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Letter to the Editor/ Editöre Mektup

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

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

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Dear Editor,

Kartagener's syndrome (KS) is is a rare, autosomal recessive genetic mucociliary disorder that consist of a triad of situs inversus, chronic sinusitis and bronchiectasis.[1,2] Patients with primary ciliar 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, maligancy 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, adenosquamous carcinoma 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

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