KARTAGENER SYNDROME IN CHILDREN

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 et al, 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, dyspnecic with flaring of alanas 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 mm³, hemoglobin content was 7,3 g%, leucocyt 18800 per mm³, hematocrit 23%, reticulocyte 7%, differential count: eosinophil 3%, stab 53%, lymphocyte 40%, monocyte 4%.

Electrolyte were in normal limits. The immunoglobulin 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 23 mEq/l, B.E. + 3,1, saturation 91,0%, SBe 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 parasanalitis showed Maxillary Sinusitis (Figure 3).

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

He was given Ampicillin, Chloramphenicol due to his bronchopneumonia.

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 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.

Received 1st September 1981.
His condition is getting worse with
apneic attacks and transferred 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
triple in two sisters among four siblings
and later emphasized 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 et al., 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. Some of them having occurred in twins and
siblings.

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

It has been estimated that the incidence of situs inversus is \( \frac{1}{8000} \) (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 \( \frac{1}{8000} \).

In the Department of Child Health, Dr. Cipto Mangunkusumo General Hospital since 1974 - 1979 from 871 congenital
heart disease patient, five had dextro-cardia 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.

Splenomegaly or splenosis was suspected in this case because of the known association of this anomaly with serious
abnormalities of the situs inversus tota-
lis. 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 : maxilla-
ry sinusitis, situs inversus totalis and
dextrocardia confirmed by Barium meal examination. A plain X-ray film of the
chest showed bronchiectasis and bron-
chopneumonia.

E.C.G. showed left ventricle hypertrophy and true mirror image. Concern-
ing 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; Dic-
key, 1953).

According to Bard (1922) there was a basic faulty disposition of the suppor-
ting tissue (muscle elastic fibers, cartil-
ge) of bronchial wall but according to
Churchill (1949) there were an altered
secretory activity of the mucous mem-
branes of the respiratory system.

Afzelius (1976) has attempted to clari-
fy 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 mi-
rotubules. He proposed that cilia on
embryonic epithelia have a fixed beat which is instrumental in determining
visceral situs, Fisher et al. (1978) repor-
ted 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 ci-
liary micro structural abnormalities. The
abnormal ciliary function predisposes
the individual to recurrent upper and
lower respiratory problems, immotile spermatoza.

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

REFERENCES


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

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


To better define the indications for diagnostic biopsy, 239 children who underwent peripheral lymph node biopsy were reviewed. The duration of the lymphadenopathy by history, the consistency of the lymph nodes, and the presence of more than one site of palpable adenopathy were not specific in differentiating serious diseases involving lymph nodes from reactive hyperplasia. The differential diagnosis of specific causes for lymph node enlargement is approached based on the child's age, the location of the adenopathy, and the presence or absence of lymph node fixation and tenderness.

Most children with supraclevicular adenopathy, children sick with fever of one week's duration or with weight loss for which a specific diagnosis is not readily made, and some children with fixation of the lymph node to the overlying skin should undergo early biopsy. Excluding the above findings, when a specific diagnosis is not apparent, serial measurements with a ruler over several weeks appears to be the most reasonable method, at the present time, of discriminating hyperplastic lymph nodes from nodes that are involved by a progressive disease process.


The clinical and laboratory features of urinary tract infections in 100 infants aged 5 days to 8 months are presented. Of the patients in the first three months of life 75% were boys, and of infants aged 3 to 8 months only 11% were boys; 95% of the infants were uncircumcised.

Sepsis was documented in 31% of neonates, 21% of infants aged 1 to 2 months, 14% of those aged 2 to 3 months, and 5.5% of infants > 3 months of age.

Roentgenographic abnormalities of the urinary system were found in 45% of female and 7% of male infants. All infants responded promptly to antimicrobial therapy. The possible factors related to the predominance of male infants with urinary tract infections in the first three months of life are discussed.