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CHRONIC LUNG DISEASE PRESENTING AS SHORT STATURE IN CHILDHOOD

Paediatrics

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ABSTRACT

Conditions leading to chronic pulmonary inefficiency can affect growth of children, by causing growth failure, delayed puberty and short stature. In addition to respiratory consequences, there is ample evidence that decreased growth is a result of decreased energy intake and increased energy expenditure (1). Enhancing the long term outcome for these conditions may therefore require consideration of both improved pulmonary management and aggressive nutritional management to limit growth failure and potentially enhance developmental outcome (1). Normal growth is considered as a barometer of good health and short stature (SS) might be the first sign of various pathological conditions. Here authors have reported an 11 year male presented with chronic respiratory complaints and short stature .Child was diagnosed with generalized fibrocystic bronchiectatic changes, due to cystic fibrosis.

KEYWORDS

INTRODUCTION

Growth is a continuum of biological processes, subject to genetic, environmental, nutritional and hormonal influences. Any deviation from normal growth potential may result from disturbance in any of these factors. Short stature, a common problem in child population of developing countries (3), is defined by height or length below 3rd centile or less than 2 standard deviation for that specific age and sex (4).

Etiology of short stature in children is very diverse, ranging from constitution causes, genetic causes and endocrinal causes to chronic diseases. Short stature is a common pediatric problem. Since normal growth is a barometer of health in childhood, any child who is growing normally virtually excludes any chronic physical or mental illness per se. Most common cause of short stature is normal variant short stature (36.1%) and familial short stature(15.9%), endocrine causes constitute 30% and chronic diseases constitute 7.8% of short stature.(5) Normal variant short stature was the most common cause of short stature followed by endocrine causes.(6)

Chronic childhood diseases, if sufficiently severe, can lead to growth failure and short stature, important examples include; renal, pulmonary and cardiac diseases, malignancy, cystic fibrosis and celiac disease (7).

Pulmonary biology and nutrition are closely interrelated. Conditions of malnutrition can cause significant effects on pulmonary function, and chronic pulmonary disease can lead to poor growth and delayed development (1).

CASE REPORT

An 11 year old male child presented to emergency department with chief complaints of persistent productive cough, with odorless, colorless sputum since 3 years of age, gradually progressive in nature. Along with complaint of ear discharge, decreased appetite, not gaining weight, dyspnoea on exertion, since last 7 years. Patient presented with respiratory distress in emergency department. No history of fever, chest pain, palpitations, haemoptysis ,haemetemesis, cyanosis, joint pain, oligouria, pedal edema, swelling, vomiting , loose stools, abdominal pain ,allergy, dermatitis etc present. There was negative history of tubercular contact. Antenatal history was insignificant. Birth weight was 3 kg, no significant postnatal history. Child is vaccinated for age.

Family history was insignificant, as there was no h/o sibling death, abortions or still born issues. No family history of chronic diseases, asthma, DM, hypertension. Pedigree analysis was normal .Developmental milestones were achieved at appropriate age. Child studies in 5th class and a good student, scores good in class.

Child has history of frequent hospitalizations started at 2 months of age for recurrent respiratory symptoms and ear discharge relieved partly by nebulisation and oral antibiotics. Patient had multiple renal stones (left kidney) and underwent open surgery at 2.5 yrs of age.

Examination revealed a nondysmorphic, proportionate child who appeared much younger than his chronological age. Height was 106cm (3rd centile height 128.2cm), weight was 16 kgs (3rd centile weight 22.6 kg) and Body mass index was less than 3rd centile. Upper: lower segment ratio, arm span, vital signs, optic fundi and visual fields were normal. There was no thyromegaly, scoliosis or any dysmorphism. Sexual maturity rating (TANNER STAGING) was 2. Patient has tachypnea, nasal flaring present with sub costal retractions. Patient was pale but no clubbing seen. Chest was asymmetrical and left sided rib cage was depressed. Air entry was decreased on right side of lung, bronchial breath sounds present on both sides with wheeze and prolonged phase of expiration.

Investigations revealed anemia with mild leucocytosis (hb-10gm%, TLC 14, 800, polymorphs -80 %) AEC- 200cells/cumm. Liver enzymes were slightly deranged, although renal function tests, electrolytes and blood glucose were normal. His bone age was equal to chronological age. Thyroid functions were normal. Pulmonary tuberculosis was ruled out as CBNAAT sputum came out negative. Chest X-Ray shows decreased right sided lung volume and patchy infiltrates .Pulmonary Function Test revealed early small airway obstruction and severe restriction,(FEF 25-75%, PEFR <70, FEV1/FVC >95% and FVC <44).

Contrast Enhanced CT scan of chest depicted fibrocystic bronchiectatic changes in left lower lobe, and inferior lingular lobe with diffuse fibro bronchiectatic changes in right middle lobe. Sweat chloride test later carried out resulted in 50 mmol/L, indicated towards cystic fibrosis. CFTR gene analysis could not be done due to financial reasons.

