



COMPILED BY :

DR ATHOTA KAVITHA

ACAD MODERATOR

JOINT SECRETARY, IADV L2022-23

**PIGMENTARY DISORDERS**  
**SIG PIGMENTARY DISORDERS (IADV L ACADEMY)**  
**(ACAD DISCUSSION 2022)**

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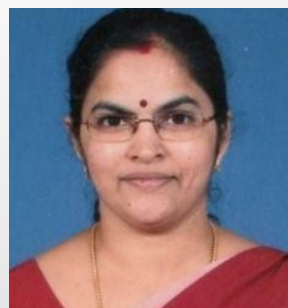
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# IADVL ACADEMY 2022



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The background of the slide is a light gray gradient, decorated with numerous realistic water droplets of various sizes. The droplets are rendered with soft shadows and highlights, giving them a three-dimensional appearance. They are scattered across the slide, with a higher concentration in the top-left and bottom-right corners.

# ACAD CASE 1 –SIG PIGMENTARY DISORDERS.

DR RASHMI SRIRAM

ASSOCIATE PROFESSOR.

DEPT OF DERMATOLOGY.

BGS GIMS, BANGALORE.

# HISTORY

- A 4 YEAR OLD FEMALE CHILD BORN AT FULL TERM BY NORMAL DELIVERY TO CONSANGUINEOUS PARENTS.
- H/O OF PIGMENTARY DISTURBANCES IN SKIN
- FROM THE AGE OF 2 YEARS.
- H/O OF RECURRENT SKIN INFECTIONS, SWELLING, AND PAIN IN THE RIGHT EAR FOR 15 DAYS.
- NO HISTORY OF RESPIRATORY INFECTION, BLEEDING, AND SIMILAR COMPLAINTS IN THE FAMILY.

# EXAMINATION

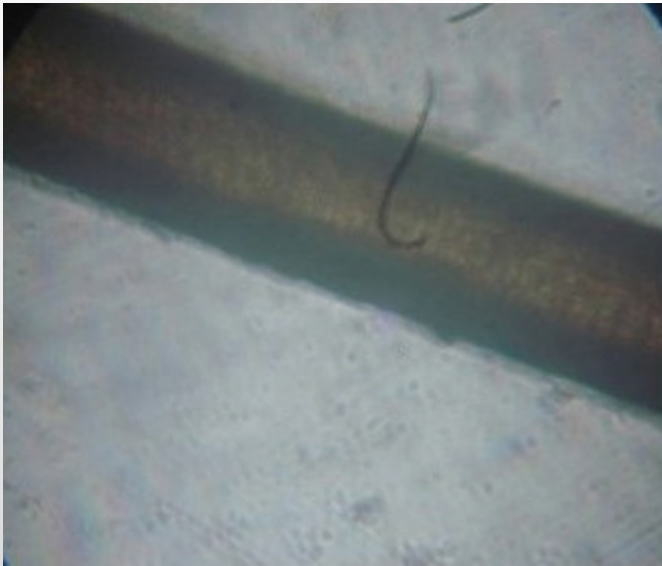
- SHE HAD SILVERY GRAY HAIR,
- MULTIPLE ILL-DEFINED HYPOPIGMENTED MACULES OVER THE FACE, NECK, PERICHONDritis OF RIGHT EAR,
- MULTIPLE EROSIONS COVERED WITH CRUSTS OVER BOTH UPPER LIMBS AND LOWER LIMBS.
- HEPATOMEGALY PRESENT.

