

SERUM IgE LEVELS IN PATIENTS WITH ATAXIA TELANGIECTASIA*

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The role of IgE in the mediation of immediate hypersensitivity reactions and its association with allergic diseases has been well established^{1,2}. It is also known that IgE has a major role in the mechanism of elimination of parasites³. The function of IgE in host resistance to infection is, however, less clear. Serum IgE levels were investigated in many immunodeficiency diseases and found to be high in Wiskott Aldrich, Nezelof's and hyper IgE syndromes and selective IgA deficiency. IgE deficiency has been reported in patients with severe combined immunodeficiency, X-linked agammaglobulinemia, common variable immunodeficiency, X-linked immunodeficiency with hyper IgM and ataxia telangiectasia⁴⁻⁶.

Ataxia telangiectasia (AT) is an autosomal recessive disease characterized by progressive cerebellar ataxia, oculocutaneous telangiectasia and recurrent sinopulmonary infections. Although varying degrees of abnormalities in both cellular and humoral immunity have been described, there are only a few studies concerning the serum IgE level in AT patients^{4,7-11}.

In the present study, serum IgE levels were determined in 15 AT patients and compared with the normal IgE levels¹²⁻¹⁵. Also, the correlation between IgE levels and other immunological parameters was investigated.

Material and Methods

Sera were obtained from 15 AT patients and stored frozen at -20 °C until used in the study.

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TABLE I
Some Clinical and Immunological Findings of AT Patients

Patients	Age (years), sex		IgE IU/ml	Sinusitis/ Pneumonitis*	Serum immunoglobulins (mg/dl)			
					IgA	IgM	IgG	E rosette (%)
1 (HA)	14	M	21.8	+ / -	90	165	755	65
2 (IB)	11	F	25.0	+ / -	80	127	795	63
3 (MB)	17	M	0.0	+ / -	96	100	1230	43
4 (AÖ)	10	M	0.0	+ / +	43	170	590	36
5 (SA)	11	F	19.9	+ / +	14	253	1400	43
6 (EK)	9	F	73.9	+ / +	140	153	880	63
7 (MK)	8	M	0.0	+ / +	80	100	7750	39
8 (TE)	7	M	0.0	+ / -	0	137	830	61
9 (ÖK)	15	M	67.4	+ / -	131	145	925	58
10 (FÖ)	6	F	7.4	+ / +	0	99	1010	49
11 (AE)	16	M	0.0	+ / +	0	107	1150	52
12 (CA)	7	F	0.0	+ / -	80	113	1090	N.T.**
13 (MY)	14	M	20.8	- / +	0	261	730	36
14 (BB)	8	F	51.1	+ / -	110	113	1170	N.T.
15 (BB)	2.5	F	52.4	+ / -	94	167	1100	N.T.
Mean $\pm$ SD		22.65 $\pm$ 26.3						
Controls (n = 13)								
Mean $\pm$ SD		58.6 $\pm$ 50.8						69 $\pm$ 8
Range		(5-210)						(49-86)
		p < 0.05						

* The + and - indicate the presence or absence of sinopulmonary infection.

** Not tested.

IgE was measured by the commercially available radioimmunosorbent test (Phadebas IgE Test, Pharmacia Diagnostic Laboratories, Uppsala, Sweden) as prescribed by the manufacturer¹². Each serum sample was tested in duplicate, and mean values were given in international units (IU). Serum immunoglobulin levels were determined by radial immunodiffusion (Behringwerke) and the E-rosette test was performed using a previously described method¹⁶.

The mean IgE concentrations in controls of the same age were taken from studies carried out by the Allergy Unit of our Institute using the same radioimmunoassay method^{17,18}.

Results

Table I shows the IgE values of the 15 AT patients along with the age, sex, sinopulmonary infection status, serum IgA, IgM and IgG levels and the percentage of E-rosette forming cells. The mean serum IgE levels of the AT patients was found to be 22.65 IU/ml (SD: ± 26.3) and this value was significantly lower ($p < 0.05$) than in the 13 age-matched controls (58.6 ± 50.8). In 6 of the 15 AT patients serum IgE could not be detected.

No correlation was found between either the serum IgE and IgA levels, or the serum IgE levels and the percentage of E-rosette forming cells ($r: 0.31$, $p > 0.05$ and $r: 0.49$, $p > 0.05$ respectively).

Further, there was no connection between the serum IgE or IgA levels and the presence of chronic sinusitis and/or pneumonitis.

Discussion

Recurrent sinopulmonary infections are one of the major components of AT and have a prognostic significance. Although varying degrees of abnormalities have been described in both the cellular and the humoral immunity of these patients, no correlation could be found between these abnormalities and susceptibility to infection. IgE deficiency in AT patients had been previously reported^{4,11}. Our results are in agreement with these previous reports but the basic mechanism of the IgE deficiency observed in our study and other reports remains to be elucidated. Since IgE deficiency is commonly observed either in combination with other immunological defects or in normal individuals, the immunological role of IgE in host resistance to infection has not been clearly defined. The low IgE levels in addition to the other immunological abnormalities in AT patients may indicate that IgE also performs some useful function in protection against sinopulmonary infections, but no association could be found between IgE deficiency and the occurrence of recurrent sinopulmonary infections. Further, the serum IgE levels did not show any correlation with other immunological abnormalities found in these patients as for instance with the IgA levels, or with the percentage of E-rosette forming cells.

Summary

The serum IgE levels in 15 At patients were investigated and compared with those of normal individuals. The correlation of IgE with various immunological parameters is discussed.

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