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Childhood Achalasia: Barium Swallow is Gold Standard Even before Histological Confirmation –Case Report.

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ABSTRACT

Achalasia in pediatric age group is rare and the causative aetiology is neurodegeneration of the neuro-myenric plexus at the lower oesophageal sphincter (LES). This may not be diagnosed properly because of inherent problems for the investigations in children. We present 14-years old boy who had been having frequent retrosternal chest pain with off and on vomiting of three years duration. There was no relief by the antacids and other symptomatic medication. He underwent barium swallow for the study of oesophagus and was diagnosed to be having oesophageal achalasia.

Keywords: Achalasia, Neuromyentric plexus, LES, Barium swallow.

INTRODUCTION

Achalasia in children can present symptoms at any age beyond five years of the age .The incidence is 0.11 in per lac population. Only less than 5% children below 15 years of age present with symptomatology. This is a neurodegeneration of the myenteric complex at the lower end of the oesophagus in the lower eoesophageal sphincter (LES). The exact diagnosis poses many problems because of variety of tests available in the modern world. We have seen that barium swallow is the gold standard which gives classical findings by which the diagnosis can be made with much of surety.

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CASE REPORT

14-years old boy reported to the children out patient department with complaints of Dysphagia and retrosternal pain of three years duration. There is history of taking antacids and painkillers in the past on symptomatic manner. He had lost 8 kg of weight in the last two years. There was no history of ingestion of any drugs responsible for the present symptoms. On examination, he was of averagely built with normal physical parameters as per the age. His present body weight was 32 kg which was low as per the standard chart. Systemic examination was unremarkable. Plain chest x-ray did not reveal any abnormality. His haemoglobin was 10 g/dL .Rest of all the biochemical investigations were within normal limits. He was subjected to barium swallow examination and the result had shown dilated thoracic oesophagus with rat tail appearance of the distal end [Figure 1 a,b & Figure 2].

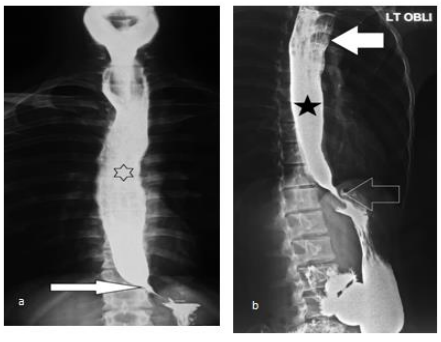

Figure 1: Barium swallow for oesophagus.(a) anteroposterior view shows dilated oesophagus (black hollow star) with "rat tail" appearance of lower oesophageal sphincter (white arrow).(b) left oblique view shows dilated oesophagus with secondary peristalsis (white arrow) with dilated oesophagus (black star) with narrow lower end of oesophagus having "bird's beak" appearance (white hollow arrow).

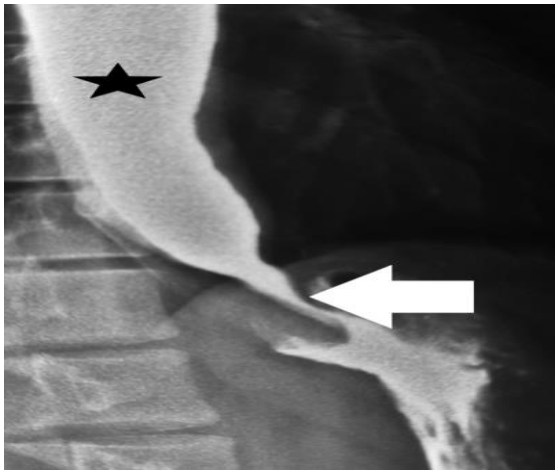

Figure 2: Barium swallow magnified left oblique view of the lower end of oesophagus. Dilated oesophagus (black star) is seen with classical "rat tail" appearance in barium swallow (white solid arrow).

In the same sitting limited computerized tomography sections were acquired with 3D reconstruction to rule out any additional findings in the surrounding regions [Figure 3 a,b & Figure 4]

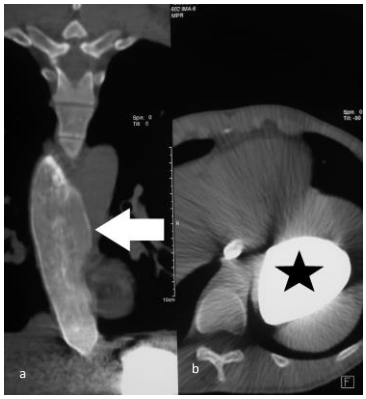

Figure 3: Computerised tomography (CT)3D reformatted images.(a) coronal section shows dilated oesophagus (white solid white arrow).(b) axial mid-oesophagus section shows dilated oesophagus without any extrinsic mass (black star).