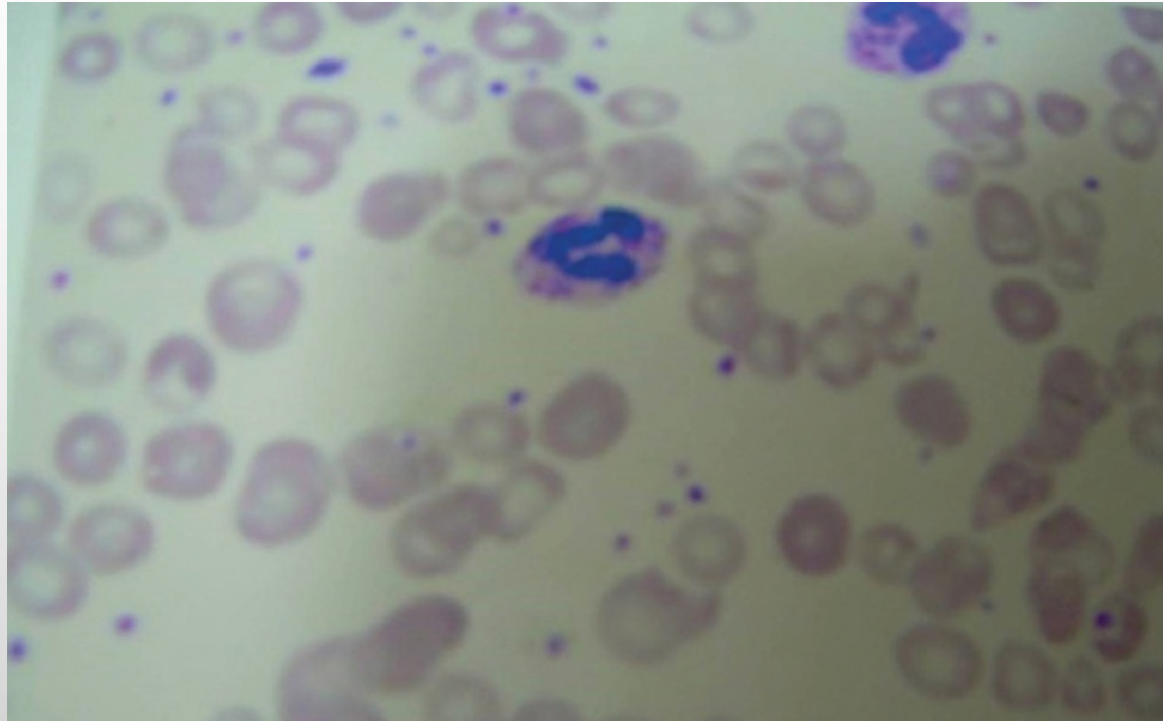




# INVESTIGATIONS

- BLOOD INVESTIGATIONS WERE WITHIN NORMAL LIMITS.
- HAIR SHAFT EXAMINATION REVEALED REGULARLY ARRANGED CLUMPS OF MELANIN.
- PERIPHERAL SMEAR SHOWED GIANT GRANULES IN LEUKOCYTES





# DIAGNOSIS AND TREATMENT.

IN OUR CASE CHILD HAD SILVERY GRAY HAIR, RECURRENT SKIN INFECTIONS, PERIPHERAL SMEAR EXAMINATION SHOWING GIANT GRANULES WITHIN LEUKOCYTES AND HAIR SHAFT EXAMINATION SHOWING SMALL CLUMPS IN REGULAR PATTERN FAVOURING DIAGNOSIS OF **CHEDIAK HIGASHI SYNDROME**. THE CHILD WAS REFERRED TO HIGHER CENTRE FOR BONE MARROW TRANSPLANTATION. WITHOUT BMT CHILDREN WITH CHS DIE BEFORE AGE OF 10 YEARS.



# CASE 2

DR VISHAL GUPTA

AIIMS, DELHI

- A 25-YEAR-OLD, OTHERWISE HEALTHY, MAN CONSULTED US WITH THE COMPLAINTS OF MULTIPLE MILDLY PRURITIC “WHITE SPOTS” ON HIS TRUNK. THE LESIONS, WHICH WERE FIRST NOTICED 7 YEARS BACK, HAD BEEN GRADUALLY INCREASING IN NUMBER.
- A PREVIOUS BIOPSY, DONE BY ANOTHER DERMATOLOGIST, WAS REPORTED NON-DIAGNOSTIC AND THE PATIENT WAS REFERRED TO US FOR FURTHER EVALUATION.

1. WHAT OTHER RELEVANT SITES WOULD YOU LIKE TO EXAMINE?
2. WHAT ARE YOUR DIFFERENTIAL DIAGNOSES?
3. HOW WOULD YOU LIKE TO INVESTIGATE FURTHER, AND CONFIRM YOUR DIAGNOSIS?



- DIFFERENTIAL DIAGNOSES SUGGESTED SO FAR INCLUDE, IN NO PARTICULAR ORDER ARE:
- PITYRIASIS VERSICOLOR
- FOLLICULAR LICHEN PLANUS
- GUTTATE LICHEN SCLEROSUS
- GUTTATE MORPHEA
- GUTTATE LEUKODERMA
- IGH
- DARIERS DISEASE (HYPOPIGMENTED)
- DOWLING DEGOS DISEASE (HYPOPIGMENTED)
- CLEAR CELL PAPULOSIS

