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Case Report

Cricopharyngeal Achalasia in elderly: A rare cause of dysphagia- Case report

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ABSTRACT

Cricopharyngeal achalasia is a rare clinical condition in which upper esophageal sphincter does not open adequately during swallowing causing dysphagia. It can be primary or secondary. We report a case of cricopharngeal achalasia in a 76yr old Parkinsonian patient.

KEYWORDS: Achalasia, dysphagia, manometry, esophageal sphincter

INTRODUCTION

Cricopharyngeal dysphagia is characterized by difficulty in initiating swallowing. Attempt to swallow lead to coughing, choking, nasopharyngeal regurgitation, aspiration, and a sensation of residual food remaining in the pharynx. Cricopharyngeal achalasia is a disabling relatively uncommon cause of dysphagia and is usually not described in adults. Treatment of mild disorders of the upper esophageal sphincter (UES) is manageable by swallowing exercises and modification of nutrition, severe disorders in this area often require surgical myotomy of the cricopharyngeal muscle.

Disorders of the lower esophageal sphincter (LES), especially achalasia, can be treated with pharmacological agents, such as nitrates, calcium channel blockers, anticholinergic agents and beta-adrenergic agonists, but frequently do not provide satisfactory relief of symptoms[1,2]. Physicians should consider this also as a possibility as a cause for dysphagia. Here we report a case of dysphagia presenting with only complaints of difficulty in swallowing and was treated surgically with PEG.

CASE REPORT

76 yr old Mr.X is a known case of parkinsonism since 10 yrs very compliant with medications without any on and off phenomenon. He also a known case of hypertension since 15 years on regular medications. He presented to the outpatient department with complaints of odynophagia. He gave history of heart burn, choking sensation and pain during swallowing. He gave complaints of "sensation of food getting stuck" in the lower third of the neck. He was symptomatic since 2 months and was treated for gastroesphageal reflux disorder.

There was no history of coughing or nasal regurgitation of food. There was similar discomfort experienced with both solid and liquid food. There was no history limb weakness, ataxia or any hemisensory disturbance. No history of inflammatory joint pains, tightening of skin on the face or arms. There is no history of chronic acid peptic disease with reflux.

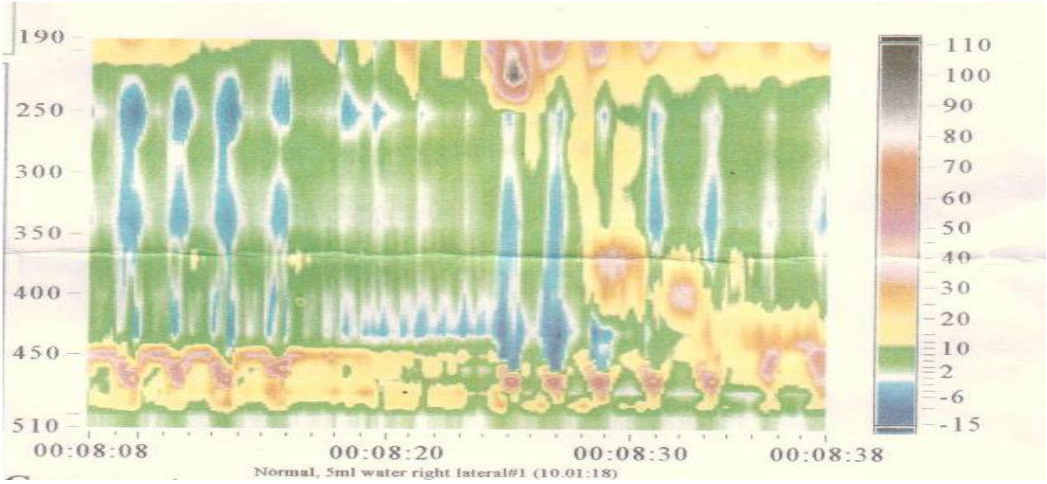
On examination there was no pooling of secretions with good gag reflex. Powers in all limbs were normal with both exteroceptive and proprioceptive sensations intact. His vitals were normal. All his blood investigations including complete blood count and liver and renal functions were normal. His chest X-ray was normal and did not show and mediastinal pathology or any mass lesion. His 2D Echo showed just concentric LVH with grade 1 diastolic dysfunction.

Upper gastro duodenoscopy was done which showed tight cricopharynx and the gastroenterologist found it difficult to pass the endoscope beyond cricopharynx. Hence esophageal manometry was done (figure 1) which showed a high basal

upper esophageal sphincter pressure of 50-70 mm of Hg and a normal lower esophageal sphincter pressure. Body peristalsis and propagation was normal with normal lower esophageal relaxation.

