CASE REPORT

KARTAGENER'S SYNDROME PRESENTED AS PARANASAL POLYPOSIS WITH RECURRENT EPISTAXIS: A RARE CLINICAL CASE REPORT

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ABSTRACT

Background: Genetically determined syndromes of ciliary dyskinesia prevent normal transport of mucus from the bronchial tree to the mouth and result in serious impairment of lung defence system.male infertility was sometimes associated with Immotile spermatozoa. Approximately half of patients with Primary Ciliary Dyskinesia have full triad of kartgeners's syndrome, give history of recurrent sinusitis and lower respiratory tract infection from early life to adulthood [1].Kartagener's syndrome has been considered to be a sub group in a heterogenous collection of disorders to which Immotile Ciliary Syndrome or Dyskinitic cilia syndrome have been applied.[2] There may also be a link with retinitis pigmentosa and hearing loss.[3]
Aim: Kartagener's syndrome with paranasal polyposis is a uncommon presentation shown in our case.
We report an adult female of 23 of age having Recurrent sinusitis,Bronchiectasis and Dextrocardia with Situs inversus and with Paranasal polyposis showing recurrent epistaxis.
Conclusion: Kartagener's syndrome with paranasal sinusitis is common but paranasal polyposis with epistaxis is uncommon way of presentation.

Keywords: Kartagener's syndrome, Sinusitis, Bronchiectasis, Situs inversus with Dextrocardia, Paranasal polyposis

CASE REPORT

A 23 years old female patient attended our OPD,Dept., of Chest ,KIMS Hospital,Amalapuram with complaints of cough with expectoration,general weakness, digginess. Last one month she is having recurrent episodes of epistaxis.She is suffering from cough with expectoration and breathlessness since her childhood.Past history of surgery to the nose at the age of 16 years details not known.She was married having 2 children.Her general condition was thin built,illnourished.Her pulse92/minute,regular,BP 110/70mm of HG,RR 18/minute.Chest was symmetrical without any deformities.Her apical impulse was ½ inch medial to midcavicular ine on 5th intercostal space on right side .Per abdominal examination , liver dullness on left side and splenic dullness on right side.On percussion ,tympanic note on right hypochondrium.Lungs were clear on auscultation without any adventitious sounds.heart sounds were louder on the right side and S1 S2 heard.No murmurs were heard.

INVESTIGATIONS

1. CXR PA: Revealed base to apex axis pointing towards right .Stomach bubbe on right side.
2. ECG: 12 lead standard ECG showed P wave ,QRS and T waves are Down in L1. P wave,QRS and T waves are UP in AVR,P wave,QRS and T waves are Down in AVL.No left ventricular deflexion in V3V4V5V6. V1V2V3V4V5V6 showed progressive left ventricular deflexion with transition zone in V3.
3. Echo cardiogram: Demonstrated dextrocardia with ejection fraction of 65%.
4. Ultrasonogram abdomen: Reversal of abdominal viscera.
5. Nasal exam: Turbinates pale,No tenderness on paranasalsinus.Post nasal drip present.Ears normal.
6. HRCT Thorax: Suggesstsive of 1.Situs inversus2.Segmental Bronchiectasis 3.Paranasal polyposis indicating Kartagener's syndrome.