- BLASCHKOID PATTERN, PARTICULARLY APPARENT IN PIC 2 WHERE LESIONS SEEM TO BE ARRANGED IN A WAVY STREAK TRAVERSING THE FLANK.
- ON EXAMINATION, THE PATIENT ALSO HAD RELEVANT CUTANEOUS FINDINGS ON BILATERAL TEMPORAL FOREHEAD EXTENDING TO THE SCALP, AND LEFT PALM (PICS ATTACHED).
- IN RESPONSE TO YOUR QUESTIONS, THE FAMILY HISTORY WAS NEGATIVE. ORAL CAVITY, GENITAL MUCOSA, AND NAILS WERE UNINVOLVED. FLEXURES (NECK AND AXILLAE) HAD A FEW SIMILAR SCATTERED HYPOPIGMENTED LESIONS, BUT WERE NOT PREFERENTIALLY AFFECTED. THERE WAS NO PHOTODISTRIBUTED PATTERN EITHER.
- KOH SCRAPING AND ANA WAS NOT DONE.
- A SKIN BIOPSY FROM ONE OF THE HYPOPIGMENTED MACULES WAS DONE

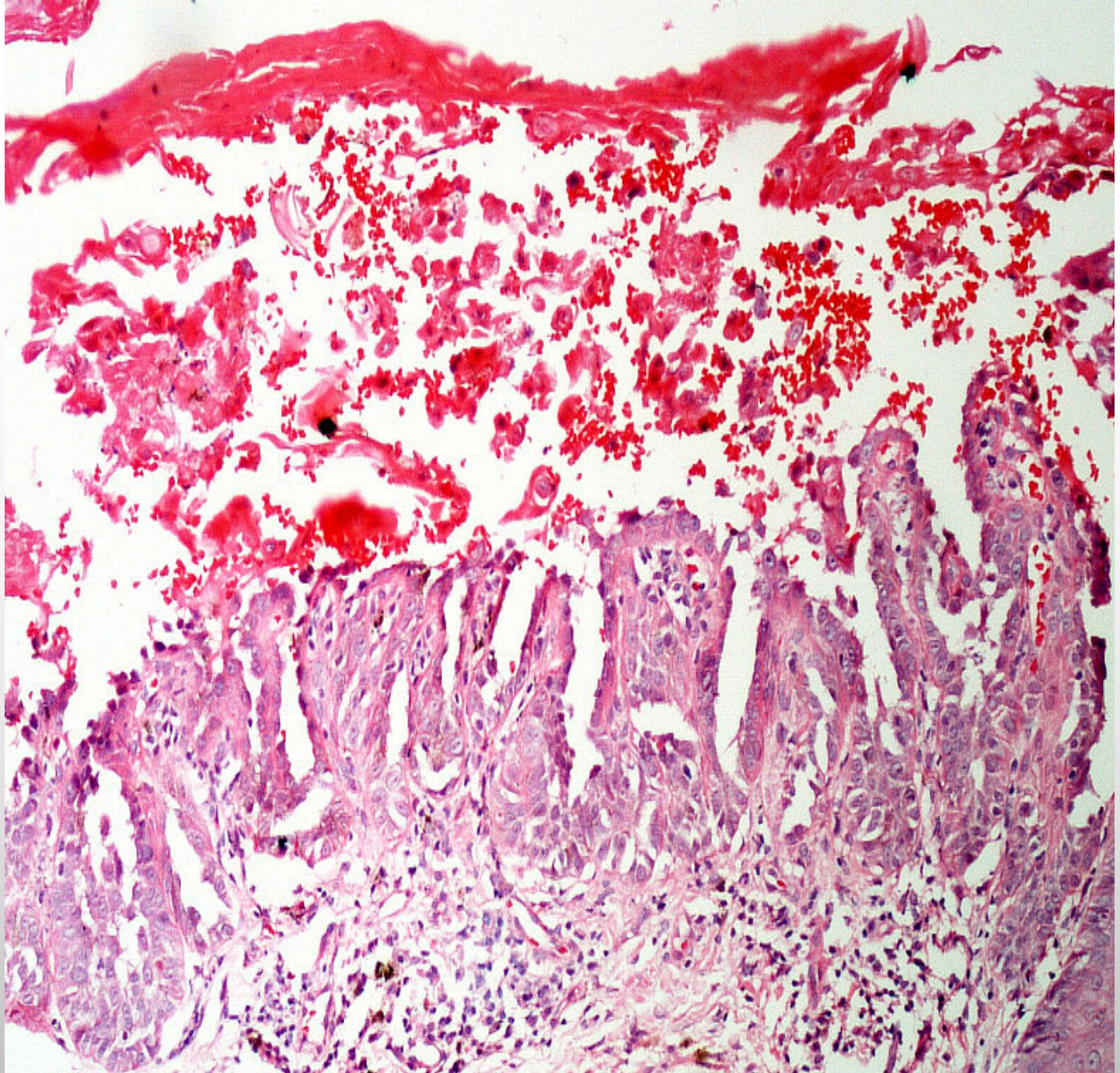
- DIAGNOSIS IS INDEED DARIERS DISEASE.
- THE HYPOPIGMENTED MACULES AND PAPULES AND EVEN THE KERATOTIC LESIONS ON THE FOREHEAD WERE IN A LINEAR PATTERN. THE PALMAR PICTURE SHOWS PITS ON THE CREASES.
- GUTTATE 'LEUKODERMA' IS WELL DESCRIBED AS A CUTANEOUS FINDING IN DARIERS DISEASE. RARELY, THIS MAY BE THE PROMINENT (OR EVEN THE ONLY) FINDING, AND OTHER CHARACTERISTIC FEATURES MAY HAVE TO BE SEARCHED FOR, LIKE IN THIS CASE. ANOTHER POINT OF INTEREST WAS THE BLASCHKOID PATTERN OF LESIONS.
- THE CLINICAL DIAGNOSIS WAS CONFIRMED ON HISTOPATHOLOGY. THE SKIN BIOPSY SHOWED CHARACTERISTIC FEATURES OF A SUPRABASAL SPLIT AND DYSKERATOTIC ACANTHOLYTIC CELLS IN THE CLEFT AS WELL AS IN THE STRATUM CORNEUM AND GRANULAR LAYER.



