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Inclusion body myositis- A rare cause of dysphagia

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Abstract: Inclusion body myositis is a rare cause of dysphagia. We report a case of 74 year old Caucasian lady, who has a background of inclusion body myositis and presented with a history of chronic dysphagia. She noticed sudden worsening of symptoms. Flexible nasoendoscopy showed normal larynx with pooling of saliva in pyriform fossa as a result of obstruction in oesophagus. Barium contrast swallow study showed complete obstruction at the level upper oesophageal sphincter. She was treated with balloon dilatation of cricopharyngeus inlet. She noticed improvement in swallow following the balloon dilatation treatment. 9 months after treatment she still continues to swallow normally. This case report focuses on etiology, pathophysiology, presentation and management aspects of dysphagia associated with inclusion body myositis.
Keywords: Dysphgia in elderly, inclusion body myositis.

INTRODUCTION:

Inclusion body myositis (IBM) is a rare chronic progressive myopathy. Prevalence of IBM can vary up to 35 per million adult over age of 50 [1]. There are two types of IBM; Sporadic inclusion body myositis (sIBM) and hereditary inclusion body myopathy (hIBM) [2]. The sporadic type is more common [2]. The clinical presentation involves slow, steadily progressive muscle weakness [1]. The pattern of muscle involvement is distal asymmetric weakness affecting the finger flexors and proximal lower extremity weakness affecting the quadriceps [1]. As disease progresses, proximal muscles also get involved [1]. It may sometime take years before patient notices any significant deterioration in function leading to seek medical advice [1]. The pathophysiology of s-IBM is still poorly understood, and currently there is no known treatment to cure [1]. Dysphagia is common in 44-66% of patients with well-established IBM and can be a presenting feature [3].

CASE REPORT:

A 74 old Caucasian lady presented with history of dysphagia for six months. She was known to suffer from Inclusion body myositis for the past 8 years. She has ongoing swallowing difficulty, making her only able to eat soft diet. Symptoms suddenly worsen overnight, when she developed total dysphagia and admitted to hospital. Flexible nasoendocpy showed pooling of saliva with normal laryngeal structures. Barium swallow showed complete obstruction at the level of cricopharyngeus muscle (see figure 1). A percutaneous endoscopic gastrostomy tube was inserted for feeding purpose during hospital stay and she was discharged home subsequently. Her case was discussed in gastrointestinal multidisciplinary team meeting. Based on radiologic features and examination findings, she was diagnosed as severe criopharyngeal spasm and sphincter dysfunction secondary to inclusion body myositis. She underwent panendoscopy (see figure 2) and balloon dilatation of cricopharymgeus inlet. There was a complete stenosis of the cricopharyngeal inlet. This area was dilated using two cook medical balloon of 18mm in diameter each simultaneously. There was only slight improvement in swallow following first procedure. Subsequent to that, she was booked for an MRI scan of her neck. MRI scan showed thickening of upper oesophagus just below cricopharyngeus without showing any other serious pathology. She underwent repeat panendoscopy and balloon dilatation. Pan endoscopy showed prominent cricopharyngeus bar. Upper oesophageal sphincter was dilated again using double balloon, each balloon inflating up to 18mm (see figure 3 and figure 4). Swallowing became normal following second balloon dilatation. A Biopsy was taken and histology showed chronic nonspecific inflammation consistent with IBM. She managed to swallow normal food and sensation of blockage was

completely gone. Initial review after one month showed patent cricopharyngeal inlet (see figure 5) and she was able to swallow normally. She was reviewed in outpatient clinic 5 months after the procedure and she was able to swallow normally. Now 9 months after dilatation of cricopharyngeus sphincter, we arranged a telephone consultation to reassess her symptoms and she still continues to swallow normally.

