CASE REPORT

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Kartagener's syndrome: A case report

Kartagenerov sindrom

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Abstract

Introduction. Kartagener's syndrome is a recessive autosomal disease which is mainly seen to affect ciliary movement. The symptoms of the syndrome are the consequence of the defective motility of the cilia found in the respiratory tract and that results with recurrent lung infections caused by mucus stasis in the bronchi. **Case report.** A 37-year-old married male, father of one child, presented with the history of productive cough, wheezing, dispnea, headache, temporary fever. In his 9th year of age, in 1986, *situs inversus*, sinusitis and *pectus excavatum* were diagnosed. In 1994 he was operated for correction of *pectus* excavatum. Bronchial asthma was diagnosed in 2008 when he was 31. In the last 2 years he had episodes of breathlessness, wheezing, cough, expectoration, headache, fever and fast declining lung function. The patient was treated with combination of inhaled bronchodilatators (inhaled corticosteroids + long-acting β-2 agonist), and occasional administration of antibiotics, oral prednisolone, mucolytics in episodes of exacerbations of disease over a period of 7−14 days. **Conclusion.** Treatment for patients with this syndrome has not been established yet, but it is important to control chronic lung infections and prevent declining of lung function.

Key words:
kartagener syndrome; respiratory tract infection; bronhiectasis; therapeutics; fertility.

Apstrakt

Uvod. Kartagenerov sindrom je recesivno autozomno oboljenje koje se uglavnom ispoljava kao zahvatanje cilijarnog kompleksa. Simptomi se ispoljavaju kao posledica poremećaja u pokretljivosti cilija u respiratornom traktu, što rezultira ponavljanim infekcijama uzrokovanim zastojem sekreta u bronhijama. **Prikaz bolesnika.** Muškarac, star 37 godina, oženjen, otac jednog deteta, javio se na pregled sa simptomima produktivnog kašlja, zviždanja u grudima, gušenja, glavobolje i povremeno povišene telesne temperature. U devetoj godini života, 1986. godine, dokazan mu je *situs inversum*, sinuzitis i deformitet grudnog koša u vidu kokošijih grudi; 2008 godine dokazana mu je bronhijalna astma. U poslednje dve godine bolesnik je imao epizode otežanog disanja (sa „kratkim dahom"), zviždanjem u grudima, kašljem i iskašljavanjem, glavoboljom, groznicom i brzim pogoršanjem plućne funkcije. Lečen je kombinacijom inhalacionih bronhodilatatora (inhalacioni kortikosteroidi + dugodelujući β-2 agonisti) i povremenom primenom antibiotika, oralno prednizolona i mukolitika, tokom 7–14 dana, u epizodama pogoršanja bolesti. **Zaključak.** Lečenje bolesnika sa Kartagenerovim sindromom još uvek nije usaglašeno, ali treba da bude usmereno na kontrolu hroničnih plućnih infekcija i sprečavanje opadanja plućne funkcije.

Ključne reči:
kartagenerov sindrom; respiratorni trakt, infekcije; bronhiektazije; lečenje; plodnost.

Introduction

Kartagener's syndrome is a recessive autosomal disease which is mainly seen to affect ciliary movement [1]. The incidence of Kartagener's syndrome is 1–2/30,000 births. Siewert first described the combination of *situs inversus*, chronic sinusitis and bronchiectasis in 1904 [2]. Manes Kartagener, a pulmonologist in Zurich, first recognized this clinical triad as a distinct congenital syndrome in 1933. Kartagener described this syndrome in detail, so it bears his name [2]. The symptoms of the syndrome are the consequence of the defective motility of the cilia found in the respiratory tract and that results with recurrent lung infections caused by mucus stasis in the bronchi [1–3]. In older children and adults with primary ciliary dyskinesia, 3 diseases of the lower

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respiratory tract have been described: pneumonia, bronchiectasis and asthma [4]. Patients with Kartagener syndrome may have immotile spermatozona as well [5, 6]. Treatment for patients with this syndrome has not be established, but it is important to control chronic lung infections and prevent declining of lung function [7, 8].

