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RESEARCH ARTICLE

A CASE SERIES OF VARIANTS IN NEVUS DEPIGMENTOSUS

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Abstract

This case series presents unique variantsof Nevus Depigmentosus in pediatric patients with widespread distribution. Nevus Depigmentosus, well known as Achromic nevus, is a rare hypopigmentary skin disorder that usually presents in childhood. Variants of this condition exhibit diverse clinical presentations and distinct distribution pattern, thereby posing diagnostic challenges. By exploring this rare dermatological phenomenon in pediatric context, we aim to contribute to the limited literature available on Generalized and Segmental Nevus Depigmentosus in this age group, ultimately improving clinical awareness and patient care.

Introduction:-

Nevus Depigmentosus (ND) is a rare and intriguing pigmentary skin lesion characterized by areas of hypopigmented or depigmented skin, resulting from the absence of melanocytes. This condition was firstly described by Lesser in 1884.[1]Variants of Nevus Depigmentosusare been documented in various age groups, but its occurrence in children as young as 3 years old, with generalized distribution is underreported in medical literature. This condition while uncommon presents with a variety of clinical manifestations that contribute to its complexity and challenge in diagnosis. Nevus Depigmentosusneeds to be differentiated from nevus anemicus, hypomelanosis of Ito, vitiligo, albinism etc.Among the diverse spectrum of nevus depigmentosus variants, the generalized distributed presentation stands out as a distinct and infrequent manifestation.

Cases Description:

Case1, a 3-year-old female child came to the dermatologic clinic with complaints of hypopigmented skin lesions. The parents reported that these lesions were first noticed a year back andhave gradually increased in size and number. There were no associated symptoms. There is no history of similar complaints in family. Upon clinical examination, the child presented with extensive hypopigmented asymptomatic macules of varied sizes over trunk and extremities. The macules were distributed bilaterally, and crossed the midline. The lesions measured of 1mm diameter. There were few hypopigmented macules of approximately 3 cm diameter over right shoulder, lower abdomen, gluteal region. The borders were well defined with irregular serrated margins that contrast with the surrounding normal skin. There were no signs of scaling, erythema or atrophy. Mucous membrane, Oral cavity, Hair and Nail appear normal. The overall development and general health were normal.Patientdid not show cutaneous lesions other than nevus depigmentosus, and no apparent congenital anomalies were observed.

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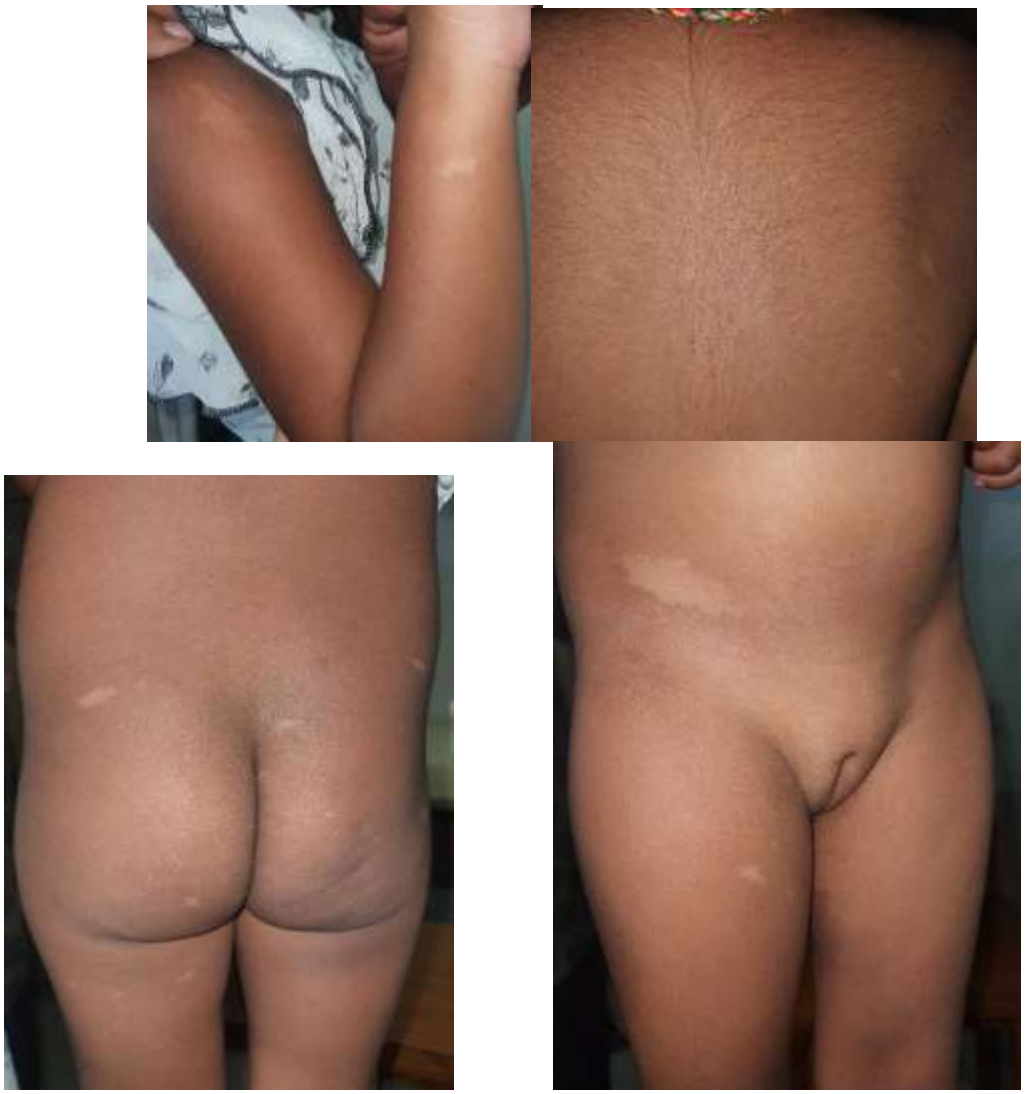
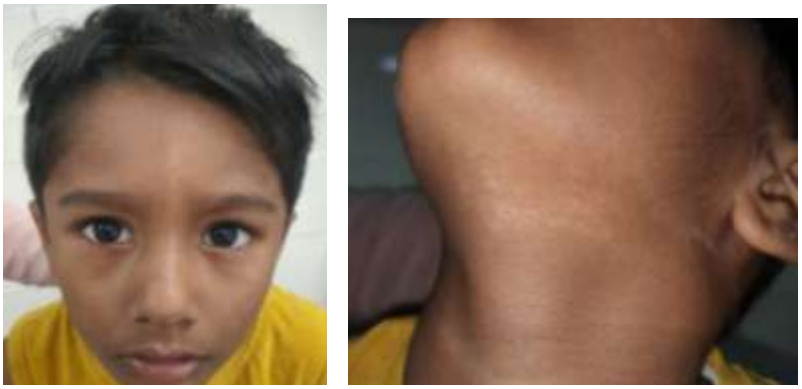
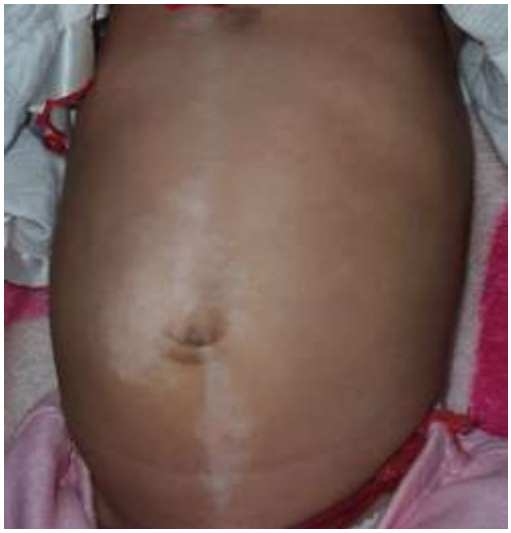