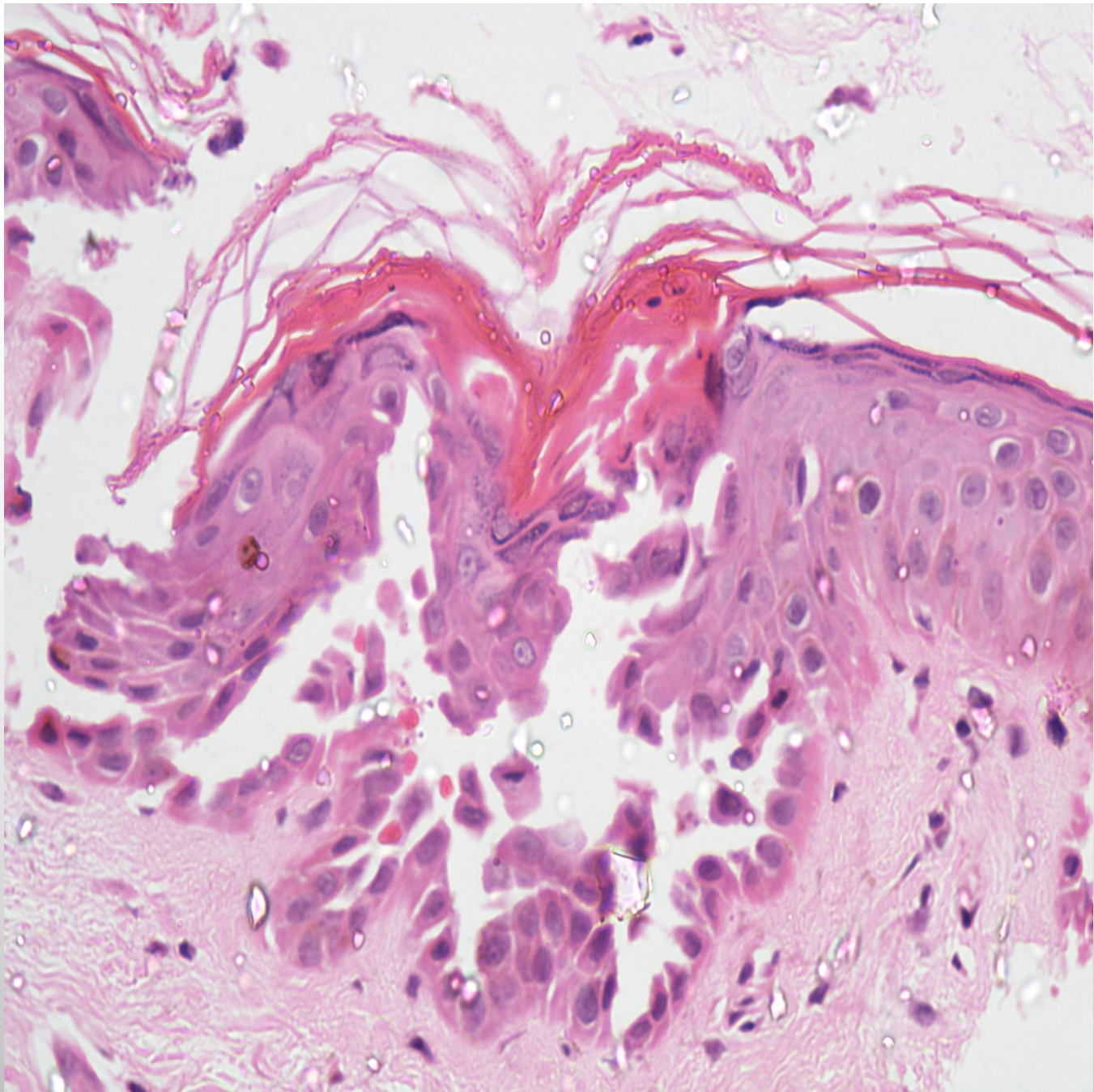