Case report

A 37-year-old male, married, father of one child, presented with the history of productive cough, wheezing, dispnea, headache, occasional fever. In 1986 *situs inversus*, sinusitis and *pectus excavatum* were diagnosed. In 1994 the patient was operated for correction of *pectus excavatum*. In 2008 bronchial asthma was diagnosed. In the last 2 years he had episodes of breathlessness, wheezing, cough, expectoration, headache, fever and fast declining lung function.

At clinical bilateral examination, predominantly on the left side, crepitations and rhonchi were evidenced.

A chest X-ray showed dextrocardia, signs of pneumonia in lower pulmonary field on the left side (Figure 1).

Chest and abdominal CT revealed dextrocardia, bronchiectasis in the lower lobes, dominantly on the left side, left liver, gastric bubble and spleen on the right (Figures 2−5).

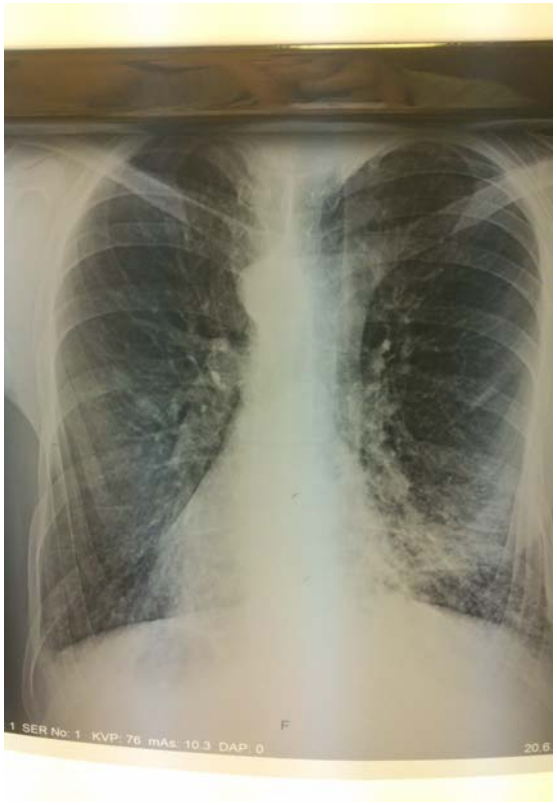

Fig. 1 – Chest radiograph shows dextrocardia and features of pneumonia in the left lower lung field.

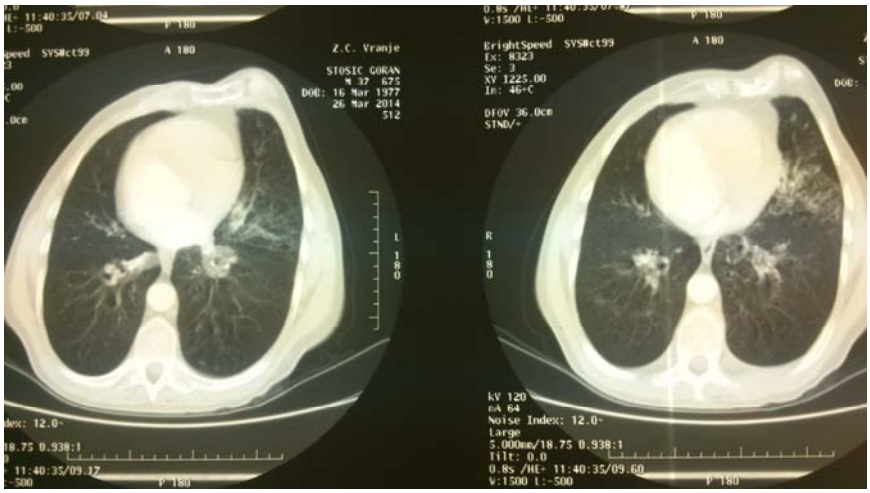

Fig. 2 – Contrast-enhanced computed tomography images show dextrocardia and bronchiestasis in the lower lobes, dominantly on the left side.

