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Case Report

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A Case of Kartagener Syndrome

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The Kartagener syndrome is an autosomal recessive disorder characterized by bronchiectasis, sinusitis and situs inversus. This research reports a 28 years old woman with dextrocardia ask for genetic counseling to have a healthy child. She suffers from recurrent chronic sinusitis and bronchitis with situs inversus including right-sided heart and pancreas and left-sided liver. Kartagener syndrome was diagnosed according to her signs, symptoms, radiological and ultrasonographic evaluations.

Key words: Dextrocardia, sinusitis, bronchitis, Kartagener syndrome

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INTRODUCTION

Kartagener Syndrome (KS) is an autosomal recessive disorder with incomplete penetrance (Gorham and Merselis, 1959), which characterized by the triad of situs inversus, bronchiectasis and sinusitis (Casanova *et al*., 2006). The primary defect is abnormal function of cilia, which causes immotile cilia syndrome. The immotile cilia cause recurrent sinopulmonary infection (Chodhari *et al*., 2004). KS was seen in consanguinity especially first cousin (Moreno *et al*., 1965). It is likely that the Kartagener syndrome is a mendelian subgroup of situs inversus viscerum (Torgersen, 1950). There is no association with left-handedness. Dextrocardia may be absent and the patient may suffer from bronchiectasis, sinusitis and infertility (Guerrant *et al*., 1978). Patients with the Kartagener syndrome may have anosmia (Goldstein, 1979). The sperm of males with Kartagener syndrome are immotile, which is the same as every cilliary cell in the body (Afzelius *et al*., 1976). The spermatozoa have fertility capacity and with using assisted reproductive technique, they will fertilize the egg. Women with Kartagener syndrome usually are fertile, although eggs are moved down in fallopian tube by cilia. So using in vitro fertilization in the management of such patients is a possibility (Aitken *et al*., 1983).

CASE REPORT

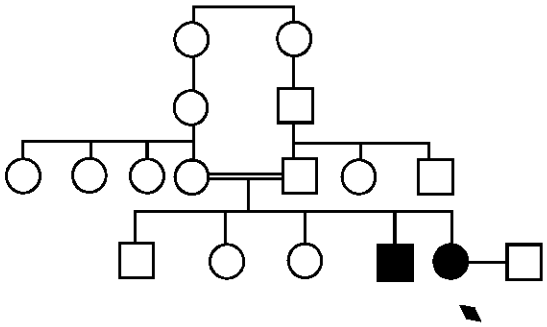
This case report described a 28 years old woman sought genetic counseling to have a healthy child. She suffered from recurrent chronic sinusitis and bronchitis and with using antibiotic; the patient got rid of that signs and symptoms for a few weeks. She has situs inversus including right-sided heart and pancreas and left-sided liver in physical examination. For complete evaluation, ultrasonography of internal organs, chest X-ray and echocardiography were asked. Right-sided pancreas and left-sided liver were visualized in sonography. Dextrocardia with bronchectasis were seen in chest X-ray. The apex of heart was right in echocardiography, but the valves and walls were normal. The sign of chronic sinousitis was seen in the radiography of sinus. Laboratory tests include fasting blood sugar, triglyceride, cholesterol, SGOT, SGPT, creatinine, sodium and potassium were normal (Table 1).

Table 1: The results of the laboratory test for this patient

Lab test	FBS	TG	CHOL	AST	ALT	CREA	SOD	P O T
Results	94*	176*	189*	34**	32**	0.9*	135*	4.5*

*mg dL^{-1}; **U L^{-1}

She had an 18 years old brother with the same problem, dextrochardia and situs inversus, but he did not accept to have more evaluation. In addition parents of them were second cousin. The pedigree of patient was drawn as follow:



DISCUSSION

Kartagener syndrome is originally described as the combination of situs inversus, bronchiectasis and sinusitis (Casanova *et al*., 2006). This syndrome is a mendelian disorder, which inherited as autosomal recessive. Narayan *et al*. (1994) suggested either X-linked or autosomal dominant inheritance for this syndrome.

Evaluation of this syndrome by genetic test is difficult because of genetic heterogeneity. In various linkage studies, this syndrome has been known as a disorder with a great genetic heterogeneity. It has not been determined any gene in the pathogenesis of this syndrome until now but some studies reported the association of Kartagener syndrome with linkage to chromosome 7 (Witt *et al*., 1999), heterozygous mutations in the DNAI1 (located on 9p21-p13) and DNAH5 genes (resides on 5p15-p14) (Guichard *et al*., 2001; Olbrich *et al*., 2002). In present study, genetic evaluation was not done, because of indefinite affected genes.

There are new reports of this syndrome which confirm diagnosis according to clinical and paraclinical findings (Ishiga *et al*., 2005; Naves *et al*., 2005).

In present study, a 28-years-old woman was described with recurrent chronic sinusitis and bronchitis. Right-sided pancreas and left-sided liver were visualized in sonography. Chest X-Ray confirmed dextrocardia with bronchectasis. Also, the signs of chronic sinusitis were seen in the sinuses graphy. Therefore, Kartagener syndrome was diagnosed for this patient.

CONCLUSIONS

According to heterogeneity of this disease, genetic evaluation of it is very expensive and usually useless. Genetic counselor recommended this couples to have a

baby because the pattern was suggested according to their pedigree is autosomal recessive. The sonography could show the position of internal organ and situs inversus in fetus at 3 months of age, which could help to diagnose Kartagener syndrome in offspring. However, rarely immotile cilia is their symptom alone.

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