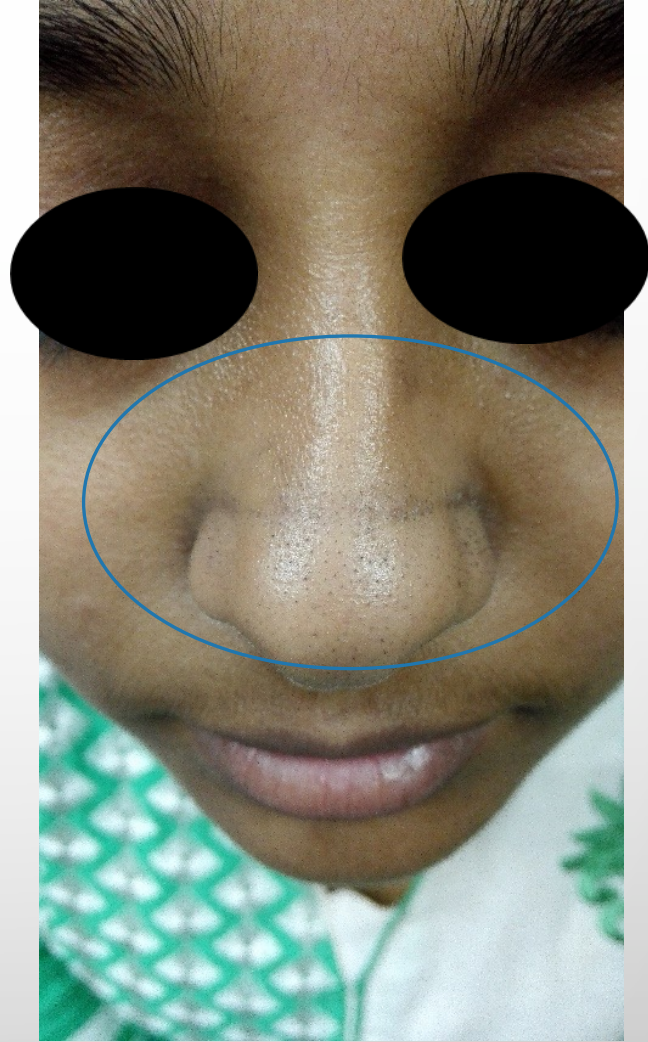
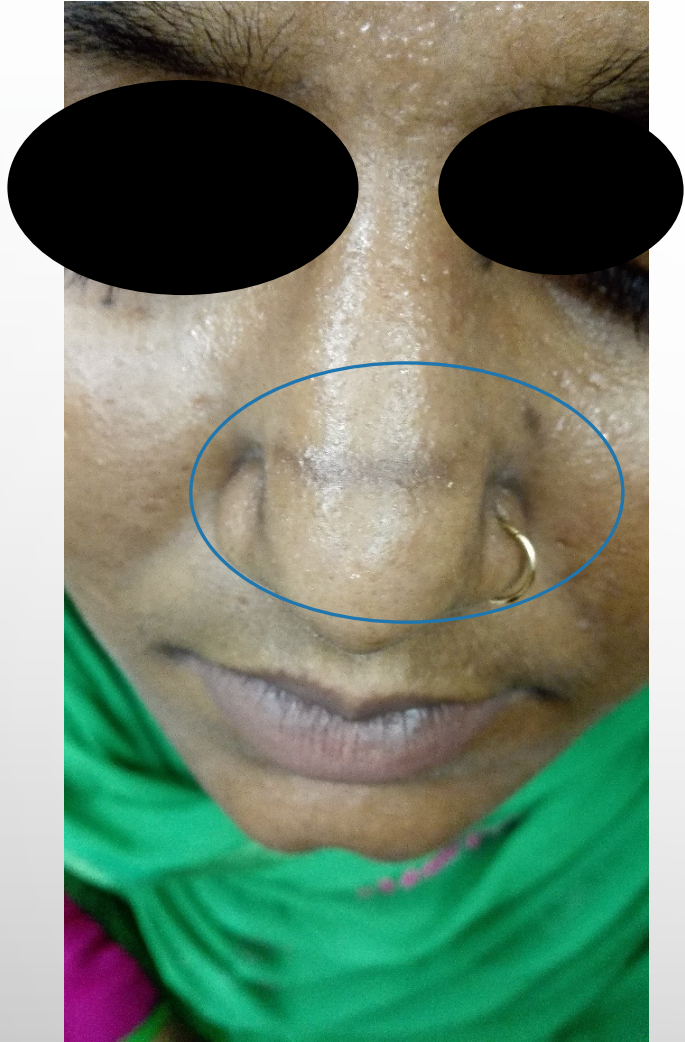