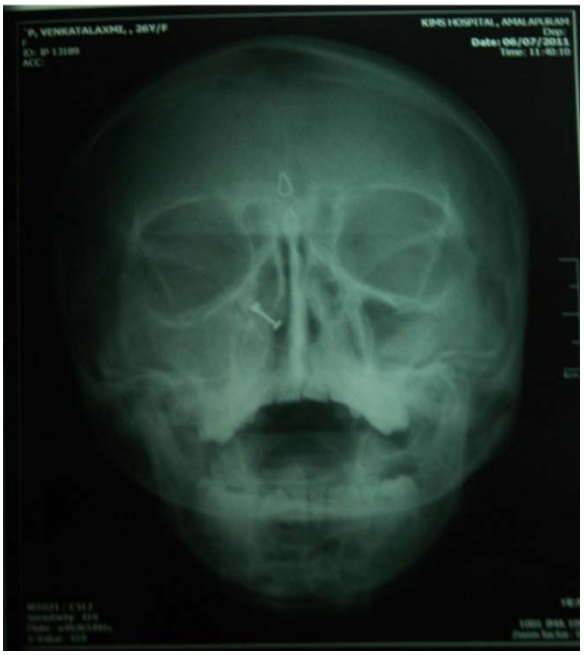

Fig 1: X ray PNS: Right Maxilary sinusitis present

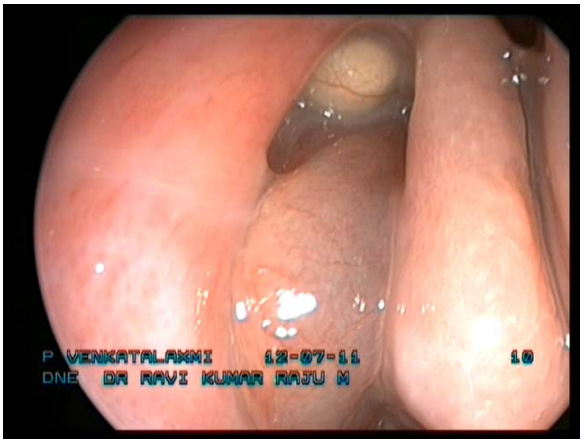

Figure 2A: Endoscopy shows whitish polyp mass

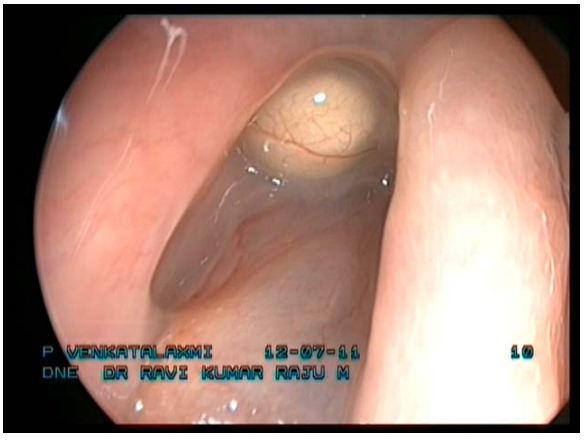

Figure 2B: Whitish polyp closure view

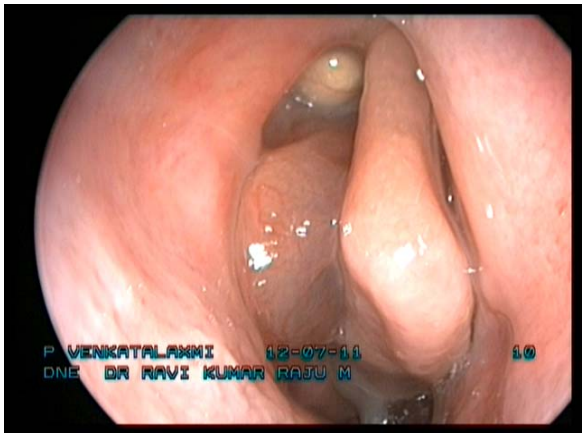

Figure 2C: Nasal polyp over view

Fig 2: Nasal Endoscopy: Bilateral extensive mucoid secretions present .Laft nasal cavity fairly normal.Right nasal cavity wide,relatively small turbinates.Whitish mass covered with membrane and blood vessels seen at roof lateral to middle turbinate.

DISCUSSION

It is suspected that visceral rotation in the embyo is dependent upon normal ciliary action,hence the association between primary ciliary dyskinesia and situs inversus abnormality. Genetically determined syndrome of ciliary dyskinesia prevent normal transport of mucus from bronchial tree to the mouth and result in serious impairment of lung defence system.The incidence of the genetic disorder is 1 in 32,000 births.Howeever ,higher incidences have been found in communities in which consanguineous marriages are common.[4] In our case she married blood relative, her own mother's brother which is very common in villages of Andhra Pradesh.Neonatal repiratory distress may occur in many cases but in our case there is no such history of childhood sufferings.[5] In our case this lady after attaining age of 23 years got reported having recurrent sinusitis ,bronchiectasis and dextrocardia with situs inversus and with paranasal polyposis.

She was presented with episodes of epistaxis and heamoptysis.Nasal examination done by ENT surgeon suggested of paranasal sinusitis and paranasalpolyposis. Chest X- ray suggestive of bronchiectasis and situs inversus.

Efforts to standardize the clinical criteria for the diagnosis of Kartageners'syndrome have centred on dextrocardia, a ciliary beast frequency of less than 10 Hz/s and a mean cross-section dynein arm count of less than 2.If the patient does not have dextrocardia,the identification of primary ciliary dyskinesia becomes the mainstay of diagnosis.Genetic testing ultimately may become the principal means of establishing this diagnosis.[6]

Obstructive lung disease/ bronchiectasis should be treated with inhaled bronchodilators,mucolytics and

chest physiotherapy.[7]Lobectomy is sometimes required for the associated bronchiectasis.[8] Lung transplantation and heart-lung transplantation have occasionally been tried in sever cases with some success. [9,10]

This episode of heamptysis and epistaxis make us to confuse but finally HRCT Thorax and CT Scan PNS suggestive of paranasal sinusitis,bronchiectasis and situs inversus and confirmed Kartagener syndrome.

