ejpmr, 2018,5(8), 42-45

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

A CASE OF ACHALASIA MISDIAGNOSED AS UVULITIS IN A 41 YEAR OLD WOMAN

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Article Received on 26/05/2018 **Article Revised on 16/06/2018** **Article Accepted on 06/07/2018**

ABSTRACT

The rarity of achalasia in Nigeria and its misconceptualized knowledge and treatment may result in missed diagnosis with the attendant risk of malignant transformation. This is a case report of a 41 year old woman diagnosed with achalasia, confirmed using various radiological modalities but previously misdiagnosed as a case of Uvulitis. The role of the physician and radiologist in contributing to intense medical awareness with a bid to discourage the ancient practice of crude uvulvectomy cannot be over emphasized. Prevention of complications and death from preventable pathologies like achalasia should be the ultimate goal of every physician.

KEYWORDS: Achalasia, lower eosophageal sphincter, uvulitis.

INTRODUCTION

Achalasia is a Greek word for "does not relax''. It is a well-known eosophageal motility disorder involving the smooth muscle layer of the distal esophagus characterized by reduced to absent primary peristalsis, incomplete relaxation of the lower esophageal sphincter in response to deglutition and lack of a coordinated lower oesophageal sphincter relaxation in response to swallowing.[1,2] Achalasia has been classified as primary or secondary[3,4,5]

Primary achalasia has no known underlying cause or precipitating factor hence also known as idiopathic achalasia and present clinically with dysphagia (difficulty swallowing), odynophagia (pain when swallowing) and/or chest pain. The index patient had dysphagia and chest pain, both of unknown cause.

Secondary achalasia is relatively uncommon and is due to recognized pathologic causes of esophageal motility disorders often indistinguishable from primary achalasia. Secondary achalasia condition exists when a process other than intrinsic disease of the esophageal myenteric plexus is the etiology. These conditions may include Collagen vascular disease (scleroderma), Diabetes, Chagas' Disease, Amyloidosis, Alcoholism, Aging process (presbyesophagus) and Invasive Cancer.[3,4,5]

Achalasia is a rare motility disorder which can occur at any age but is rare before adolescence. It is most frequently seen in middle and late adulthood (30-70 years of age) with no racial nor gender predilection with an annual incidence of 1 case per 100,000 and a prevalence rate of 1: 10,000. In sub Saharan Africa, 3-4 patients are seen annually.[6]

The rarity of this condition in Nigeria and misconceptualized knowledge and treatment of achalasia which may result in missed diagnosis and risk of malignant transformation, necessitated the report of this case.

CASE REPORT

A.W is a 41 year old house wife who presented at the medical out -patient department of the University of Calabar Teaching Hospital, Calabar, with a 2 years history of gradual onset of dysphagia, initially to solids and later to liquids. There was no history of odynophagia. She also gave a history of easy satiety, a feeling of fullness in the chest after eating and burning chest pain often radiating in between the shoulder blades and was also said to have lost considerable weight evidenced by her loosely fitted skirts. Patient was said to have undergone traditional Uvulectomy about six months prior to presentation in clinic after which she was given herbal drugs for a period of one month. There was no antibiotic administration after the procedure and patient was said to have suffered very high fever which lasted for more than a week after the procedure, most probably due to infection.

Physical examination revealed a chronically ill, cachectic young woman, mildly pale, anicteric, afebrile with normal vital signs.

No significant findings were seen in the body systems examined. A tentative diagnosis of achalasia was made

and chest x-ray, and barium swallow and upper GI endoscopy were requested.

Chest radiograph (Fig.1) revealed a widened mediastinum, more on the right side with a tubular differential soft tissue density seen posterior to the heart not silhouetting the heart with its upper extent at the level of the aortic arch and the lower extent seen inferior to the diaphragmatic outline; the cardiac size was within normal limit.

Barium swallow showed a grossly dilated esophagus with significant tertiary peristalsis; hold up of contrast and tapering of the distal esophagus, giving the "rat tail appearance" (Fig. 2).

Endoscopy done for the patient showed retained saliva with puckered gastroesoghageal junction.

Patient was re-evaluated and booked for surgery five weeks after presentation. The modified Heller's oesophagocardiotomy was done for the patient. The repeat chest x-ray was satisfactory. Patient was discharged home, however lost to follow up.

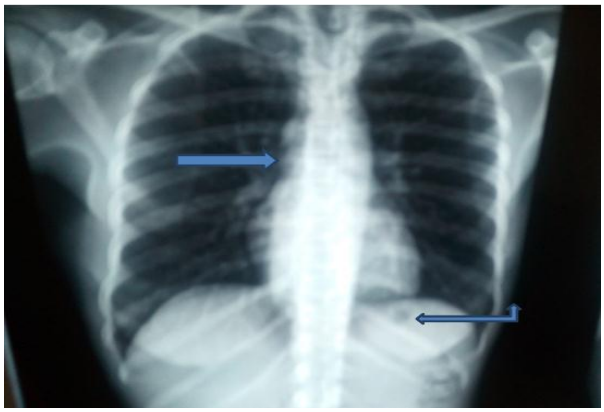

Fig.1: Posterioanterior chest radiograph shows a widened mediastinum, more rightward (see horizontal arrow. The angled arrow shows a small fundal gas shadow beneath the left hemidiaphragm.