# CASE 3

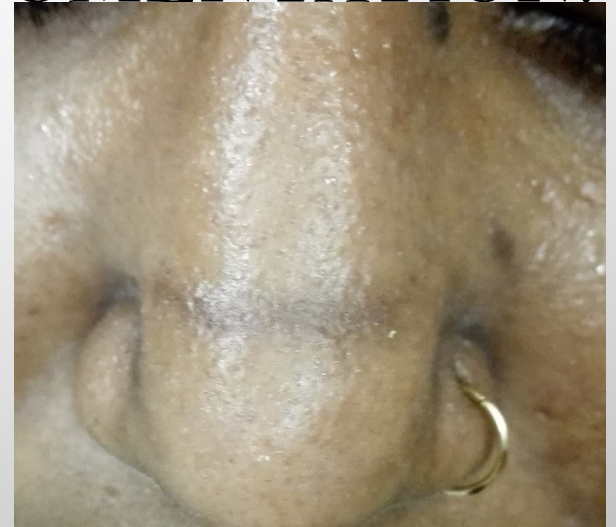
DR SANJAY RATHI

CONSULTANT DERMATOLOGIST

SILIGURI



- NOMENCLATURE / DIAGNOSIS?
- DO YOU SEE THIS IN YOUR PRACTICE?
- DO YOU WORK-UP? ANY ASSOCIATIONS?
- HOW TO TREAT THIS PIGMENTATION?



**DIAGNOSIS: PIGMENTED TRANSVERSE**

**NASAL BAND**

✓ **PIGMENTED TRANSVERSE NASAL**

**BAND** IS AN UNDER-REPORTED

ENTITY IN DERMATOLOGY

✓ IT REPRESENTS TRANSVERSE NASAL

LESION HAVING VARIOUS

MORPHOLOGICAL TERMS AS GROOVE,

STRIP, LOOP, RIDGE

✓ IT MAY BE AN EMBRYOLOGICAL

FAULT LINE

✓ **ON DERMOSCOPY,** TRANSVERSE NASAL  
CREASE DEMONSTRATES INTERMITTENT,  
THIN, WHITE TRANSVERSE STREAKS WHICH  
ARE MORE PROMINENT AT THE CENTER OF  
NOSE AND GRADUALLY FADE AWAY TOWARD  
THE PERIPHERY. IT IS HYPOTHESIZED  
THAT THESE STREAKS REPRESENT FIBROSED  
ADHESIONS TO THE NASAL CARTILAGE.

✓ IT MAY BE ASSOCIATED WITH SEBORRHEIC DIATHESIS ,  
DPN,

ICHTHYOSIS, ATOPIC, SEBORRHEIC MELANOSIS ,ACNE.  
AMONG ALL OF THE ABOVE, IT IS **MOST COMMONLY**  
**ASSOCIATED WITH SEBORRHEIC DERMATITIS, SEB**  
**MELANOSIS**

