Case report

Claude Bernard-Horner syndrome associated with colonic interposition grafting after esophagectomy

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SUMMARY

A 34 year-old male was admitted in our hospital because of double vision, ptosis of the eyelid, miosis and enopthalmos of the left eye together with impaired sweating of the upper body quarter for the last two weeks. Ten years ago the patient underwent total esophagectomy with colonic graft interposition because of esophageal rupture during large caliber bougienage for congenital esophageal stenosis. The endoscopy showed a large diverticulum below the upper esophageal sphincter and numerous ulcerations of the lower part of the colonic graft in the area of the former gastro-esophageal junction. The patient was diagnosed with post-ganglionic Claude Bernard-Horner syndrome due to stellate ganglion compression from the colonic diverticulum. The patient was discharged with recommendations for liquid diet and body positions that could facilitate left stellate ganglion decompression. In the case of persisting symptoms the patient was advised about the possibility of a new surgical intervention including diverticulectomy. To the best of our knowledge this is the first reported case of Claude Bernard- Horner syndrome as a long term complication of colonic graft diverticle after esophagectomy for congenital esophageal stenosis

Key words: Claude Bernard-Horner syndrome, colonic interposition, colon grafting, esophagectomy, neuro-opthalmic complications, esophagus

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INTRODUCTION

Claude Bernard-Horner syndrome [C. Bernard 1813-1887, J.F. Horner 1831-1886] is characterized by ptosis of the eyelid (paralysis of the smooth superior tarsal muscle), together with more or less marked miosis (paralysis of the dilator pupillae), with less impressive hyperemia of the conjunctiva (vasomotor paralysis), with enopthalmos (paralysis of the smooth orbital muscle or Muller muscle) and, often, with impaired sweating of the upper body quarter.

Claude Bernard-Horner syndrome can result from lesion of the central sympathetic homolateral pathways passing among the hypothalamus, the dorsolateral medulla oblongata and the lateral columns of the spinal cord or from lesions of the paravertebral sympathetic chain and its radicular afferents.

The common causes of the last are neoplastic (i.e thyroid carcinoma) or inflammatory involvement of the cervical lymphnodes or of the proximal part of the brachial plexus, surgical and other types of trauma to cervical structures such as jugular venous catheters, carotid artery dissection, syringomyelic or traumatic lesions of the first and second thoracic spinal segments and infarcts or other lesions of the lateral part of the medulla (Wallenberg syndrome). Congenital stenosis of the esophagus is an exceptional condition, for which three main types are described: a mucous diaphragm, to be compared with membranous atresia, fibrous stenosis and stenosis caused by bronchial heterotopy[1]. The diagnosis is established in the newborn, and less frequently in children and adults, in cases of vomiting, regurgitation or dysphagia. It is based on esophago-gastro-duodenal follow through and fiberendoscopy, which allows ruling out other causes of benign narrowing. Treatment resorts to endoscopic dilatations (bougienage) and to

surgery with esophageal substitution, which remains the safest method.

CASE REPORT

A 34 year-old male was admitted to our hospital because of double vision, ptosis of the eyelid, miosis and enopthalmos of the left eye together with impaired sweating of the upper body quarter for the last two weeks (Figure 1a).

Ten years ago the patient underwent total esophagectomy with colonic graft interposition because of esophageal rupture during large caliber bougienage for congenital esophageal stenosis.

The patient fulfilled the criteria of Claude Bernard-Horner syndrome and underwent investigation with brain and neck computed tomography, which was negative. In addition, routine laboratory tests were within normal limits. As pre-ganglionic (stellate ganglion) or central nervous system damage was excluded, a post-ganglionic cause of the syndrome was speculated. Barium meal showed a slightly dilated spiral colonic graft with a left mediastinal position (Figure 1b) and neck computed tomography was suggestive of stellate ganglion compression from the colonic diverticulum (Figure 1c). Although the patient did not complain of dysphagia and long-term food impactions, an upper gastrointestinal endoscopy (using a peadiatric type endoscope) was performed. The endoscopy showed a large diverticulum below the upper esophageal sphincter and numerous ulcerations of the lower part of the colonic graft in the area of the former gastro-esophageal junction. The colonic graft was spiral and slightly dilated with no evidence of stenosis or fistulae co-existence.

The patient was diagnosed with post-ganglionic Claude Bernard-Horner syndrome due to stellate ganglion compression from the colonic diverticulum. He was discharged with recommendations for soft food diet and body positions that could facilitate left stellate ganglion decompression. In case of persisting symptoms the patient was advised about the possibility of a new surgical intervention including diverticulectomy. Two years later, the patient remained in a stable condition and good nutrional status, and his opthalmological complications were significantly improved.