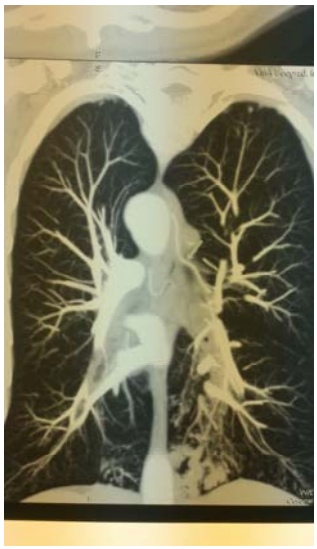

Fig. 3 – Chest contrast-enhanced computed tomography scan showing bronchiestasis in both lower lung lobes, dominantly in the left lower lobe.

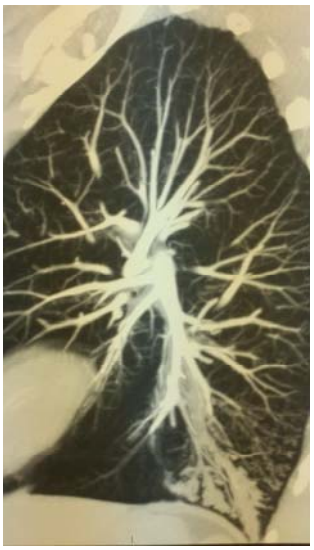

Fig. 4 – Chest contrast-enhanced computed tomography scan showing bronschiectasis, as previously described.

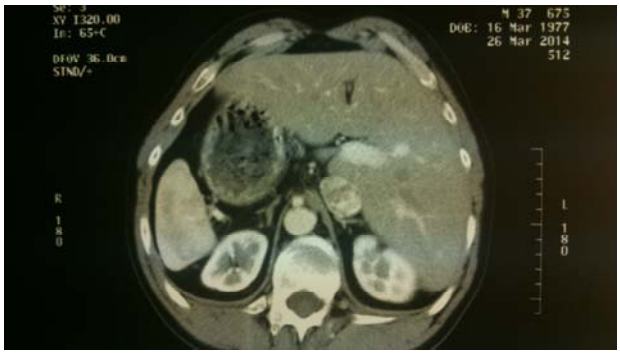

Fig. 5 – Abdominal computed tomography scan shows complete inversion of the internal organs: liver lying on the left side of the abdomen, gastric air bubble and spleen on the right side.

It was difficult to obtain correct images during echocardiography because of the chest abnormality (after the operation of *pectus excavatum*), but transthoracic echocardiogram and transesophageal echocardiography confirmed dextrocardia, too.

Spirometry demonstrated bronchial obstruction forced expiratory volume in 1s (FEV1) 1.6 L, 38% of predicted value (4.26 L), forced vital capacity (FVC) 58% (3.00 L) FEV1/FVC 64%.

As the patient was diagnosed with congenital Kartagener's syndrome 28 years ago, in the last two years he was treated with a combination of inhaled bronchodilatators (inhaled corticosteroids+long-acting beta-2 agonists), and occasional administration of antibiotics, oral prednisolone, mucolytics in episodes of exacerbations of disease over a period of 7–14 days. In the last year the patient was admitted to hospital 4 times because of exacerbations of bronchial asthma and inflmmation bronchiectasis.

Discussion

Kartagener's syndrome is seen in 50% of patients with primary ciliary dyskinesia. Kartagener's syndrome is characterized by *situs inversus*, bronchiectasis, sinusitis and otitis media. In some cases it is inherited with bronchial asthma. Our case is unusual for 2 reasons. Firstly, the presented patient was fertile and had a child. He refused to give a spermogram. Munro et al. [9] their show that 30% of their patients with primary ciliary dyskinesia have normal spermatozoa and that 2 patients are fertile. Secondly, the presented had *pectus excavatum*. In a study by Kennedy et al. [10] it was found that 10% patients with Kartagener's syndrome had *pectus excavatum*.

The presented clinical case demonstrated a progressive course of bronchiectasis, declining lung function, because of recurrent infections which were treated inappropriately in recents period.

Conclusion

We presented this case because the Kartagener's syndrome is a very rare condition.

The prognosis is generally considered favorable, and life expectancy is usually normal. An important part of the clinical visits at regular intervals should be monitoring the progression of the lung disease.

R E F E R E N C E S

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