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General Medicine

YOUNGS SYNDROME - A RARE INHERITED SYNDROME IN YOUNG MALE ADULTS

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ABSTRACT Youngs syndrome also known as azoospermia sinopulmonary infections,sinusitis -infertility syndrome and Barry - Perkins -Young syndrome is a rare, inherited syndrome commonly seen in middle aged men with chronic reccurent rhinosinusitis, bronchiectasis, infertility due to azoospermia. Diagnosis of youngs syndrome is based on the occurrence of early onset progression in adult life with the presence of clubbing, sinusitis, and cystic bronchiectasis1.Azoospermia is seen due to hypomotility and decreased sperm count.

KEYWORDS : Azoospermia,cystic bronchiectasis,recurrent rhinosinusitis

Case Report-

Here we report a case of 25 year old married male patient who is a shopkeeper by occupation came to opd of GGH,Ananthapur with complaints of shortness of breath,cough with mucopurulent expectoration since 5 months with recurrent attacks of exacerbation since early childhood. There was no history of mumps,trauma or any history of drug intake ,no history of surgeries like adenoidectomy and tonsillectomy in early childhood.No consanguity or similar complaints in the family.

On General examination showed grade 4 clubbing of both fingers and toes.On chest examination AP to transverse diameter is increased.On auscultation ,bilateral inspiratory crepts present all over both lungs.chest xray shows right lobe and left lower lobe consolidation. Xray PNS suggestive of frontal and maxillary sinusitis. Sputum for CBNAAT is negative for 3 samples.HRCT chest suggestive of extensive cystic bronchiectasis involving right medial segment and left lower lobe of lungs.semen analysis report shows nil sperm count with normal viscosity,suggestive of azoospermia.patient treated with IV piperacillin and tazobactum ,along with bronchodilators and steroids for about 2 weeks.clinical response is seen and advised regular inhaled beta 2 agonists and steroids.

This image suggestive of cystic bronchectactic changes in right medial segment and left lowerlobe

DISCUSSION-

Young′s syndrome is a common cause of recurrent sinopulmonary infection and azoospermia.one of the most important differential diagnosis for youngs syndrome is cystic fibrosis,there fore we must differentiate from each other . Cystic fibrosis is an autosomal recessive disease with equal presentation in both sexes where as youngs syndrome occurs in males.

The bronchiectasis in cystic fibrosis is generally progressive and characteristically colonized initially with staphylococcal pyogenes and later with pseudomonas, as disease progresses to respiratory failure. The lower respiratory disease observed in Young′s syndrome is nonprogressive without any characteristic microbial culture. They have normal sweat chloride test and testicular dysfunction[2]. In contrast cystic fibrosis has decreased spermatogenesis and abnormal sperms.

Another hypothesis suggests that Young′s syndrome may be a variant of immotile cilia syndrome which is an autosomal recessive defect characterised by immobility or poor motility of cilia in airway and sperms. Kartagener′s syndrome is a subgroup of immotile cilia syndrome associated with situs invertus, chronic sinusitis and bronchiectasis. The structural deformity leading to impaired motility of cilia can usually be defined by electron microscopic appearance showing defect in dynein, or microtubule doublets[3]. Cilia from respiratory epithelia and sperm tails exhibit the same defect but the pulmonary manifestation may be minor. In primary ciliary dyskinesia sperms are hypomotile and ultrastructurally shows decreased mucus clearance, hence more prone for recurrent sinopulmonary infection[4].

The present case report shows pattern of chronic sinopulmonary infection with azoospermia. Clinical picture of chronic lung infection such as clubbing, mild polycythemia were present. X-ray showed hyperinflation with basal peribronchial thickening and or cystic bronchiectasis of both lungs. X-ray of paranasal sinuses shows maxillary and frontal sinusitis with area of central opacification. CT thorax shows alveolar destruction and cystic bronchiectasis. Pulmonary function test showed mild airflow obstruction mostly in smaller airways.

Management of Young′s syndrome in form of control of infection with antibiotics and bronchodilators. Hormonal supplementation for treatment of azoospermia.Microsurgical techniques to restore fertility is the modality of treatment for Young′s syndrome. Microsurgical epididydmovasostomies have been performed however with poor results. As a complication it causes further obstruction by thickened secretions even after a technically adequate anastomosis. Therefore epididydmovasostomy appears to be worse in patients with Young′s syndrome than in those with epididymal obstruction.patients with youngs syndrome may benefit from the use of ICSI (intracytoplasmic sperm injection) or intrauterine insemination(IUI) ,using their own sperms extracted from the epidydimis,if the surgical treatment has failed to restore fertility.

REFERENCES:

1. Seaton A, Seaton D. Bronchiectasis Crofton & Douglas. 20025th edn:7
2. Wellesy D. Schwartzs, Cystic fibrosis, Young′s syndrome & normal sweat Lancet. 1998;352:38
3. Banshi S, Guiris A, et al Immotile cilia syndrome with azoospermia Br. J Chest dis. 1998;82:194–196
4. Hanndelsman DJ, Conway AJ, Turtle, et al Obstructive azoospermia and sinopulmonary infection N Engl J Med. 1984;310:3–9