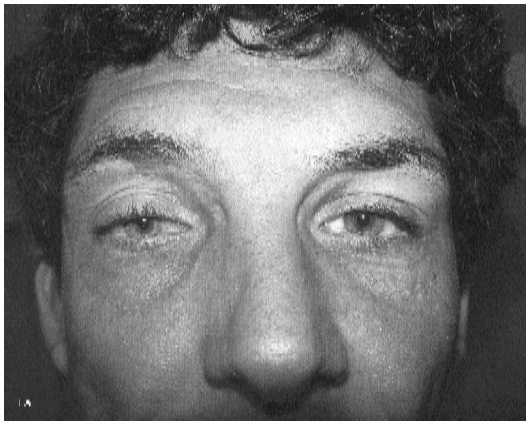

Figure 1a. Ptosis of the eylid, miosis and enopthalmos of the left eye in a patient with Claude Bernard-Horner syndrome associated with colonic interposition grafting after esophagectomy

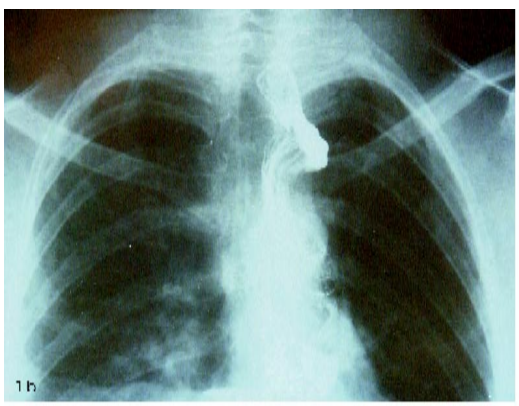

Figure 1b. Barium meal after esophagectomy for congenital esophageal stenosis showing the colonic graft with left mediastinal position.

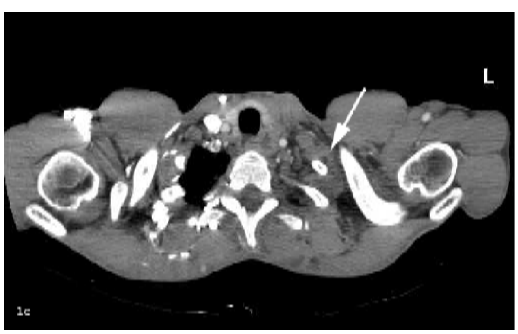

Figure 1c. Neck computed tomography showing stellate ganglion compression from the colonic diverticulum

DISCUSSION

Claude Bernard-Horner syndrome is a well-recognized but extremely rare complication of esophagectomy.[1] By contrast, colonic graft diverticula causing this syndrome as a long-term complication after esophagec-

tomy has not been previously described.

Differential diagnosis must be carefully done. With lesions on the stellate ganglia, Claude Bernard-Horner syndrome occurs in association with anhidrosis of the face. With paravertebral sympathetic chain lesions immediately caudal to the stellate ganglion, the result is isolated anhidrosis of the face without Claude Bernard-Horner syndrome, which was not the case reported here. Instillation of 1% of phenylephrine drops will cause dilatation of the miotic pupil when damage is post-ganglionic, as in this patient, but not in pre-ganglionic or central nervous system damage.

Congenital esophageal stenosis is the least common congenital tracheoesophageal anomaly. It is estimated to occur per 25,000 live births. The anomaly is thought to result from failure of normal embryonic separation of trachea and esophagus. Unlike atresia and tracheoesophagic fistulas, congenital stenosis often is not diagnosed until later in childhood and several cases have been reported in adults.[2] The question of whether "congenital" esophageal stenosis in adults really means "congenital" or just early onset of presently unknown causation has been recently posed.[3] In addition, esophageal stenosis, mimicking that of congenital type, may result from tracheobronchial remnants remaining with the esophageal wall after repair soon after birth. In such instances, dysphagia that occurrs later in life fails to respond to dilatation, but promptly improves after surgical resection of the stenotic segment. Unfortunately, the exact type of congenital stenosis could not be retrospectively identified in this patient.

With adequate insulfation during endoscopy the appearance of multiple rings with normal overlying mucosa can be seen in most cases of congenital esophageal stenosis. Segmental resection has been advocated for these patients; however properly performed bougienage has been reported to be safe in children and adults.[3] However, repeated bougienage in this patient resulted in esophageal wall rupture and subsequent surgical intervention with colon grafting.

In general, the stomach remains the first choice for an esophageal substitute. During colon grafting the overall complication rate can be up to 60%, including anastomotic insufficiency, fistulae, graft necrosis, arterial hypoperfusion and peritonitis.[4] Furthermore, secondary obstructive disorders of the esophageal substitute, including strictures[5,6] and diverticles[7-9] are generally surgically treated and this is what the patient was advised to do in case of petsisting symptoms. Despite these limitations, colon interposition for esophageal replacement is required in some cases. Some surgeons recommend the use of the colon graft especially in benign diseases of the esophagus since a better long-term function has been reported.[10-12]

Remarkably, in this patient there was evidence of colon graft ulcerations in the lower part of the colon graft. This mucosal damage is of exceptional interest in this kind of substitution surgery. In fact, thirty-six patients, subjected to colon interposition for benign esophageal disease or carcinoma of the esophagus or gastric cardia, were studied by endoscopy for signs of mucosal disease in the interposed colon. The alterations were unexpectedly few and mild considering the marked change in the location and function of the colonic segment.[13]

To the best of our knowledge this is the first reported case of Claude Bernard Horner syndrome as a long term complication of colonic graft diverticle after esophagectomy for congenital esophageal stenosis

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