Clinical Significance of Heat Shock Protein 70 in Autoimmune Inner Ear Disease Patients

Thesis
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الأهمية الإكلينيكية لبروتين الصدمة الحرارية 70 في مرض الأذن الداخلية ذاتي المناعة

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Eman Soliman

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LIST OF ABBREVIATIONS

ABR	Auditory brain stem response
AECAs	Anti-endothelial cell antibodies
AIED	Autoimmune inner ear disease
ANA	Anti-nuclear antibodies
ANCA	Antineutrophil cytoplasmic antibody
anti-DNA	Anti- deoxyribonucleic acid
APHA	Antiphospholipid antibodies
APS	Antiphospholipid syndrome
ASNHL	Autoimmune sensorineural hearing loss
C	Complement
CD	Cluster of differentiation
CIC	Circulating immune complex
CIEAg	Crude inner ear antigen
COCH5B2	Coagulation factor C homolog, cochlin
CRP	C-reactive protein
CSF	Cerebrospinal fluid

CT	Computed Topography
dB	Decibels
ECOG	Electrocochleography
ELISA	Enzyme-linked immunosorbent assay
ELS	The endolymphatic sac
ENA	Extractable nuclear antigen
ENT	Ear, Nose, and throat
ER	Endoplasmic reticulum
ESR	Erythrocyte sedimentation rate
FTA	Fluorescent Treponemal Antibody Absorption
GGA	Geranylgeranylacetone
HL	Hearing loss
HLA	Human leukocyte antigen
HSF	Heat shock factor
HSP	Heat shock protein
ICAM-1	Intercellular adhesion molecule 1

IDT	Intradermal dilutional testing
IF	Immunofluoresence test
IFN-γ	Interferon-gamma
Igs	Immunoglobulins
IK	Interstitial keratitis
IL	Interleukin
IPBSNHL	Idiopathic progressive bilateral SNHL
IPSNHL	Idiopathic progressive SNHL
ISSNHL	Idiopathic sudden SNHL
kDa	kilo Daltons
LTT	Lymphocyte transformation test
MD	Meniere's disease
MHC	Major histocompatibility complex
MRI	Magnetic resonance imaging
mRNA	Messenger ribonucleic acid
M.W	Molecular wieght
NK	Natural killer

NSAIDs	Nonsteroidal anti-inflammatory drugs
PAGE	Polyacrylamide gel electrophoresis
PBS	Phosphate buffer saline
PHA	Phospholipids antibodies
PHL	Progressive hearing loss
PMNCs	The polymorphonuclear cells
PVDF	Polyvinylidene fluoride membrane
rbHSP70	Recombinant bovine HSP70
RF	Rheumatoid factor
RAST	Radioallergosorbent testing
rhHSP70	Recombinant human HSP70
RPSNHL	Rapidly Progressive SNHL
SAD	Systemic autoimmune disease
SHL	Sudden hearing loss
sHSPs	Small heat-shock proteins
SLE	Systemic lupus erythematosus
SMV	Spiral modiolar vein

SNHL	Sensorineural hearing loss
SSNHL	Sudden sensorineural hearing loss
T cells	T-lymphocytes cells
TNF-α	Tumor necrosis factor-α
TSH	Thyroid stimulating hormone
WB	Western-blot
WG	Wegener’s granulomatosis

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INTRODUCTION

The immune system is designed to ward off infection. However, there are times when the immune system malfunctions and instead of fighting infection it turns on our own body parts and attacks them. Over the years, there have been sporadic reports of sudden or rapidly progressive hearing losses that have been found to be immune-mediated or in the setting of other systemic autoimmune disease ***(Hain, 2003).***

Some cases of idiopathic sensorineural hearing loss (SNHL) of adult onset are attributable to immune recognition of inner ear proteins as foreign or non-self, a phenomenon referred to as autoimmune inner ear disease (AIED) **(*Tebo et al., 2006).***

Lehnhardt (1958) first introduced the possibility that some of these cases of rapidly progressing SNHL were caused by an immune process in the cochlea. ***Schiff and Brown (1974)*** further hypothesized that, the process was most likely of an autoimmune etiology, as the rapidly progressing SNHL could be reversed or stabilized by immunosuppressant drugs.

Although the mechanism(s) in the pathogenesis of AIED is not known, autoimmunity may be induced either within the inner ear, in a primary end organ response, or outside the inner ear and gain access to the inner ear as a secondary response **(*Tebo et al., 2006).***

AIED is a relatively newly described disease process of which little is known of the etiology, diagnosis and treatment. Many studies in the past 20 years have done much to further understanding of the immune function of the ear and its involvement in systemic disease ***(Bonajuri et al., 2007)***

The importance of diagnosing AIED is highlighted in the context of its being one of few forms of treatable inner-ear disorders with a good response to immunosuppressive therapy. Early diagnosis of AIED with prompt treatment may prevent irreversible damage to inner-ear structures ***(Agrup and Luxon, 2006).***

The diagnosis of AIED is ascertained by the history, clinical findings, response to immunosuppressive medication and an immunologic evaluation of the patient's serum ***(Bovo et al., 2006).***