Case Report

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

B B Sharma1, Shashi Sharma2, Naveen Bhardwaj3, Sakshi Dewan4, Mir Rizwan Aziz4

1Professor & HOD, Dept. of Radio-diagnosis, SGT Medical College Budhera (Gurgaon) Haryana 122505.
2Associate Professor, Dept of Pediatrics, SGT Medical College Budhera (Gurgaon) Haryana 122505.
3Assistant Professor, Dept. of Radio-diagnosis, SGT Medical College Budhera (Gurgaon) Haryana 122505.
4Senior Resident, Dept. of Radio-diagnosis, SGT Medical College Budhera (Gurgaon) Haryana 122505.

Received: February 2017
Accepted: March 2017

ABSTRACT

Achalasia in pediatric age group is rare and the causative aetiology is neurodegeneration of the neuro-myentric 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 oesophageal 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.

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).

Name & Address of Corresponding Author
Dr. B B Sharma,
Professor & HOD,
Dept.of Radio-diagnosis,SGT Medical College Budhera (Gurgaon) Haryana 122505
C 35 First Floor,Anand Niketan,New Delhi - 110021
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. 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].

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 hyperventilation 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.\(^4\)

Conclusions

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.

Acknowledgement

We are thankful to Mrs Shilpa, Mr Nitish Virmani, Mr Rajdeep Thidwar and Miss Nitika of the department of Radio-diagnosis SGT University, Budhera (Gurgaon) for carrying out the investigation and providing the images.

REFERENCES