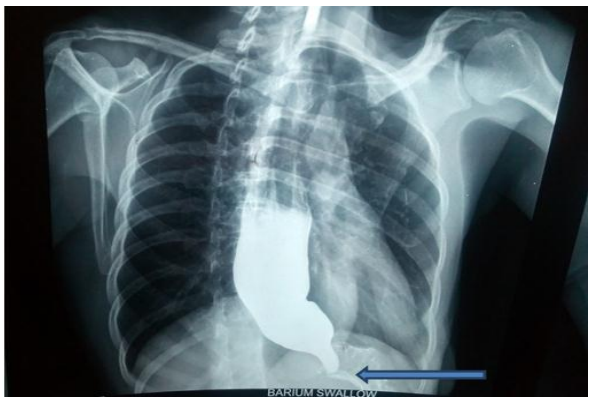

Fig. 2: Left anterio-oblique view of barium swallow showing a dilated tortuous barium-filled esophagus with narrow tapering at its distal end, producing a rat tail appearance.

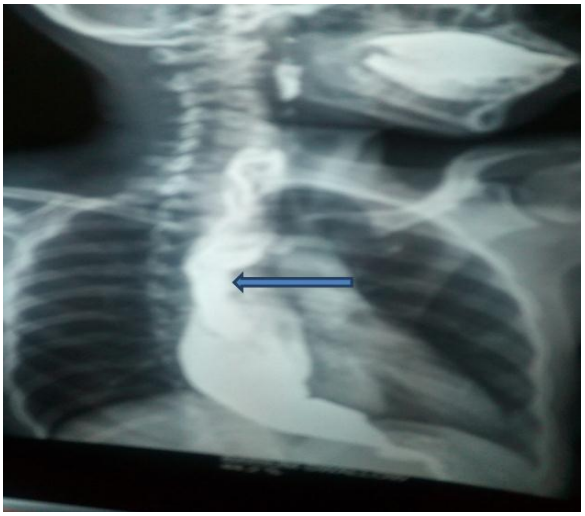

Fig. 3: Left anterio-oblique view of the esophagus shows grossly dilated contrast filled esophagus with wavy outline and mottled lucencies within suggestive of contrast hold up due to food debris along its mid portion (see arrow).

DISCUSSION

Achalasia is a primary motor disorder of the esophagus characterized by insufficient lower esophageal sphincter relaxation and loss of esophageal peristalsis. This results in patients complaints of dysphagia to solids and liquids, regurgitation, and occasional chest pain with or without weight loss or barium swallow showing dilated esophagus with rat tailing in a symptomatic patient should prompt appropriate diagnostic and therapeutic strategies.[3,4] The index case had progressive dysphagia to solids and later liquids and occasional chest pain and significant weight loss.

Uvulectomy by traditional practitioners in Africa has been an age-long practice.[7,8] It involves the complete or partial removal of the uvula. It is a common procedure in Nigeria and several other African countries with documentations of this practice in countries outside Africa like Israel, Saudi Arabia, and some Middle Eastern countries.[7,8]

These practitioners are primarily traditional healers or lay men or women who double as barbers performing their acts with a sickle knife and other unsterilized instruments.[6] The commonest indications include pains in the throat, dysphagia, cough, and loss of appetite recorded. Some of these traditional practitioners' patients do well after the procedure but this unwholesome, unscientific, and unsupervised practice is potentially dangerous resulting in complications such as hemorrhage, anemia, septicemia, tetanus, risk of the Human Immunodeficiency Virus (HIV) infection, and death. This index patient most probably suffered from infection and her earlier presenting symptoms of dysphagia to both solids and liquids persisted. Therefore, this practice should be actively discouraged by media health campaigns.

Achalasia misdiagnosed as a tumour and treated by traditional uvulectomy in this case reported results from degeneration of intramural innervation in the esophagus and decreased numbers of ganglion cells in myenteric plexus in patients. The remaining ganglion cells in myenteric plexi are surrounded by lymphocytes and eosinophils. These Inhibitory neurons are selectively destroyed (nitric oxide producing neurons) net increased tone in LES and aperistalsis in esophageal body.

Clinically, diagnosis is usually reached based on the presenting symptoms which may include dysphagia to solids and later progressively to liquids; odynophagia, recurrent regurgitation and chest infections (basal pneumonitis from frequent aspirations). Chest pain, mostly retrosternal is a prominent feature, often sequel to gastro esophageal reflux.[1-4] Difficulty in belching and weight loss is seen in 85% and 68% of patients respectively.[1,3] Weight loss was seen in the index case.