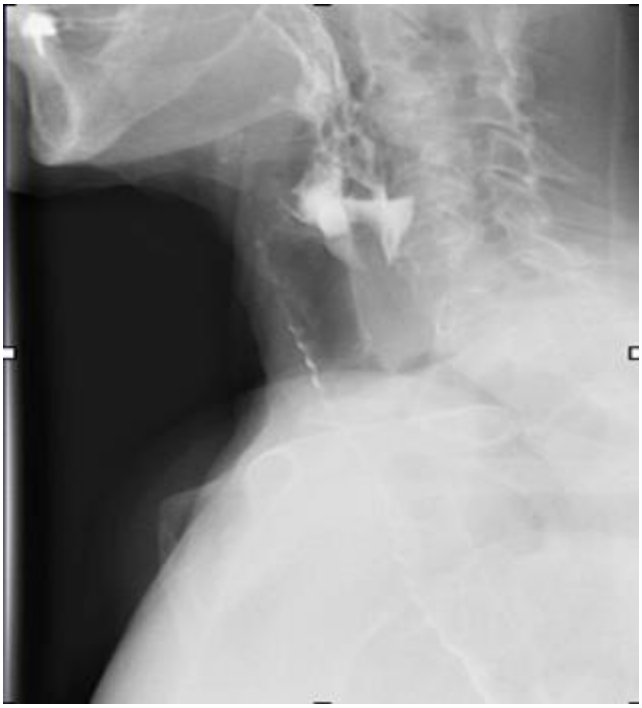

Fig 1: Barium swallows study showing complete blockage at the level of cricopharyngeus muscle

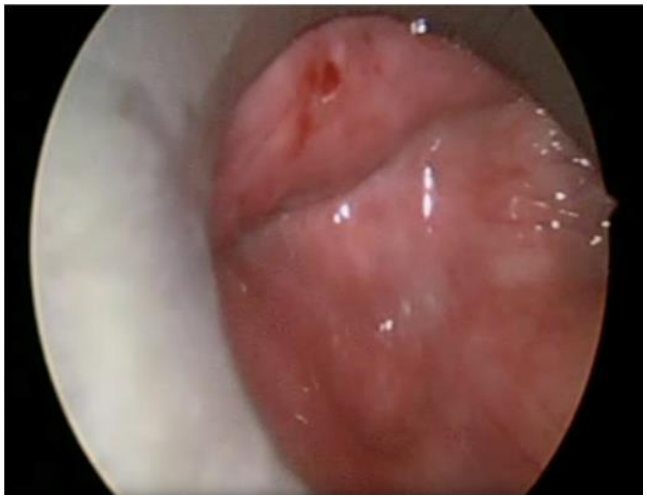

Fig 2: Upper oesophagoscopy showing complete obstruction of cricopharyngeal inlet

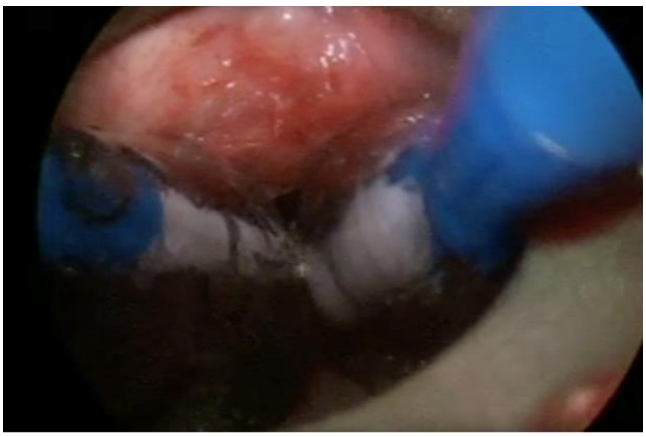

Fig 3: Upper oesophagoscopy and double balloon dilatation of cricopharyngeal inlet

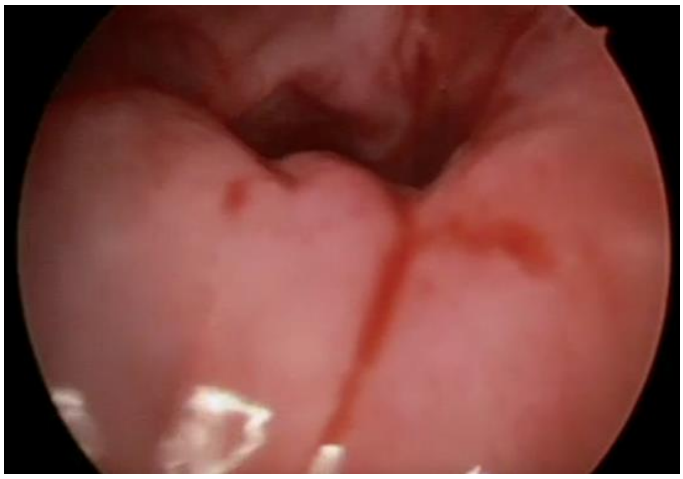

Fig 4: Appearance of cricopharyngeal inlet post dialatation

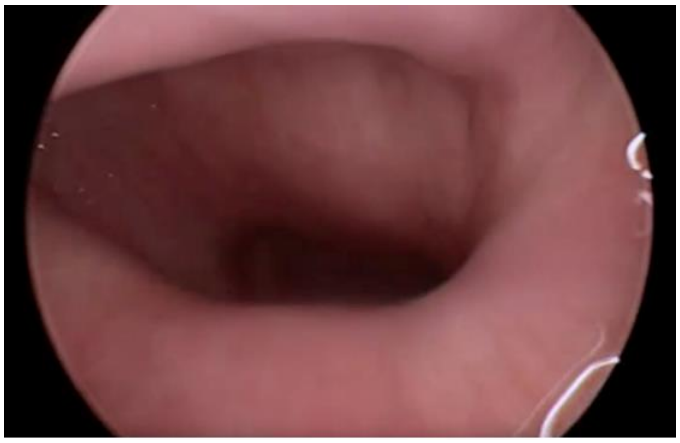

Fig 5: Appearance of cricopharyngeal inlet one month after dilatation

DISCUSSION:

In year 1971, the term inclusion body myositis was used for the first time by Ynuis and Samaha [6]. The term was used to describe a type of myopathy which phenotypically resembled chronic polymyositis but histological examination revealed cytoplasmic vacuoles and inclusions [6]. S-IBM should be considered as an important differential diagnosis when evaluating an older Caucasian patient with progressive muscular weakness [6].

Familial or hereditary IBM (h-IBM) could be differentiated with sporadic form by the absence of inflammation [6]. Familial IBM is classified into autosomal recessive form and autosomal dominant form [5, 6]. Gene mutations of chromosome 9 and chromosome 17 have been described as possible aetiology.

Sporadic IBM (s-IBM) commonly designated as just "IBM", is now considered as most common acquired inflammatory myopathy in patients above the age of 50 years [6,7]. Theories proposed about pathophysiology of disease include: muscle degenerative disorder, or triggered by a virus or an autoimmune disorder [5, 6]. IBM is more prevalent in males as compared to females and white as compared to blacks [5,6]. S-IBM is classified under the common umbrella of 'idiopathic inflammatory myopathies' (IIM) along with polymyositis, dermatomyositis and the rarer eosinophilic polymyositis and focal myositis [6]. These myopathies differ from one another on the basis of pathogenicity and histology [6].

Disease process includes patchy involvement of muscular tissue so repeated biopsy should be considered [6]. Light microscopy shows ragged-red, atrophic muscle fibres, with lymphocytic mono nuclear inflammation and muscle fibres with vacuoles containing red and green staining material with the Congo-Red stain (60– 80% of the vacuolated muscle cell fibres and denoting amyloid) [6]. Electron microscope shows 15–18 nm diameter paired helical filaments [6]. Vacuolated muscle fibres of IBM contain prion protein, acetylcholine receptor and proteins that are typically characteristic of Alzheimer brain (β-amyloid, N- and C-terminal epitopes of β-amyloid precursor protein, alpha 1 antichymotrypsin, phosphorylated tau, apolipoprotein E and ubiquitin) [6].

Sporadic IBM tends to progress gradually over the period of time. Muscular atrophy and weakness is typically asymmetric [6, 9]. Characteristic pattern is early involvement of distal musculature [1]. Quadriceps, iliopsoas, ankle dorsiflexor and volar forearm muscles typically gets affected first [1].

Complain of swallowing difficulty is noticed in 40-50% of cases although >80% of patients get some degree of narrowing of lumen of oesophagus on contrast swallow [6]. Inflammatory reaction of cricopharyngeus muscle is considered as the cause of dysphagia, as evidenced by presence of inflammatory or rimmed vacuole in biopsy specimen taken from cricopharyngeus muscle [7]. Inflammation not only reduces lumen but also affects the function of oesophageal sphincter [7]. Dysphagia tends to get worse with time in majority of the cases. Associated risk of aspiration pneumonia tends to reduces life expectancy [7, 8]. Severe dysphagia could be a cause of social embarrassment, resulting in having meals in isolation due to audible deglutition [7].

Treatment of dysphagia includes oesophageal dilatation and surgical myotomy. In some patients local injection of Botulinum A is found to be beneficial [6]. Combination of intravenous imunnonglobulins (IVIg) and prednisolone has shown benefit in cases of severe oesophageal constriction of life threatening nature [6].

In general disease process is slowly progressive. Individuals with progressive dysphagia tend to have poor outcome and poor quality of life as compared to individuals with non-progressive dysphagia [6].

CONCLUSION:

Inclusion body myositis is a rare myopathy. The disease is more common in caucasians population over 50 years of age. Dysphgia is a common problem associated with the disease involving cicopharyngeus muscle in upper oesophagus. We recommend oesophageal dilatation as we have demonstrated in our case report as the first treatment as this is minimally invasive and can be repeated if necessary. Other treatment options include cricopharyngeal myotomy, percutaneous gastrostomy feeding tube. Disease tends to progress slowly with passage of time so long term follow up may be required.

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