Case 2 was a 3-year-old male child presented withhypopigmentedpatch over face and neck. The lesions presented since 6 months. Thepatch over forehead is arranged in a linear pattern with irregular border.They were asymptomatic with no signs of scaling, erythema or atrophy.Mucous membrane, Oral cavity, Hair and Nail appear normal. The overall development and general health were normal.Patient did not show cutaneous lesions other than nevus depigmentosus, and no apparent congenital anomalies were observed.



Case 3 was a 1.5-year-old female child presented with hypopigmented patchoverabdomen.The borders were well defined with irregular and serrated margins.They were asymptomatic.There were no signs of scaling, erythema or atrophy. Mucous membrane, Oral cavity, Hair and Nail appear normal.



Diagnostic Evaluation:
Under Wood's lamp examination, the lesion of ND showed an off-white accentuation without fluorescence.On Diascopy, the lesions were non-blanchable with intact border. To confirm the clinical suspicion a skin punch biopsy was performed on a hypopigmented macules. The histopathological analysis revealed reduced number of melanocytes along with reduced melanin pigment. Although complete loss was not evident, the biopsy was suggestive of Nevus Depigmentosus.

Discussion:-

Nevus Depigmentosus (ND) presents as a stable, discrete, hypopigmented macule or a patch, whose size and distribution remains the same throughout life.[2-4]El-Sayed S. Hewedy et al[5] found age incidence mostly present before 3 years (73.33% cases had the initial lesion before the age of 3 years) as in our cases (100%).Our cases had lesions with serrated borders which is in line with El-Sayed S. Hewedy et al.[5]So, the diagnosis of ND can be considered if patient develops hypopigmented macule with irregular or serrated border at an early age.Nevus Depigmentosusmay be focal, segmental or generalized in distribution most commonly on the trunk.The main pathology is secondary to aggregation of melanosomes in the melanocytes and a defective transfer of the same to the surrounding keratinocytes.There is reduced melanosomalcontentoflesional keratinocytes and melanocytes. Immature and aggregated melanosomes are present more in lesional keratinocytes.Given the benign nature of Nevus Depigmentosus and absence of associated symptoms, a conservative management was done initially. Counselling regarding photoprotection had been given. Regular follow up was done to monitor any changes. No specific treatment was effective or was necessary. But for cosmetic purpose 308 nm excimer laser and surgical procedures like blister roof grafting, cultured epidermal grafting, autologous epidermal cell grafts had been used with good results which has been consistent with Abhijeet Kumar Jha et al.[6] Park et al.[7] also stimulated melanogenesis in ND by using a 308-nm excimer laser and observed marked improvement of the ND lesion.

Conclusion:-

This case series highlights the clinical presentation, diagnosis, and management of Generalized Nevus Depigmentosus in Case1,Segmental Variant in Case 2 and Case3 in paediatric context. The patients hypopigmented macules remained stable during 12 months since presentation. So, though conservative management would be a better option in management of cases ofnevus depigmentosus, excimer laser and surgical procedures like blister roof grafting, cultured epidermal grafting, autologous epidermal cell grafts can be used with good cosmetic results.

Further research is needed to understand the genetic factors and potential treatments for Nevus Depigmentosus.

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