The diagnosis and management of achalasia can be done conveniently without misinterpretation as in this case reported using the following modalities namely plain chest radiography, barium swallow, computerized tomography, magnetic resonance imaging as well as manometry and upper gastro intestinal endoscopy.

The diagnosis of achalasia is made easy with detailed chronological medical history, radiography and esophageal motility testing. Patients with dysphagia should be properly investigated to prevent misdiagnosis as in the case presented. A number of entities may mimic achalasia, forming the so called 'achalasia pattern'. In achalasia**,** the distal segment of narrowing is less than 3.5 cm, in reflux esophagitis (long segment stricture and hiatus hernia); scleroderma (GOJ will be open; less severe dilation); esophageal malignancy or gastric (barium swallow and endoscopy will show esophageal obstruction by the tumour) carcinoma: commonly referred as pseudoachalasia; esophageal stricture and Chagas' disease are all possible differentials.

The radiologic examination of choice in the diagnosis of achalasia is a barium swallow study performed under fluoroscopic guidance.[3.5,6] The diagnosis of achalasia supported by the results of radiologic studies must always be confirmed by performing upper gastrointestinal endoscopy and esophageal manometry. These tests allow the direct evaluation and inspection of the esophageal mucosa and an objective measurement of esophageal contractility.[6]

Esophageal manometry is the gold standard for diagnosis of achalasia. The three classic manometric features in achalasia are: a) aperistalsis of the esophageal body, b) an elevated LES pressure greater than 45 mmHg (normal 15-30 mg Hg), and c) impaired relaxation of the LES during swallowing.[7.9] Manometry was not done for this index patient due to non-availability¨

The value of endoscopy in patients with achalasia is not to make the diagnosis, but to exclude other disease entities and to diagnose complications. The endoscopic findings in primary achalasia are normal mucosa and a mild to moderate degree of resistance in passing the endoscope through the EG junction. The endoscope allows visualization of mass and mucosal lesion and tissue specimens may be obtained.[10]

Plain chest radiography often reveals a widened mediastinum caused by a fluid filled esophagus; a double mediastinal stripe is also occasionally seen and in severe disease a large, dilated esophagus with air fluid level at or above level of the aortic arch; gastric air bubble frequently small or absent.[1,9,10] The gastric air bubble was small in the index case.

Barium studies gives 95% diagnostic accuracy as early stages caused by Primary peristaltic waves absent with abnormal distal peristalsis results in only minimal narrowing of the gastroesoghageal (GE) junction; Occasionally non-propulsive peristaltic waves in the esophageal body ("vigorous achalasia" secondary to tertiary waves) are seen. As the disease progresses, the "Bird's beak "or rat tail appear at the GE junction where the distal esophagus makes right angle before entering stomach.

The dilated, aperistaltic esophageal body; may assume a sigmoid shape. In Severe cases, significant esophageal body dilation with large amounts of fluid/food retention is noted and the entire esophagus becomes atonic in late stages.

Computerized (CT) findings are nonspecific as diagnosis cannot be made using only CT. Contrast enhanced CT may help demonstrate structural esophageal abnormalities such as the dilated luminal structure with retained debris and narrowing at level where it enters the stomach, a feature often seen in advanced stages.[10]

CT and MRI may not be indicated in routine cases of primary achalasia, except in complicated cases or in secondary achalasia to help determine the specific disease entity which resulted secondarily in achalasia. They can be vital, however, in complicated cases to help confirm the diagnosis or to detect atypical features. Features of symmetric or asymmetric wall thickening, mass lesion involving the esophagus or causing its extrinsic compression or adenopathy raise suspicion of secondary achalasia.[9,10] Because no specific radiographic or clinical criteria are established which predictably distinguish primary achalasia from secondary achalasia in all instances, diagnosis may require use of numerous modalities. Patients with long-standing primary achalasia are predisposed to development of infection and esophageal carcinoma. Increase in cancer and infection risk is thought to result from inflammation caused by chronic stasis.[9]

Several management options currently includes palliative care which involves systematic relieve of the syndrome such as endoscopic pneumatic dilatation or surgical myotomy. Life style modification is encouraged and involves eating slowly, chewing food very well, drinking plenty of water with meals and avoiding food before bed time.[1,5,10]

CONCLUSION

This is a case report of a 41 year old woman diagnosed with achalasia, confirmed using various radiological modalities but previously misdiagnosed as a case of Uvulitis. The role of the physician and radiologist in contributing to intense medical awareness with a bid to discourage the ancient practice of crude uvulvectomy cannot be over emphasized. Prevention of complications and death from preventable pathologies like achalasia should be the ultimate goal of every physician.

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