On attempted swallowing the upper esophageal pressure increased up to 90-100mm of Hg, hence confirming the diagnosis. Considering age and Parkinsonian state he was not advised pneumatic esophageal dilatation and he underwent feeding gastrostomy.

Figure 1: Esophageal manometry of the patient



Basal UES pressures are high ranging from ranging from 50-70 mm of Hg.Basal LES pressures are normal ranging from 30-35 mm of Hg.On attempted swallowing the UES pressures increase up to 90-100 mm of Hg and there are multiple attempts made at swallowing.Body peristalsis is normal and propagative LES relaxation is adequate.

DISCUSSION

The term Cricopharyngeal achalasia was first coined by Asherson[3] but its clinical features were first described by Brown and Kelly in 1919. The patients are usually above 40 years of age and most commonly over 60 [4]. Rarely, it may be present in infants also .Swallowing is normally an exceedingly well coordinated sequence of movement carried out by mouth, tongue, palate and pharynx which causes food and fluids to pass on towards into the upper esophagus. In the condition of cricopharyngeal achalasia, there is partial or complete failure of the sphincter to relax, or a delay in relaxation. Cricopharyngeal achalasia may be primary or secondary. Primary cricopharyngeal achalasia implies that the abnormality that leads to the persistent spasm or failure of relaxation of the cricopharyngeus muscle is confined to the muscle, with no underlying neurologic or systemic cause.

This primary group can be further subdivided into primary cricopharyngeal achalasia with no underlying cause (ie, idiopathic) or cricopharyngeal achalasia caused by intrinsic disorders of the cricopharyngeus muscle (eg, polymyositis, muscular dystrophy, hypothyroidism, inlclusion body myositis).In many instances, the cricopharyngeal spasm may be secondary to neurologic disorders such as polio, oculopharyngeal dysphagia, stroke, and amyotrophic lateral sclerosis (ALS). Peripheral neurologic disorders,such as diabetic neuropathy, myasthenia gravis, and peripheral neuropathies, can also cause cricopharyngeal dysfunction. The picture is analogous with achalasia of the cardia and achalasia of the pyloric sphincter. The mechanism is a neuromuscular imbalance with loss of normal reflex function. The achalasia is the result of a broken or 'imbalanced' reflex arc resulting in failure of relaxation of the horizontal fibres of the inferior constrictor muscle of the pharynx during the process of deglutition.

The diagnosis of cricopharyngeal achalasia can often be made by a carefully taken history and physical examination. Endoscopy and radiographic examination of the pharynx, larynx, trachea and esophagus are essential, however, since by these examinations the presence or absence of a pharyngeal diverticulum can be determined, and other causes of dysphagia, such as carcinoma of the proximal esophagus, can be excluded. In patients with cricopharyngeal achalasia, the characteristic radiographic features in the pharynx are:

a)The presence of posterior shelf like semicircular filling defect caused by hypertrophy of the cricopharyngeus muscle.

b)Pooling of barium in the vallecular folds which gives rise to a "cross-bow" appearance in the antero-posterior view of the pharynx

c) Distension of the hypopharynx during swallowing and delay in the passage of barium into the esophagus; and

d) Folding and redundancy of the pharyngeal mucous membrane, which in some cases simulates a web in appearance.

Treatment is directed by the severity of symptoms. Injection with botulinum toxin has been beneficial as a temporizing treatment in adults with cricopharyngeal achalasia, as well as serving as a diagnostic test[5]. In a small study,

percutaneous injection of botulinum toxin into the cricopharyngeus muscle resulted in improvement of swallowing in all treated patients. The patients who responded to an injection of botulinum toxin could receive repeated injections or myotomy should symptoms recur.

However, if no clinical response was noted after botulinum toxin injection, then myotomy would not be indicated, the rationale being that relief of the hypertensive cricopharyngeus muscle with botulinum toxin should result in improved swallowing; if injection was not followed by clinical improvement, the swallowing dysfunction is not due solely to achalasia.

In such cases, surgery would not be addressing the complete problem. Dilation by bouginage has been shown effective for the relief of symptoms associated with cricopharyngeal achalasia, even in children[6].Development of balloon dilation has maintained effectiveness while improving safety, often requiring a single treatment**[**7].In a small adult study, the maximum balloon size required for symptomatic relief was 20 mm. In severe or recurrent achalasia, surgical intervention is warranted; myotomy of the UES is typically used[6]. Patients tend to have dramatic and complete, long-term symptomatic relief after myotomy[8].

CONCLUSION

In our everyday practice, the majority of deglutition disorders in elderly occur secondary to cerebrovascular accident. They would have other associated symptoms with the dysphagia. However when an isolated symptom of dysphagia is present Cricopharyngeal dysphagia should be kept as a part of differential diagnosis.

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