✓ **REASSURANCE** IS NEEDED

✓ TOPICAL RETINOIDS/ MILD TS MAY BE USED TO LIGHTEN  
THE PIGMENTATION TEMPORARILY

✓ SURGICAL INTERVENTION TO BE AVOIDED TO PREVENT  
UNDERLYING STRUCTURE DAMAGE





# **CASE: 4**

**DR. ATIYA YASEEN**

**Srinagar, J&K**

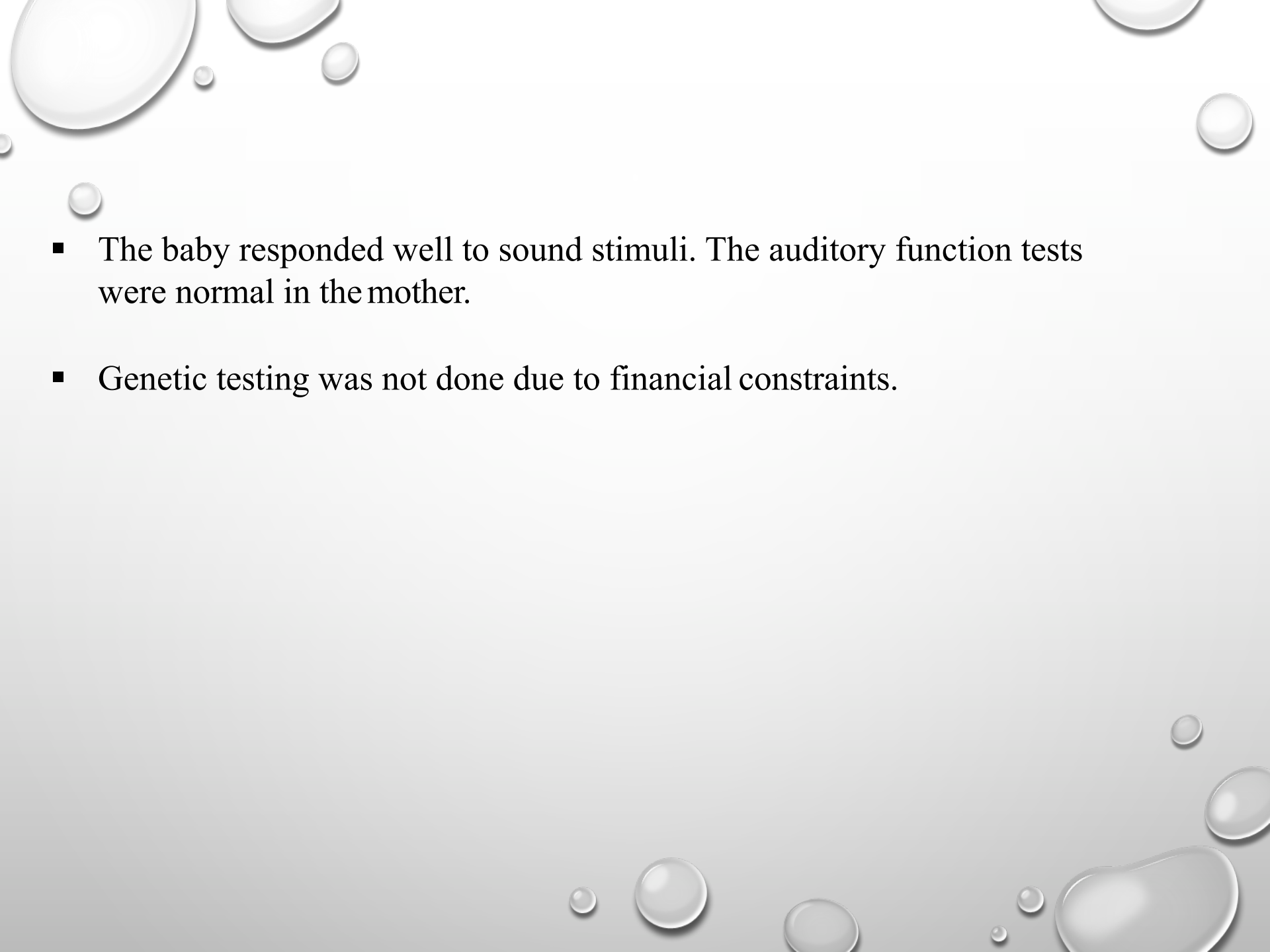


## HISTORY:

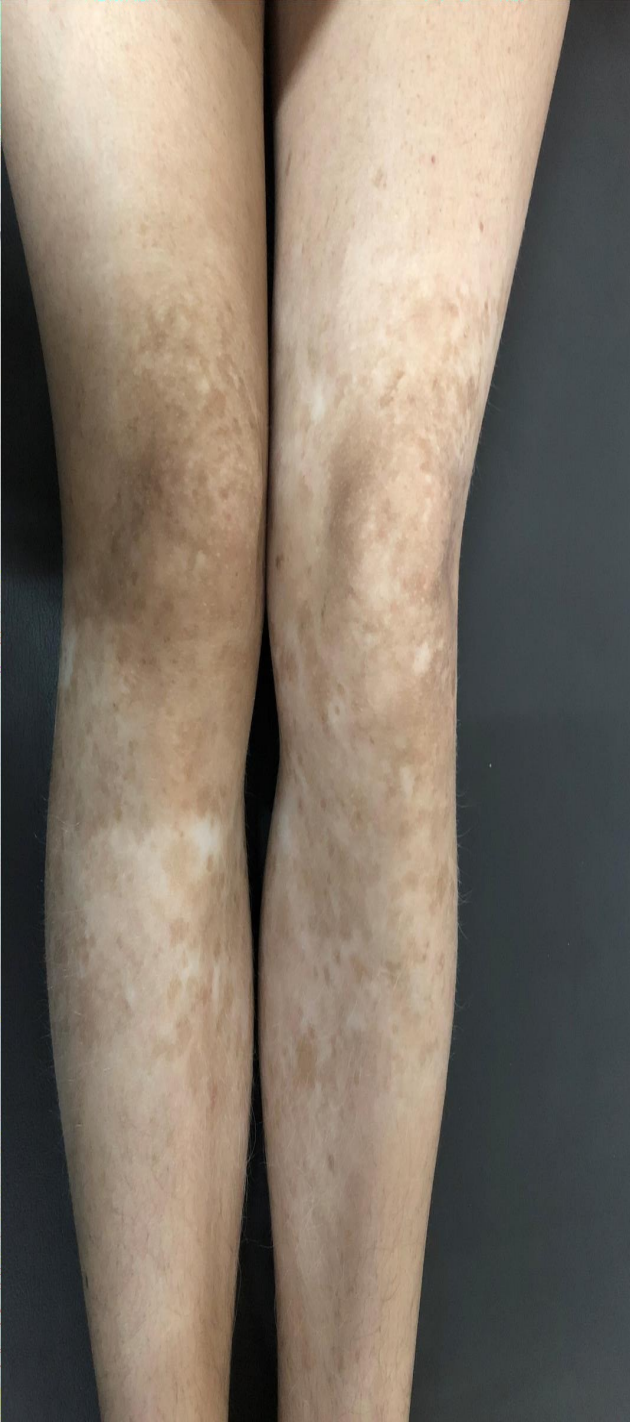
- Parents of a 6 month old boy (1<sup>st</sup> in birth order) complained of asymptomatic white hairs & white patches on his body since birth.
- The child's mother had white hairs & similar white patches on her trunk & legs since birth.
- The lesions were stable since birth in both mother & son.
- There was no history of consanguinity amongst the parents of the child or his mother.
- The child had normal development and milestones.

# EXAMINATION:

- Large depigmented macules were seen over the ventral part of mid trunk of the child . Macules of normally pigmented skin were interspersed within the depigmented macules.
- A triangular well circumscribed forelock in the midfrontal region with a depigmented macule on the forehead was also seen. The hairs of the left eyebrow at the medial end also showed depigmentation.
- Mother had similar macules over the mid portion of the lower extremities on the anterior aspect, lower chest & abdomen. The depigmented macules were associated with leucotrichia. She also presented with some depigmented hairs in the mid frontal region & an underlying depigmented macule.

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- The baby responded well to sound stimuli. The auditory function tests were normal in the mother.
  - Genetic testing was not done due to financial constraints.





# *Diagnosis:* **Piebaldism**

Piebaldism is a condition characterized by the absence of cells called melanocytes in certain areas of the skin and hair. Melanocytes produce the pigment melanin, which contributes to hair, eye, and skin color. The absence of melanocytes leads to patches of skin and hair that are lighter than normal. Approximately 90 percent of affected individuals have a white section of hair near their front hairline (a white forelock). The eyelashes, the eyebrows, and the skin under the forelock may also be unpigmented. The unpigmented patches are at increased risk of sunburn and skin cancer related to excessive sun exposure. Aside from these potential issues, this condition has no effect on the health of the affected individual.

Piebaldism can be caused by mutations in the KIT and SNAI2 genes. Piebaldism may also be a feature of other conditions, such as Waardenburg syndrome; these conditions have other genetic causes and additional signs and symptoms. This condition is inherited in an autosomal dominant pattern

THANK YOU