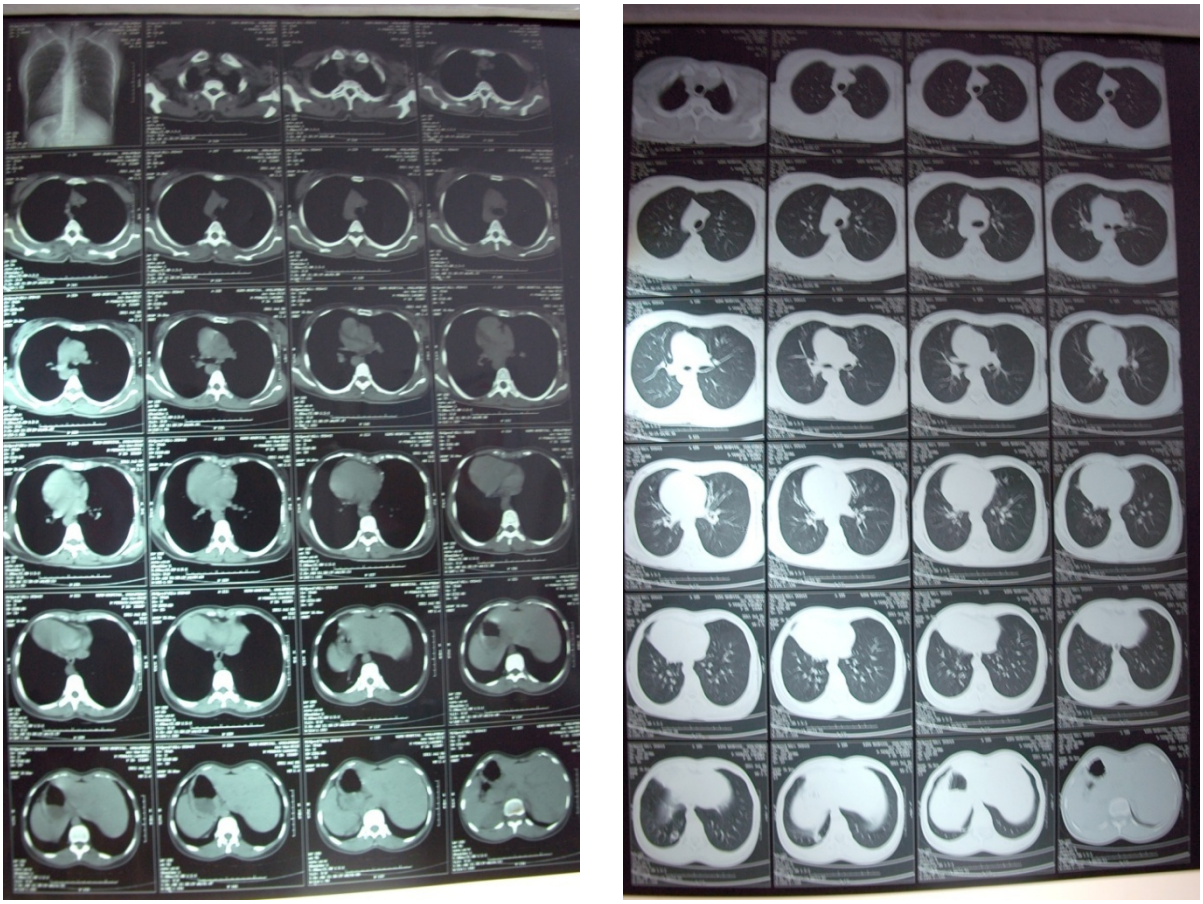

Figure 3: HRCT Thorax-Showing Dextro Cardia and Bronchiectatic changes in lungs

CONCLUSION

Kartagener's syndrome presenting with episode of epistaxis is rare .Nasal endoscopy examination revealed paranasal plyposis.But later we did Chest X ray which suggestive of Bronchiectasis with dextrocardia.HRCT Thorax reveaed the truth of Kartagener's syndrome with Situs inversus,Bronchiectasis and Paranasal sinusitis.With episodes of epistaxis and heamoptysis we think of heamatological abnormalties.But we confirmed it is a Kartagener's syndrome with paranasal polyposis which results in epistaxis.

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