Endoscopic biopsy had confirmed the diagnosis as cause being idiopathic in one of our sister concerned hospital. Patient had been contemplated for pneumatic dilatation in one of our sister concerned hospital. The patient will also undergo biopsy from the lower end for histological confirmation of diagnosis. We were sure in this case of the diagnosis as the barium investigation had shown classical appearance.

DISCUSSION

Achalasia is related to non relaxation of the lower end of the oesophageal junction. There are various variety of inhibitory and excitatory reflexes originate at the neuro-myenteric plexus.[1] The inhibitory reflexes fail to operate because of the neurodegeneration which leads to this entity. This can lead to the dilatation of the distal part of the oesophagus [Figure 5].

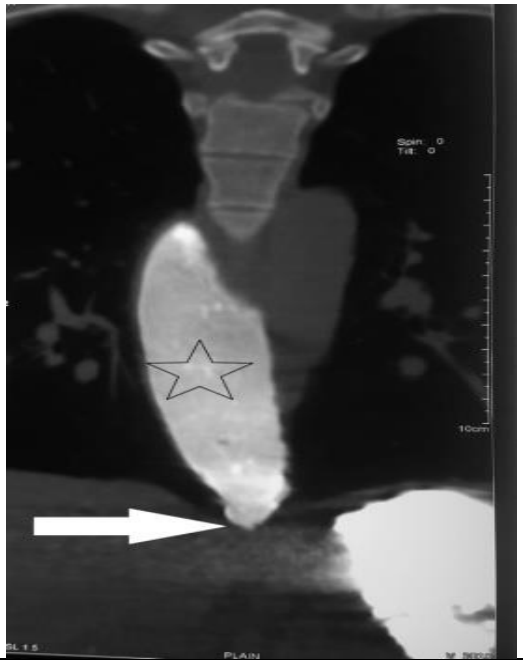

Figure 4. CT reformatted coronal section shows dilated upper part (black star) and with narrowed non relaxing lower end of the oesophagus (white arrow).

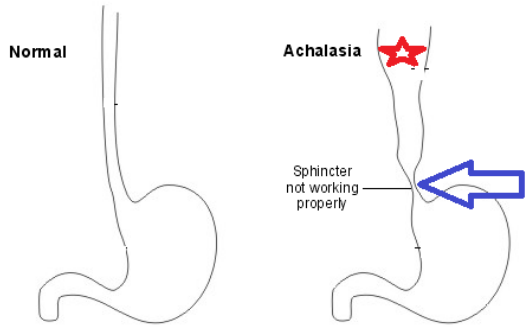

Figure 5: Diagrammatic representation of achalasia as compared to normal structure. The upper part of the oesophagus is dilated (red star) and the sphincter at the lower end is not functioning properly (blue hollow arrow).

The aetiology in most of the cases is idiopathic. There are many other diseases which can also cause similar type of conditions viz mixed connective tissue disease like scleroderma, oesophageal cancer, Trisomy 21, congenital hypoventilation syndrome, familial dysautonomia, Chaga's disease and few other syndromes. Pathophysiology remains as the increased sphincteric pressure and failure to relaxation. This leads to proximal oesophageal dilatation. Initially the diagnosis is mistaken as gastrooesophageal reflux disease (GERD) and

children are treated with antacids which delay the diagnosis. Dysphagia, feeding difficulties and vomiting are the frequent complaints. This usually leads to the weight loss. Some children can present with aspiration pneumonias and hoarseness.[2]

The diagnosis is confirmed by barium swallow and can be confirmed by manometric studies. The classical presentation of the study is "rat tail" or "bird's beak' appearance of the lower end of the oesophagus. The proximal part of the oesophagus shows dilatation and some residual material may be seen in that. Endoscopy is avoided in younger children as barium studies shows ample evidence of underlying aetiology.[3]

Management has got different options as medical, endoscopic, pneumatic dilatation and surgical. Calcium channel blockers like nifedipine is used for the relaxation of LES but the results are not that encouraging compare to other managements. Endoscopic botulinum toxin injection at LES may give temporary relief but because of frequent intervention this is also not considered as the choice of management. Pneumatic dilatation is advocated in children > 8 years old but this is also accompanied with some complications like rupture and retrosternal pain.Laproscopic Hellar Myotomy (LHM) remains the management of choice because of definitive results and cure.[4] Pasricha et al (2007) had shown peroral endoscopic myotomy (POEM) by natural oral transluminal endoscopic surgery (NOTES) as the most favorable surgical management in adolescent paediatric age group. There are minimum complications in this procedure.[5]

CONCLUSION

Children Achalasia remains undiagnosed for a long period because of the lack of investigating the entity. Child is subjected to barium swallow study when there is no relief and the diagnosis is confirmed. Endoscopic interventions are avoided for the diagnosis. Once the diagnosis is made than surgical management is the best option for the permanent relief.

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