Opinion

Ectodermal Dysplasia patients susceptible to COVID-19

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ABSTRACT

Ectodermal dysplasia (ED) is a rare heterogeneous disorder. Defects in the development of the ectoderm cause symptoms in tissues derived from the ectoderm layer such as skin, nails and hair. One of the genes involved in ED is Nuclear Factor Kappa-B (NF-κB) that mutation in this gene causes immunodeficiency. There are also some ED subgroups such as X-Link Hypohidrotic Ectodermal Dysplasia (XLHED) at risk of severe pneumonia and respiratory infections. The patients with ED due to the susceptibility of the immune system defect as well as the respiratory system, we hypothesized that these patients are sensitive to COVID -19. So the main organ involved in the respiratory system and the main cause of mortality in patients with COVID-19 is the respiratory system involvement. Therefore, they are at higher risk of developing symptomatic COVID-19 that requires further clinical care.

***Keywords*:** Ectodermal Dysplasia; COVID-19; infection

INTRODUCTION

Ectodermal dysplasia (ED) is a congenital disorder characterized by disturbances in the tissues originating from ectoderm. Therefore, sparse hair, peg-shaped teeth, and lack of sweat glands are typical of this congenital disorder. There are approximately 200 subtypes of ED have been identified. Hypohidrotic Ectodermal

Dysplasia (HED) is the common form of ED [1]. HED shows a decrease in mucus production in the respiratory system; therefore, patients are susceptible to severe infections. HED can be inherited as X-linked recessive, autosomal dominant, and autosomal recessive. X-Link Hypohidrotic Ectodermal Dysplasia (XLHED) is caused by a mutation in the EDA gene. EDAR and EDARADD genes cause autosomal recessive and autosomal dominant types of ED. Many respiratory infections have been reported in patients with an autosomal recessive mutation in the EDARADD gene. One of the ED types is associated with immunodeficiency, a kind of immunological disorder caused by mutations in the coding genes of Nuclear Factor Kappa-B (NF-κB). NF-κB is an essential transcription regulator factor of ectodermal growth and adaptive immune function. Activation of NF-κB is a critical signaling event in immune responses downstream of different immune receptors such as Toll-like, interleukin-1, T cell, and B cell receptors [2,3]. In addition, in almost all patients, signs of infection in the form of combined immunodeficiency indicate EDA. People suffering from XLHED are at risk of severe pneumonia and respiratory infections at young ages. Respiratory infections, wheezing, and frequent sinus infections can be continued in adults. Immunodeficiency caused sensitivity to progenies bacteria, such as *Streptococcus* pneumonia, Haemophilus influenza, Mycobacteria and Herpes virus [4]. Deletion of four exons in the EDARADD gene shows frequent respiratory infections in childhood. Sometimes these chronic respiratory infections are seen in patients with X-Linked recessive inheritance pattern and who have p.Trp135-Val136del EDARADD gene mutations. Other genes include PKP1 and TP63, also cause infections in ED [5]. SARS-CoV-2 affects different parts of the body, but respiratory involvement is the main cause of death. One of the most common infections is pneumonia, which in COVID-19 leads to respiratory tract infections. Mutations in genes NF-κB, EDA, EDARADD, PKP1, and TP63, are the main cause of bacterial, fungal and viral infections in ED. Therefore, ED patients with mutations in these genes are at high risk for respiratory tract infections [6]. Also, patients who have a mutation in the NF-κB gene and have COVID-19 cause over-activation of this gene, causing cytokine storm syndrome, one of the most important causes of death in patients with COVID-19 [7]. A study also looked at 14 patients with HED who tested positive for COVID-19 and found

that they had a higher risk of long-term COVID-19 symptoms such as hair loss and chronic fatigue. So, this group of patients should be vaccinated against COVID-19 [8]. Therefore, COVID-19 is very dangerous for patients with ED, and families with ED patients should pay more attention to health protocols.

CONCLUSION

The COVID-19 affects different body systems. Therefore, all healthy and patients are prone to COVID-19, especially patients with a history of other diseases. ED is one of the diseases that is susceptible to severe COVID-19, these patients have immunodeficiency and respiratory defects, so these patients need special care not to get COVID-19.

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