Chiari Malformation Type 1 as Cause of Recurrent Pneumonia with Failure to Thrive

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Abstract

Diagnosing the cause of recurrent pneumonia with failure to thrive (FTT) can be difficult. A nine months age presented with recurrent pneumonia and failure to thrive. Investigation for the same led to the diagnosis of Chiari 1 malformation. Screening for latent neurological disorders should be done in such patients.

Key words: Recurrent, Pneumonia, Failure to thrive, Chiari 1 malformation

Introduction

Recurrent pneumonia is defined as more than one episode of pneumonia in one year or more than three episodes in a lifetime with documented radiologic clearance in between[1]. It is often a challenge to diagnose the aetiology.

The Case

Nine months old male child presented with complaints of cough and mild to moderate respiratory distress with failure to thrive. He was previously admitted at our institute at three months of age and was under follow-up over the last seven months. He frequently suffered from cough and cold and during which six sequential chest X-Rays done by family physician which showed no evidence of hyperinflation or consolidation.

Child was born at term with birth weight 2.6 kg and had uneventful perinatal history. However his body weight at three months of age was 3 kg and at nine months of age was 4.7 kg, which was less than the 1st percentile of the IAP standard[2]. However the family was affluent, the parental bonding was observed to be affectionate and there was no history of chronic vomiting or diarrhea. Child was formula fed from birth.

Investigations for pneumonia, namely blood count, CRP, ESR, Mantoux test, sweat chloride, milk scan (to check for gastro-oesophageal reflux), echocardiography and high resolution CT thorax were normal. Gastric lavage and induced sputum were negative for tuberculosis. Bronchoscopy done at four months of age was non-contributory. Broncho-alveolar lavage was negative for infections. Immunoglobulin levels, T, B, NK cell profile and Nitro blue tetrazolium test were all normal, thereby excluding any immunodeficiency.

Investigations for failure to thrive, namely arterial ammonia and lactate, thyroid profile, anti tTg IgA, Serum total IgA, blood glucose, urine reducing substance, amylase, lipase, stool microscopy, HIV screening were non-contributory. Bone age matched chronological age.

The child received various antibiotics, intravenous fluconazole and anti-reflux medications along with other supportive treatments but did not show satisfactory recovery. Finally nasogastric feeding was started, with complete stoppage of feeding through mouth. Subsequently the child improved remarkably. Upon starting oral feeds he deteriorated again.

A barium swallow was also done which failed to detect any significant gastro-esophageal reflux. Upper gastro-intestinal endoscopy was also normal.

Although the neurological examination was apparently normal, we wanted to look for any neuromuscular weakness that could contribute to oropharyngeal motor incoordination. Lactate dehydrogenase and creatine phosphokinase in blood as well as electromyography and nerve conduction velocity studies were normal, showing that the problem cannot be attributed to the peripheral nerves and muscles. Then we did an MRI of the brain which showed Chiari 1 malformation with minimal tonsillar descent.

Child was subsequently referred for neurosurgical intervention with the nasogastric tube in situ. The neurosurgeons advised that the child should be fed by nasogastric tube or a percutaneous endoscopic gastrostomy tube for the next three to six months, in the hope that with increase of body weight, his oropharyngeal muscles may acquire a better tone and consequently, dysphagia may resolve.

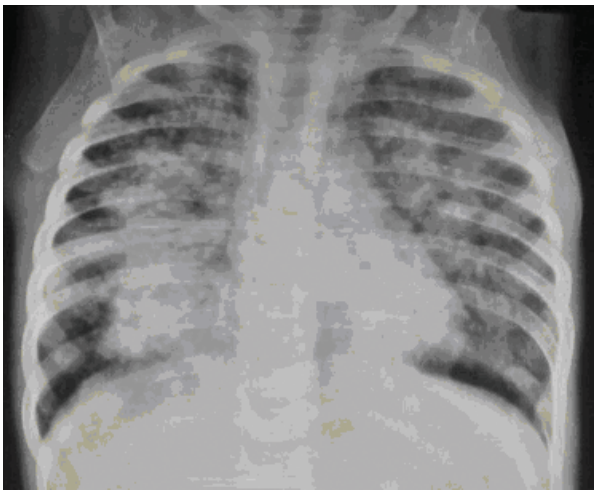

Fig 1: Showing bilateral pneumonia in the patient

Discussion

Recurrent pneumonia is defined as more than one episode of pneumonia in one year or more than three episodes in a lifetime with documented radiologic clearance in between[3]. It is often a challenge to diagnose the aetiology.

A common cause of recurrent pneumonia is aspiration[4]. Abdullah et al[4] found oromotor incoordination to be the most common cause of aspiration. It must be borne in mind that gastro-esophageal reflux disease (GERD) is a separate entity[4] from oromotor incoordination and can also contribute to recurrent pneumonia. However, in a patient of aspiration pneumonia secondary to GERD, no improvement of pulmonary symptoms is expected to occur on feeding through a naso-gastric tube. In GERD, the gastro-esophageal sphincter is expected to be incompetent, so reflux will continue to occur, until and unless the food is given through a site distal to the 'competent' pyloric sphincter (by a feeding jejunostomy). The very fact that naso-gastric tube feeding, with complete stoppage of feeding by mouth, improves the pulmonary symptoms, conclusively proves that GERD was not the etiology in our patient. Besides upper gastro-intestinal endoscopy did not show any mucosal ulceration and barium swallow was non-contributory.

Chiari malformation type 1 can be congenital or acquired, most commonly due to lack of development of posterior cranial fossa and consequent herniation of the cerebellar tonsils and medulla to upto 5 mm within the foramen magnum[5]. Although multiple symptoms can occur, some of which are mostly apparent at a further age, dysphagia may often be the sole manifestation of the disease[6]. Recurrent aspiration pneumonia due to dysphagia is also well known in this disorder[7]. Additionally, we also concluded that the protein energy malnutrition is occurring as a result of neurological dysphagia due to this palato-pharyngeal incoordination[8].

Hence, we ascertain that in a child with recurrent pneumonia with failure to thrive, neurological cause has to be considered even when no apparent neurological complaints are present.

Conclusion

It is very common for the paediatricians to encounter cases of both recurrent pneumonia and FTT. Although there are occasional case reports describing Chiari malformation type 1 to be associated with aspiration pneumonia, however it must be borne in mind that in most

cases this malformation is benign and asymptomatic. Thus pediatricians need to be well informed of this rare association so as to start appropriate management as early as possible and thereby avoid causing irreversible lung damage from long standing aspiration.

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