DISCUSSION

Although short stature is defined as an absolute height ≤3rd percentile for age and sex, a child's growth velocity is an even stronger indicator of their general health. Short stature can be due to physiological or pathological etiologies. Presenting manifestations include postnatal growth failure and delayed bone age (8).

The most common causes of short stature beyond the first year or two of life are familial (genetic) short stature and delayed (constitutional) growth, which are normal, non-pathologic variants of growth .

The goal of the evaluation of a child with short stature is to identify the subset of children with pathologic causes (such as Turner syndrome, inflammatory bowel disease or other underlying systemic disease, or growth hormone deficiency).

Bronchiectasis is defined as an abnormal dilation of the bronchi,

mostly irreversible, associated with airway obstruction, chronic productive cough and recurrent infections [10]. It begins with airway inflammation, almost always after an infectious process , linking the release of toxins and immune mediators with the subsequent permanent damage to the respiratory tract. Moreover, this lead to bacterial colonization and recurrent infections, enhancing tissue damage (11). A number of predisposing factors have been associated with the development of this condition, including infection, congenital defects, genetic predisposition and systemic inflammatory disorders (12). As proposed by Cole this inflammatory process is progressive and results in a cycle of worsening pulmonary damage (15). A common feature of bronchiectasis is malnutrition, which causes short height for age, secondary to slowing skeletal growth, as well as low weight in children (9). Henceforth, chronic pulmonary diseases such as Cystic fibrosis, causes Growth failure in children.

Bronchiectasis can be further classified into two major groups; cystic fibrosis (CF) that typically occurs in childhood and non-cystic fibrosis (non-CF) [13]. In resource-rich countries, cystic fibrosis (CF) is the most common cause of bronchiectasis in children. Bronchiectasis can be caused by a variety of disease processes, including combination of bronchial obstruction and infection (14).

Cystic fibrosis was thought to be extremely rare in India. However published reports, reviews and comments indicate that CF is probably far more common in people of Indian origin than previously thought but is under diagnosed or missed in the majority of cases (16). There are no community based studies to document precise incidence/ prevalence of CF in India. A study on 950 cord blood samples investigating carrier state of F508 mutation in india calculated incidence of CF as 1: 40000 newborns (17). The estimated prevalence of CF is 1 / 43,321 to 1 / 100,323 in Indian populations.

CF represents a good example of the bilateral relationship between pulmonary function and nutritional status. Whereas nutritional deficiency can lead to poor lung growth and increased infection, poor pulmonary function also causes increased energy utilization and growth failure.

Indian children are usually diagnosed late and in advanced stages and differ from their counterparts from developed world in being frequently malnourished, having clinical evidence of fat soluble vitamin deficiencies and more chances of being colonized with Pseudomonas (18). CF is not rare in India as it was thought to be and should be seriously considered in the differential diagnosis of children with recurrent/persistent respiratory infection and/or chronic malabsorption with failure to thrive. The diagnosis must confirmed by demonstrating elevated sweat chloride levels along with genetic confirmation with mutational studies. Treatment must be initiated via pancreatic enzyme replacement therapy along with good nutritional support, rigorous airway clearance and antibiotics during pulmonary exacerbations forms the mainstay of management.

Diagnostic facilities in India are scarce. Mutation profile is different with a lower prevalence of ΔF508. Management of CF in India is difficult due to inadequately trained manpower, lack of financial support, limited availability and high cost of pharmacologic agents (18).

The determinants of early death in Indian children with CF include: severe malnutrition, colonization with Pseudomonas at the time of diagnosis, more than four episodes of lower respiratory infection per year and age of onset of symptoms before 2 months of age. There is need to create awareness amongst pediatricians, develop diagnostic facilities and management protocols based on locally available resources.

CLINICAL IMAGES

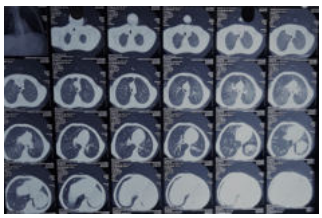

Image 1
CECT scan of chest shows fibro- bronchiectatic changes in right middle lobe and diffuse fibrocycstic bronchiectatic changes in left lower lobe.

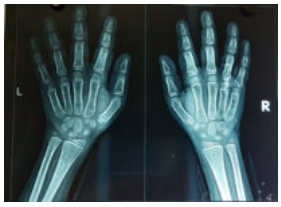

Image 2
X ray of Bilateral hands and wrist AP view shows bone age of 9-10 years

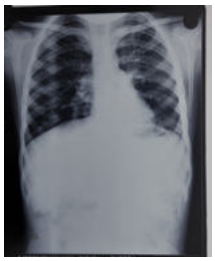

Image 3
Chest X ray PA view shows reduced lung mass in left lung, with fibro bronchiectatic changes in right middle lung field.

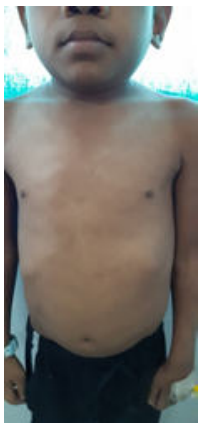

Image 4
Symmetrical Short stature patient with assymetrical chest wall and protrubent abdomen.

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