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CASE REPORT

Kartagener's Syndrome in Children

by

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Abstract

Kartagener's syndrome is manifested by the association of sinusitis, situs inversus and bronchiectasis. The case described below occurred in a boy. Asplenia was also suspected with a strong family history of chronic respiratory infection. One of the four dead siblings and grandmother from mother's side has similar symptoms of respiratory infection and foul smelling breath. Symptoms of respiratory infection and running nose had started since infancy.

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Introduction

Although the association of situs inversus, bronchiectasis and sinusitis had been noted previously by Siewert (1940). Oeri (1909) and Guenther (1923) the first report of this combination as a syndrome were by Kartagener (1933) in four cases. Later he reported seven further cases in 1935, and since then the syndrome has been associated with his name (Holmes etal, 1968; Karani, 1952).

In 1962 Kartagener and Stuchi reviewed three hundred thirty four reported cases. Purnaman and Tandari (1974) reported the first case in a twenty years old male in this country. This paper reports a Kartagener's syndrome in a child with suspected asplenia.

Case report

A., an Indonesian boy, was admitted at the age of 4 years because of his bronchopneumonia. He is the ninth child of the family, one of the four dead siblings and his grandmother had chronic respiratory tract infection since he was seven months old. His cough had been much more troublesome and he brought up foul smelling sputum some times copiously.

Physical examination revealed a boy aged 4 years, body weight of 10 kilograms, body length of 85 centimeters, with cough and running nose and foul smelling breath, dyspnoic with flaring of alanasi and intercostal retraction; body temperature was 38° Celcius.

There were fine and coarse moist rales in inspiratory and expiratory through out the chest. Dextrocardia was present, and liver dullness was on the left side, spleen was not palpable. There is clubbing of the fingers. Tuberculin skin test was negative. The central nervous system was normal.

Erythrocyte count was 2.72 million per mm^3, hemoglobin content was 7,3 g% leucocyte 18800 per mm^3, hematocrit 23%, reticulocyte 7‰, differential count : eosinophil 3%, stab 53%, lymphocyte 40%, monocyte 4%.

Electrolyte were in normal limits. The immunoglobin content were normal with IgM 121 mg%, IgA 249,8 mg%, IgG 2961 mg%. Blood gas examination revealed pH 7,433, PCO2 41,9 mmHg, PO2 72,4 mmHg, HCO3 26,9 mEq/l.

PCO2 28 mEq/1, B.E. + 3,1, saturation 91,0%, SBc 27,1. Howell Yolly bodies was found in blood smear.

A plain X-ray film of the chest was suggestive of bronchiectasis and bronchopneumonia (Figure 1). Situs inversus totalis and Dextrocardia were confirmed by Barium meal (Figure 2). Rontgenogram of paranasalis showed Maxillary Sinusitis (Figure 3).

Electrocardiogram showed left ventricle hypertrophy and true mirror image ventricle heart position and sinus tachycardia.

He was given Ampicilllin, Chloramphenicol due to his bronchopneumonia.

His condition is getting worse with apneic attacks and transfered to intensive care unit, but passed on the tenth day hospitalization.

Discussion

These hereditary and familial tendencies of the syndrome were first noted by Guenther (1923) who reported the triad in two sisters among four siblings and later emphasied by Kartagener and Stucki (1962).

The genetic pattern is compatible with an autosomal recessive inheritance with some degree of pleotropic effect. It seems noteworthy that frequently more than one member of the family is affected (Kartagener and Stucki 1962; Bergstrom etal, 1950). In such families an accumulation of bronchiectasis, chronic bronchitis or recurrent sinusitis can be noted even with situs solitus or situs inversus without any respiratory disease. Someo of them having occurred in twins and siblings.

In this case one of the four dead siblings and his grandmother had chronic respiratory infection.

It has been estimated that the incidence of situs inversus is 1 : 8000 (Bergstrom et al, 1950; Holmes et al, 1968) and that approximately 12-23% of the individuals with situs inversus also have sinusitis and bronchiectasis. On this basis the incidence of Kartagener's syndrome would be 1 : 40.000.

In the Department of Child Health, Dr. Cipto Mangunkusumo General Hospital since 1974 - 1979 from 871 congenital heart disease patient, five had dextrocardia and situs inversus and this case is the first Kartagener's syndrome in a child ever been reported.

Purnaman and Tandari (1974) reported the first case in a twenty years old male in this country. The larger percentage of those in whom history was available, the symptoms dated back to infancy or early childhood, and ninety percent symptoms were present before the age of fourteen years with recurrent respiratory infection and mucopurulent nasal discharge.

