



Relationship Between Kartagener's Syndrome and Internal Diseases: Coincidence or Coexistence?

Kartagener Sendromu ile dahili hastalıkların ilişkisi: Tesadüf mü? Birliktelik mi?

Muhammet Limon, Nurdan Yıldırım

Mersin University, Faculty of Medicine, Department of Rheumatology, Mersin, Türkiye

Dear Editor,

Kartagener's syndrome (KS) is a rare, autosomal recessive genetic mucociliary disorder that consists of a triad of situs inversus, chronic sinusitis and bronchiectasis.^[1,2] Patients with primary ciliary dyskinesia (PCD), including KS, may present with various manifestations, such as infertility, olfactory disorders, ophthalmopathy and hydrocephalus resulting from ciliary dysfunction. Ciliopathy has become recognized as a multisystem disorder, of which PCD is an important subgroup. In our review of the literature, the coexistence of rheumatic disease, renal disease, malignancy and KS have been reported. We write this letter to draw attention to this issue.

The coexistence of rheumatic diseases and KS relationships have been rarely reported. Only 10 cases reported the association of KS and rheumatoid arthritis (RA).^[3,4] One case described the association of systemic lupus erythematosus and dextrocardia.^[5] One case described the association of undifferentiated connective tissue disease and dextrocardia.^[6] An association between RA and bronchiectasis has been reported, although the underlying pathogenic mechanism remains unclear. There is no evidence that KS is associated with specific HLA antigens, indicating that the occurrence of RA may be coincidental.^[7] Beutler et al. proposed that continuous exposure to infections resulting from mucociliary dysfunction in the airways may play an important role in pathogenesis of RA.^[8]

Ali Momeni et al. reported KS with focal segmental glomerulosclerosis.^[9] El Houssni S et al. reported renal amyloidosis revealing a KS.^[10] Also cyanotic heart disease

may be relationship with focal segmental glomerulosclerosis as reported by Flanagan, the probable cause of which could be hyperfiltration of the glomerulus.^[11] Chronic hypoxia may cause many medical problems. Chronic hypoxia is mentioned as a cause of secondary focal segmental glomerulosclerosis. Hida and colleagues reported a case of congenital heart disease with focal segmental glomerulosclerosis.^[12] A case of KS and polycystic kidney disease was reported by Sayarlioglu and colleagues.^[13]

The coincidence of malignancy and KS have been reported. These reported cancers to date are renal cell carcinoma, testicular germ cell tumor, lung cancer, carcinoma of the ethmoid labyrinth, colon carcinoma, angioimmunoblastic T cell lymphoma, adenocarcinoma of the paranasal sinuses, adenocarcinoma of the cervix uteri and endometrium. It is seen that most of these tumors are tissue or organ cancers with ciliated epithelium. The majority of these tumors can be explained by impaired mucociliary clearance and exposure to chronic irritations.^[14]

In conclusion, we think, as suggested by other authors^[8,9,14] that chronic infectious processes, impaired mucociliary clearance and chronic hypoxia may play an important role in the development of KS coincidence with internal diseases. As the number of cases demonstrating the coexistence of KS and internal diseases increase, it will be possible to obtain more detailed information about this issue.

Keywords: Kartagener's syndrome, relationship, internal disease



ETHICAL DECLARATIONS

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

REFERENCES

1. Sleight MA. Primary ciliary dyskinesia. *Lancet*. 1981;2(8244):476.
2. Afzelius BA. A human syndrome caused by immotile cilia. *Science*. 1976;193(4250):317-9.
3. Mastaglia GL, Goatcher P. Rare association of rheumatoid arthritis and Kartagener's syndrome: a common denominator?. *Med J Aust*. 1980;2(7):400.
4. Rébora ME, Cuneo JA, Marcos J, Marcos JC. Kartagener syndrome and rheumatoid arthritis. *J Clin Rheumatol*. 2006;12(1):26-9.
5. Martínez-Cordero E, Aracena-Taborga G, López-Zepeda J. Systemic lupus erythematosus and dextrocardia. *Clin Exp Rheumatol*. 1990;8(3):327-8.
6. Limon M. Kartagener's syndrome and undifferentiated connective tissue disease. *Int J Rheum Dis*. 2024;27(5):e15179.
7. Neffen H, Oehling A, Crisci CD. Kartagener's syndrome: a report of six cases with special reference to humoral and cellular immunity. *Respiration*. 1980;40(3):161-7.
8. Beutler A, Mackiewicz SH. Association of rheumatoid arthritis with Kartagener's syndrome in a patient with HLA-DR1-DR4-B27 haplotype. *Z Rheumatol*. 1992;51(5):253-5.
9. Momeni A, Doroushi B, Taheri N. Kartagener syndrome with focal segmental glomerulosclerosis. *Iran J Kidney Dis*. 2013;7(6):499-501.
10. El Houssni S, Laine M, Bziz A, Alhamany Z, Eddine Bourkadi J, Bayahia R. Amylose rénale révélant un syndrome de Kartagener [Renal amyloidosis revealing a Kartagener's syndrome]. *Nephrol Ther*. 2015;11(1):50-52.
11. Flanagan MF, Hourihan M, Keane JF. Incidence of renal dysfunction in adults with cyanotic congenital heart disease. *Am J Cardiol*. 1991;68(4):403-6.
12. Hida K, Wada J, Yamasaki H, et al. Cyanotic congenital heart disease associated with glomerulomegaly and focal segmental glomerulosclerosis: remission of nephrotic syndrome with angiotensin converting enzyme inhibitor. *Nephrol Dial Transplant*. 2002;17(1):144-7.
13. Sayarlioglu H, Dagli CE, Dogan E, Sayarlioglu M, Koksak N. Kartagener's syndrome and polycystic kidney disease. *NDT Plus*. 2009;2(2):189-90.
14. Kara B, Seher N, Eren-Karanis M, et al. Nasopharyngeal carcinoma in a child with Kartagener's syndrome. *Turk J Pediatr*. 2021;63(1):155-60.