

CHRISTIAN MEDICAL COLLEGE
VELLORE - 4
DEPARTMENT OF HAEMATOLOGY
MEDICAL REPORT

Name : ANWIN ANTO

Hospital Number : 122477D

Age : 2 **Sex :** Male

Visit Date : 20-Oct-2007

Address : THALOVE
THALORE
THRISSUR
THIRSSUR

KERALA

Pincode : 680306

DIAGNOSIS :

WISKOTT ALDRICH SYNDROME

HISTORY

Baby Anwin Anto, first born child to non-consanguineous parents presented with recurrent skin lesions from 6 months of age, recurrent epistaxis of 3 months duration and an episode of bloody diarrhoea 3 months back. There was no history of bleeding disorders in the family.

For skin lesions he was on homeopathic medications for 2 months, switched over to ayurvedic medications and continued the same for next 6 months. As there was no appreciable response to either of these medications, received treatment with prednisolone for 6 months following which the lesions got controlled.

For the bleeding episode he was evaluated from royal hospital, Muscat and was diagnosed to have Immune thrombocytopenic purpura based on clinical and bone marrow study. He was started on prednisolone 25mg OD which was tapered over months to the present dose of 5mg on alternate days.

ON EXAMINATION

Afebrile. HR: 100/mt. AF: closed

No pallor/icterus/edema/bleeds/significant lymphadenopathy.

Bilateral frontal prominence+.

Popular scaly skin eruptions all over the body more over the face & scalp. systems:

Gastro-intestinal system: no palpable spleen /liver /nodes /kidneys.

other systems: unremarkable.

INVESTIGATIONS

17-Oct-2007	RETICULOCYTE COUNT	2.19	%
17-Oct-2007	PT WITH INR		
	PATIENT	10.5	SECS
	NORMAL RANGE	9.7-13.3	SECS
	INR	0.977	
17-Oct-2007	CBC PROFILE		
	DC: NE:60 EO:20 Ba:0 MO:8 LY:12 B.Pic	PLT ON SMEAR 15,000/CUMM	
OCCASIONAL	SCHISTOCYTES PRT; OCC IRREGULAR CONTRACTED CELLS PRT;	DR.S.JAIN/DR.AP.	
	MCV	75.6	f1
	MCH	25.9	PG
	MCHC	34.2	G/DL
	RDW	18.8	%
	TOTAL WBC	15300	/CUMM
	HB	12.5	GM/DL
	PLATELET COUNT	24000	/CUMM
	RBC	4.82	
	HCT (PCV)	36.4	%
	RETICS	2.19	%
17-Oct-2007	APTT		
	PATIENT	24.0	SECS
	NORMAL RANGE	23.8-37.4	
17-Oct-2007	COOMBS TEST -DIRECT	NEGATIVE	
18-Oct-2007	BLOOD GROUPING & RH TYPING	B POS	
17-Oct-2007	LFT		
	BILIRUBIN TOTAL	0.6	mg%
	DIRECT	0.2	mg%

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	PROTEIN TOTAL	7.6	g%
	ALBUMIN	4.4	g%
	SGOT	32	U/L
	SGPT	15	U/L
	ALKALINE PHOSPHATASE	112	U/L
17-Oct-2007	CREATININE	0.4 R	mg%
17-Oct-2007	GLUCOSE RAN PL	93	mg%
18-Oct-2007	IMMUNOGLOBULINS. (IgA, IgG & IgM)		
	IGG	1060	mg%
	IGA	224	mg%
17-Oct-2007	BLOOD BORNE VIRUS SCREEN		
	HIV	NEGATIVE	
	HbsAg	NEGATIVE	
	HCV	NEGATIVE	

SPECIAL INVESTIGATIONS

IMMUNOPHENOTYPIC ANALYSIS FOR WISKOTT ALDRICH SYNDROME:
GATING ON MONONUCLEATED CELLS:

	IgG2a	%WSAP-B9
Patient (Anwin)	10.6	10.5
Father	9.6	95.7
Mother	8.2	95.9

Impression: Consistent with Wiskott Aldrich syndrome

DISCUSSION

Baby Anwin Anto was evaluated and was found to have eczema, microthrombocytopenia (MPV-8.1fl) and absent WASP protein on immunophenotyping. Sample for DNA analysis of the patient and the parents has been sent and the report is awaited. Based on the above features he was diagnosed to have Wiskott Aldrich syndrome.

[ESID diagnostic criteria for WAS: (Definitive-presence of # with at least one of *)
#Male patient with congenital thrombocytopenia- YES

*Absent WASP protein in lymphocytes- YES

*Mutation in WASP- awaited

*Absent WASP mRNA on northern blot analysis of lymphocytes-not done

*Maternal cousins, uncles or nephews with small platelets & thrombocytopenia - UNKNOWN (to be evaluated)]

The nature of the diagnosis and the curative treatment option ie; bone marrow transplant in the event of HLA matched related donor and option for splenectomy if bleeding due to thrombocytopenia becomes severe and persistent, was discussed with the parents. As he is the only child HLA typing of parents was done and the father is a 4/6 match only so transplant carries a significant risk. However he can be considered for a Haplotransplant. Dermatology consult was done for advice on skin care. As the patient was on alternate day prednisolone it was decided to continue the same and stop in next 3 months.

RECOMMENDATIONS

1. Emoderm cream twice daily
2. Desonide cream twice daily
3. Elocon cream daily once
4. Syp. Atarax 5mg PRN
5. Lacticare HC for local application
(Use the above medications as advised by the dermatologist)
6. Avoid live virus vaccines
7. Treat infections promptly with appropriate antibiotics
8. Avoid frequent bath; use bath oils during the bath
9. Use skin moisturisers after bath and frequently thereafter to avoid dry skin
5. Platelet transfusion in the event of bleeding
6. Consider immunoglobulin therapy if frequent infections occur.
7. Review in 6 months.

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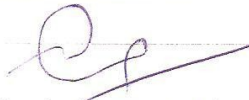
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