In this case the boy gave history of recurrent upper respiratory infections since he was seven months old and his cough had been much more troublesome and he brought up foul smelling sputum sometimes copiously. The immunoglobulin content were normal with IgM 121 mg%, IgA 249,8 mg%, IgG 291 mg%. Holmes et al. (1968) reported 2 out of 13 affected patients the immunoglobulin were deficient, they noted transient immunoglobulin deficiency with a persistent low level of IgA.

Splenic agenesis or asplenia was suspected in this case because of the known association of this anomaly with serious abnormalities of the situs inversus totalis. The finding of Howell Yolly bodies on examination of the blood smear was sufficiently specific to permit the ante mortem diagnosis to be suspected even in the absence of severe associated anomalies (Bush and Lorin 1950; Polhemus and William 1952; and Samuel, 1956).

Martin (1826), observed absence of the spleen associated with situs inversus and congenital malformation of the heart in an infant.

Ventilatory obstruction at least one of the abnormality of pulmonary function. The abnormal findings ranged from mild to severe and the PCO2 measurement were normal in all patients except who had chronic pulmonary insufficiency. In this case blood gas analysis revealed PH. 7.433; PCO2 21,9; PO2 72,4 mm-Hg; saturation 91,9%.

Radiologic finding revealed : maxillary sinusitis, situs inversus totalis and dextrocardia confirmed by Barium meal examination. A plain X-ray film of the chest showed bronchiectasis and bronchopneumonia.

E.C.G. showed left ventricle hypertrophy and true mirror image. Concerning bronchiectasis with situs inversus, most authors agree that congenital or constitutional factors seems to be the main causative factors. The tendency of the triad to occur in one family also points to congenital or even hereditary or at least predisposing factor (Karani, 1952; Kartagener and Stucki 1962; Dickey, 1953).

According to Bard (1922) there was a basic faulty disposition of the supporting tissue (muscle elastic fibers, cartilage) of bronchial wall but according to Churchill (1949) there were an altered secretory activity of the mucous membranes of the respiratory system.

Afzelius (1976) has attempted to clarify the relationship between immotile cilia and Kartagener's syndrome. He suggested that the gene may be the one responsible for synthesis of the dynein protein which binds dynein to the microtubules. He proposed that cilia on embryonic epithelia have a fixed beat which is instrumental in determining visceral situs. Fisher et al. (1978) reported Kartagener's syndrome with middle dynein arms that lacked the typical "hook".

Previous studies in adult and older children have shown that microciliary clearance is lacking in patients with ciliary micro structural abnormalities. The abnormal ciliary function predisposes the individual to recurrent upper and lower respiratory problems, immotile spermatozoa.

Unfortunately we can not continue further examinations because the parents refused to perform autopsy.

REFERENCES

1. AFZELIUS, B.A. : A Human syndrome caused by immotile cilia. Science 193 : 317 (1976).
2. BARD, L. : Les dilations idiopathiques des Organes tubules ou cavitares J. Med. Lyon. 193 (1922) (Cited by Kartagener 1962).
3. BERGSTROM, W.H.; COOK, C.D.; SCANNELL, J. and BERENBERG, W. : Situs inversus bronchiectasis and sinusitis. Pediatrics 6 : 573 (1950).
4. BUSH, A.J. and LORIN, A. : Congenital absence of the spleen with congenital heart disease. Report of a case with

ante morfem diagnosis on the basis of hematologic morphology. Pediatrics. 8 : 93 (1950).

5. CHURCHILL, E.D. : The segmental and lobular physiology and pathology of the lung J. Thorac. Surg. 18 : 279 (1949) (Cited by Kartagener 1962).

6. DICKEY, L.B. : Kartagener's Syndrome in children. Dis. Chest 23 : 657 (1953).

7. FISCHER, J.T.; JAMES, A.M:; GREGORY, N.; ROBIN, C.; JOSEPH, E.G.; ROBERT, W.A. : Middle ear ciliary defect in Kartagener's Syndrome. Pediatrics 62 : 443 (1978).

8. GUENTER, H. : Biol. Zbl. 43 : 175 (1923) (Cited by Karani 1952).

9. HOLMES, L.B.; BLENNERHASSETT, J.B.; AUSTEN, K.F. : A reapprasil of Kartanener's Syndrome. Am. J. med Sci. 255 : 13 (1968).

