humanitarian Use Device (HUD); it generates a low-energy and high-frequency neurostimulation of smooth muscle cells through two electrodes placed within the muscularis propria of the stomach and connected to an adjustable generator. Previous studies have shown that GES would be useful to relieve vomiting and nausea. However, given the limited data and system status as a HUD, more additional clinical trials are necessary to prove its effectiveness in reducing the symptoms of gastroparesis [9–12].

In the current literature it has also been described the laparoscopic approach for the treatment of gastroparesis, not only for gastric resections, but also in the placement of GES device. The results described were excellent in addition to the indisputable advantages of the approach: a decrease of post-operative pain, reduced morbidity, early food resumption, shorter hospital stay [10,13,14].

On the basis of encouraging results [15–17], although limited to a small number of case studies focused mainly on post-surgical or diabetic refractory gastroparesis, we decided for total gastrectomy, deeming it necessary to remove “the organ which causes the disease”, although our patient and we were well aware of morbidity and mortality associated to such an extreme approach. The success was evident from the immediate post-operative phase. In the six months after surgery, the patient reported a considerably improved quality of life, resulting from the complete resolution of clinical picture, the resumption of normal eating and social habits and a slight weight gain (Table 1).

4. Conclusion

Syringomyelia is a rare neurological disorder rarely associated with gastrointestinal dysfunction. Our case report describes the association between syringomyelia and gastroparesis, whose first treatment envisages dietary changes and medical therapy up to surgical intervention, if a refractory and highly disabling condition persists. Among surgical options we mention total gastrectomy; today, it is considered as the last chance and it works mainly in gastroparesis cases related to gastric surgery outcomes. We chose it as first surgical option, as we were convinced of the necessity to remove the affected organ; in the event of gastroparesis associated with a neurological disorder the results were outstanding, given the complete resolution of clinical picture and the indisputable improvement of patient's quality of life.

Table 1
Modified Visick Grading System [15].

Grade 1	Perfect result, an asymptomatic patient
Grade 2	Mild, intermittent symptoms that are easily controlled by diet
Grade 3	Moderate symptoms without substantial interference with lifestyle
Grade 4	Unsatisfactory outcome and includes all patients with recurrent ulcer

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent was sent to the Editor of this journal.

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Author contribution

Zizzo: surgical management, data collection, analysis, interpretation, writing.
Bonilauri: surgical management, data collection, analysis.
Lanaia: surgical management, data collection, analysis.
Negro: clinical management.
Santi: clinical management.

Conflict of interest

The authors declare that they have no conflict of interest.

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