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Department of Radiology and Imaging Diagnostics of the District Hospital No. 2 in Rzeszów
2nd Department of Radiology, Medical University of Lublin

RADOSŁAW RAMOTOWSKI, WIESŁAW GUZ, EWA ZIĘBA,
GRAŻYNA ZŁOMANIEC

Clinical and radiological aspects of Kartagener's syndrome

Inverse position of the viscera associated with bronchial distensions and inflammatory changes in the mucosa of paranasal sinuses was termed Kartagener's syndrome – KS (3). Its underlying factor is a recessive genetic defect inherited in the autosomal way, responsible for the syndrome of primary ciliary dyskinesia. It coexists in 50% cases with inversed viscera (2).

Functional impairment of the ciliary apparatus involves the respiratory system, where it causes bronchial distensions, lateral sinusitis, sinuso-bronchial syndrome, middle lobe syndrome (4, 11). This results from abnormalities of bronchial tree structure (anomalies of the cartilaginous ring, fibrosis of bronchial wall elements) (7, 9). Anomalies of the alimentary tract (pancreocystosis, meconium impatency) and of the reproductive system (abnormal migration of the ovum to the endometrium, defective functioning of sperm cells). Disturbances of ciliary function also underlies anomalous rotation of viscera in the fetal life and immune disturbances (impairment of migration and chemotaxia of neutrophils, decreased immunity of delayed type, considerable increase in IgG level) (1).

Other disturbances coexisting with KS include: 1) disorders or bone-articular system (spondyloschisis, costal anomalies, disorders of dentition), 2) defects of the auditory organ (hypoplasia of the external ear, disturbances of pneumatisation of the mastoid process, deafness), 3) heart defects and defects of big vessels, 4) abnormal structure of the neural tube with consequences in the form of oligophrenia, epilepsy, 5) changes in the organ of vision (congenital cataract, congenital pigmental degeneration of the retina with short-sightedness, astigmatism) (12, 13).

Scientific literature presents KS cases mainly in adults (6). In children this syndrome is diagnosed and treated late. This is connected with more evident intensification of changes involving pathology of the bronchial tree in older children. In 90% of cases they get revealed about the age of 15 years and sometimes even much later.

CASE REPORT

A boy P. J. aged 14 years reported at a Paediatric Outpatient Unit because of persistent cough with throwing up sputum, sore throat, headaches, conjunctivitis and fever (37.8°C). His history showed frequent recurrent inflammations of nasal mucosa with serum-pus secretion. Physical examination revealed: tenderness to pressure of frontal sinuses, conjunctivitis, inflammation of oral mucosa, palate

tonsillitis with a lot of muco-pus secretion on the posterior pharyngeal wall. On auscultation scattered wheezes, dry rales and coarse rales were heard over pulmonary fields. Chest X-rays showed dextral heart position and streak condensation under the lower hilar pole on the left (Fig. 1). X-ray picture of nasal lateral sinuses showed opacities of maxillary sinuses. Clinical ailments considerably subsided owing to treatment.

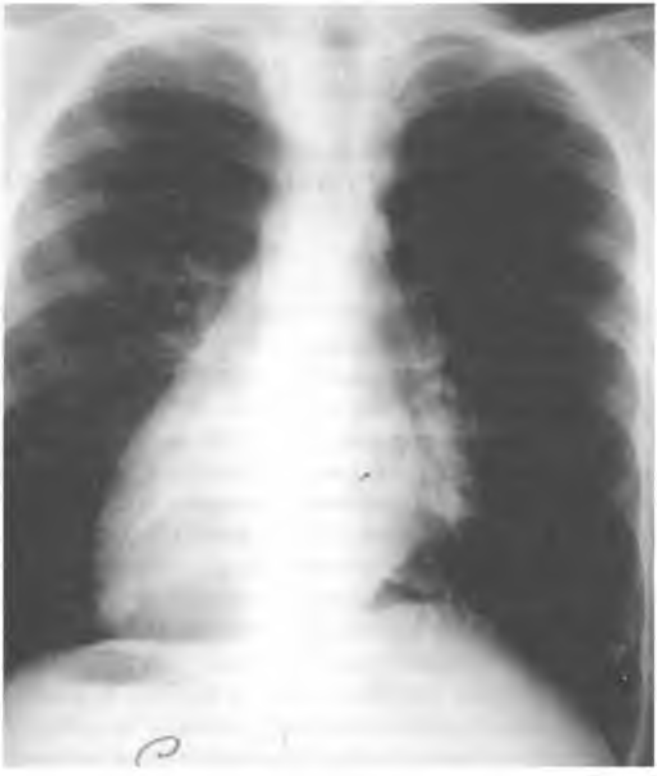

Fig. 1. Chest X-ray. Dextral heart position, streak condensations under the lower hilar pole on the left

After 3 months the patient was again admitted to the Unit with symptoms of respiratory insufficiency of obturative type, dyspnoea at rest increasing on even slight exertion. On auscultation diffuse dry rales and wheezes prevailed. USG of the abdomen showed situs viscerum inversus. ECG RZN showed dextrocardia. Laryngologic examination revealed rhinitis allergica chronica. Clinical improvement was achieved but despite treatment the child returned to the Unit again after next two weeks with symptoms of pneumonia. Recurrent episodes of bronchopneumonia, persistent cough, inflammatory changes within the middle lobe made doctors order chest CT. It showed inverse position of abdominal organs (Fig. 2) inflammatory-atelectatic changes of the middle lobe (Fig. 3), inflammatory condensations of pulmonary parenchyma in lower lung fields of bronchial origin.