10. KARANI, S. : Kartagener's Syndrome. Br. med. J. 12 : 74 (1952).

12. KARTAGENER, M. and STUCKI P.. : Bronchiectasis with situs inversus. Arch. Pediatr. 79 : 193 (1962).

13. MARTIN, G. : Observation d'une deviation organisue de lestomac, dune anomalie dans la situation, dans la configuration du cocur et des vaisseaux qui partent ou qu sy rendent. Bull. Soc. Anat de Paris. 1 : 40 - 51 (1926) (Cited by Polhemus 1952).

14. OERI, R. : Frankf. Z. Path. 3 : 393 (1909) (Cited by Karani 1952).

15. POLHEMUS, D.W.; WILLIAM, B.S. : Congenital absence of the spleen; syndrome with atrioventricularis and situs inversus. Pediatrics 9 : 696 (1952).

16. PURNAMAN, H.; TANDARI, R. : Sindroma Kartagener. Maj Kedok. Indones. 3 : 311 (1974).

17. SAMUEL, J.N. : Kartagener's Syndrome in a newborn infant. J. Am. med. Ass. 161 : 166 (1956).

18. SIEWERT, A.K. : Berl. klin. Wschr. 41 : 139 (1904) Cited by Karani 1952

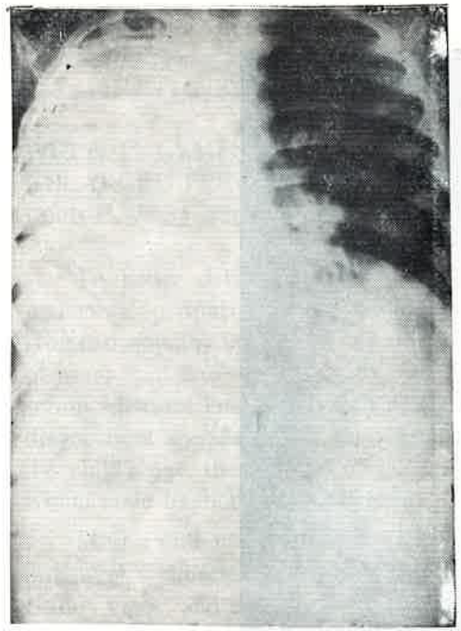

FIG. 1 : *Irregular density in the base of the left lung with honey comb pattern suggestive for bronchiectasis*

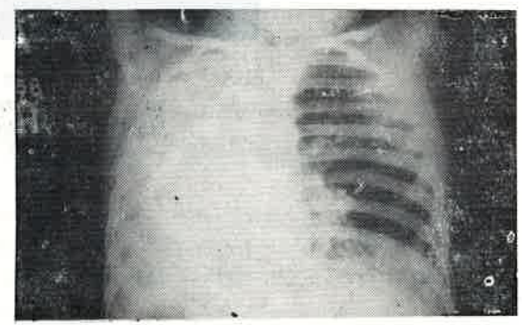

FIG. 2 : *Barium meal in frontal projection, showing Dextrocardia associated with transposition of all of the thoracic and abdominal organs (Complete Situs Inversus)*

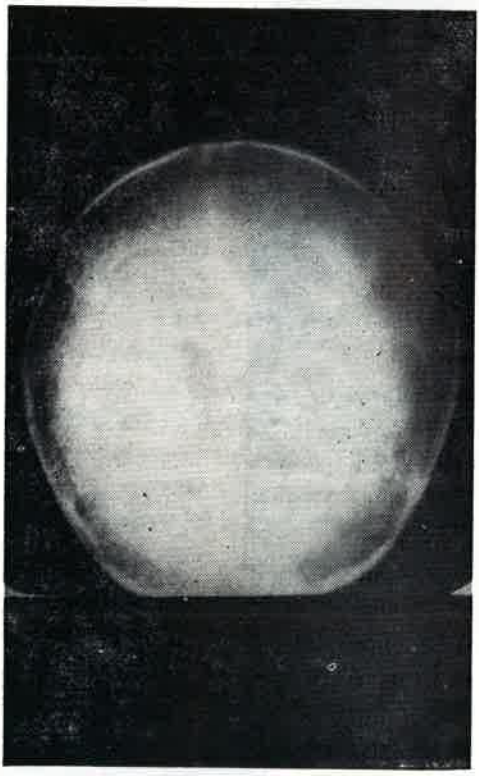

FIG. 3 : *Clouding of the Maxillary Sinuses in Waters projection showing Maxillary Sinusitis*