The administered treatment gave subsidence of clinical ailments.

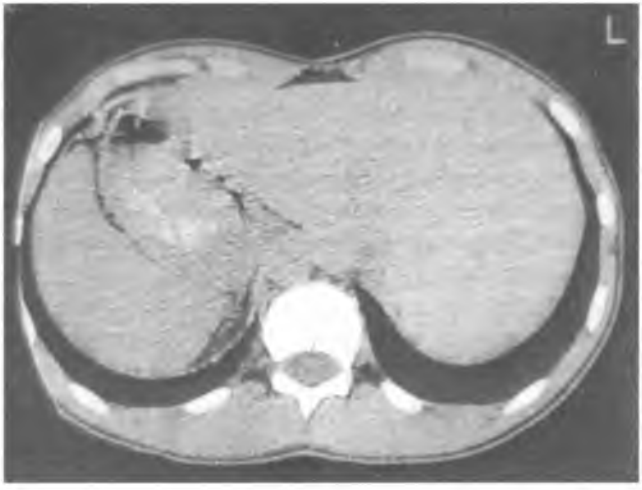

Fig. 2. CT inversed position of abdominal organs

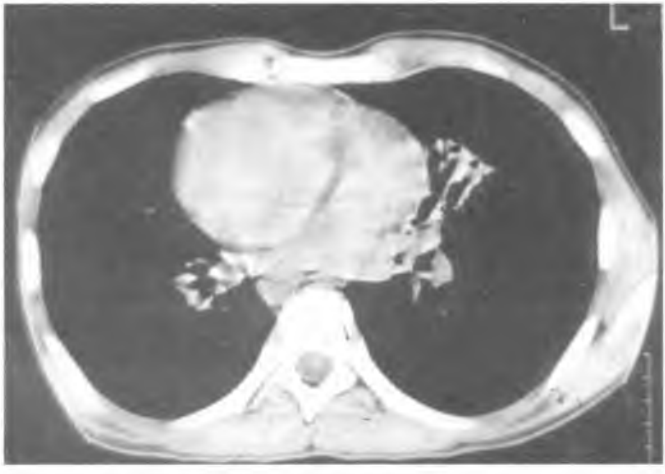

Fig. 3. CT inflammatory-ectatic-cirrhotic changes within the middle lobe

DISCUSSION

The presented case of KS initially regarded as an incidence of pneumonia made doctors, as time elapsed and no clinical improvement was observed, further look for causes. Performed examinations (X-ray, USG) led to the suspicion of Kartagener's syndrome. Final confirmation of bronchial distensions with CT imaging made diagnosis possible (10). Difficulties in treating recurrent respiratory inflammations resulted from a definite background of the illness which was ciliary diskinesia of

respiratory epithelium and secondary changes in the form of bronchial ectasiae and middle lobe syndrome. Literature emphasises that recurrent respiratory inflammations in these cases are caused by opportunistic germs (5, 8). Their treatment is primarily based on prevention, a big role being played by immune-modulating preparations administered orally as well as antibacterial and antiviral vaccinations. They can be used in both treatment and prevention of infections. In the case of bacterial superinfection and associated complications targeted (topical) therapy with antibiotics is absolutely indicated. Because of bronchial distension and excessively expressed mucosal function a big role in complex treatment is played by the removal of secretion residue (mucolitic drugs given orally or by inhalation, positional drainage, respiratory gymnastics).

Recognition of middle lobe syndrome and subsequent irreversible cirrhotic changes by means of chest X-ray and CT, on account of ineffective conservative treatment, imposes the decision to perform surgical treatment, resection of the cirrhotic lobe.

CONCLUSIONS

In case of frequent, recurrent respiratory infections a possibility of KS must be considered. In the diagnosis of KS radiological and imaging examinations are crucial such as chest X-ray, X-ray of lateral sinuses, chest CT and USG of the abdominal cavity.

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SUMMARY

The paper presents the picture of Kartagener's syndrome in a 14-year-old in whom inversed position od the vicera ectases of the bronchi and inflammatory changes in maxillary sinuses were found.

Recurrent infections of the respiratory system should suggest changes of this type of etiology requiring USG examinations of internal organs and especially CT of the lungs.

Kliniczne i radiologiczne aspekty zespołu Kartagenera

Przedstawiono obraz zespołu Kartagenera u 14-letniego chłopca, u którego stwierdzono odwrócone położenie trzew, rozstrzenie oskrzeli i zmiany zapalne zatok szczękowych. Nawracające infekcje układu oddechowego powinny nasuwać podejrzenie zmian tego typu etiologii, skłaniające do badań USG narządów wewnętrznych, a zwłaszcza TK płuc.