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Idiopathic Developmental Dysplasia of Hip in a Female Child with a Rare Epidermal Syndrome- A Case Report

Eshaan Mishra[1], Nirmal C Mohapatra[1], Rajesh Rana[1], Soumitesh S Das[1], Chandrakanta Mishra[2]

Learning Point of the Article:

DDH can be associated with epidermal syndromes even though the musculoskeletal system is of mesodermal origin.

Abstract

Introduction: Developmental dysplasia of the hip (DDH) describes a spectrum of disorders affecting the neonatal hip. Trachyonychia or twenty nail dystrophy refers to thin, brittle nails with excessive longitudinal ridging affecting all twenty nails. Alopecia universalis congenita (ALUNC) is a rare anomaly affecting skin and appendages. It shows a genetic preponderance with its autosomal recessive variety being the most common and severe variety.

Case Report: We report a case showing idiopathic DDH in a female child with a rare epidermal syndrome consisting of Trachyonychia and ALUNC. The cutaneous symptoms show familial inheritance in the form of autosomal dominant inheritance.

Conclusion: This case report highlights the fact that DDH can be associated with other syndromes which require multidisciplinary evaluation and research.

Keywords: Developmental dysplasia of the hip, alopecia universalis congenita, Trachyonychia, autosomal dominant inheritance.

Introduction

Developmental dysplasia of the hip is used to describe a spectrum of disorders affecting the neonatal and infant hip joints [1]. The exact etiology is not known [2]. It is believed to be of multifactorial nature: a combination of genetic, environmental, and mechanical factors [3]. Many cases are syndromic and have underlying neuromuscular disorders [4].

Alopecia universalis congenita (ALUNC) is a rare cutaneous anomaly. It can show autosomal recessive, x-linked recessive, or autosomal dominant inheritance, of which autosomal recessive is the most common and severe phenotype [5].

Trachyonychia, also known as Twenty-nail dystrophy, refers to thin, brittle nails with excessive longitudinal ridging [6]. Some common skin conditions associated with Trachyonychia include alopecia areata, psoriasis, lichen planus, atopic dermatitis, and ichthyosis vulgaris amongst others [7].

We report a case of a 4-year-old female child with Idiopathic developmental dysplasia of the hip (DDH) of the left hip in association with a rare epidermal syndrome, consisting of ALUNC and Trachyonychia, with a family history of the epidermal symptoms spanning four generations highlighting its genetic preponderance. It is the first case report of this kind in literature.

Case Report

A 4-year-old female child was brought to our OPD by her parents

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Author's Photo Gallery


Dr. Eshaan Mishra


Dr. Nirmal C Mohapatra


Dr. Rajesh Rana


Dr. Soumitesh S Das


Dr. Chandrakanta Mishra

[1]Department of Orthopaedics, SCB Medical College and Hospital, Cuttack, Odisha, India,
[2]Department of Cardiology, SCB Medical College and Hospital, Cuttack, Odisha, India.

Address of Correspondence:
Dr. Eshaan Mishra,
Department of Orthopaedics, SCB Medical College and Hospital, Cuttack, Odisha, India.
E-mail: eshaanmishra271@gmail.com

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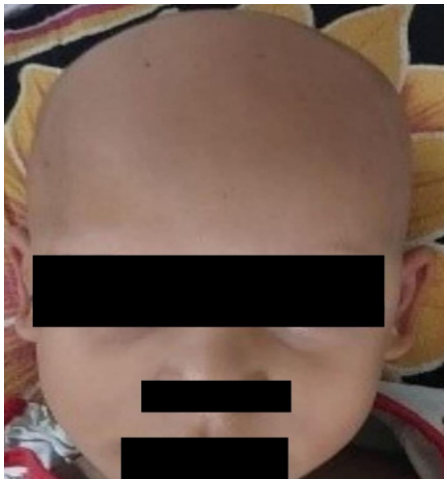

Figure 1: Alopecia universalis congenita in female child.

Figure 2: Nail dystrophy of all 10 upper limb digits of female.

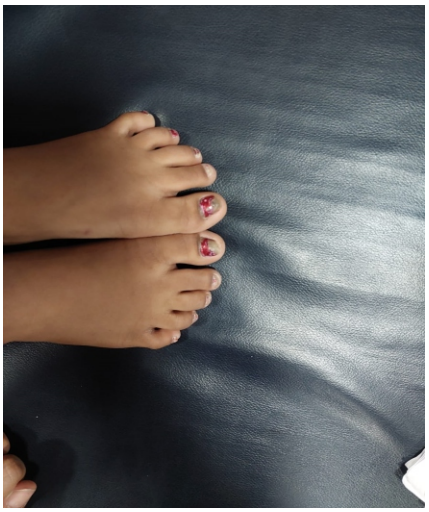

Figure 3: Nail dystrophy in all 10 lower limb toes of female.

with a chief complaint of painless limping since the child started walking. She is the first-born female child. There is no significant antenatal or perinatal history. There is no history of abnormal drug intake or radiation exposure of the mother during pregnancy. On general examination, the child was of average build for her age and appeared to have normal intelligence. During a detailed head-to-toe examination, she was found to have features suggestive of ALUNC (Fig. 1) and Trachyonychia (Figs. 2 and 3) with a family history spanning four generations (Figs. 4, 5, and 6- Alopecia universalis and Trachyonychia in father of patient). There is no history of hip dysplasia in any of the other family members.

Pedigree analysis (Fig. 7) revealed it to have an autosomal dominant inheritance pattern.

Hip examination revealed Trendelenburg gait, exaggerated lumbar lordosis, proximal migration of left greater trochanter, asymmetric thigh crease, fixed flexion deformity of left hip of 30°, restricted abduction, increased rotational range of movement of left hip, apparent and true left limb supra-trochanteric shortening of 1.8 cm, Galeazzi sign positive, Telescopy test positive, Trendelenburg test positive, vascular sign of Narath positive. There was no evidence of any other skeletal dysplasia, ectodermal dysplasia, or visceral abnormality. There were no associated neuromuscular signs.

All routine blood investigations were normal. Ultrasonography did not reveal any visceral abnormality. On roentgenography (Fig. 8), the left hip was dislocated with the femur head in the superolateral Perkins quadrant. The left acetabulum was dysplastic. The left acetabular index was 42°. Shenton's arc was broken on the left side. A non-contrast CT scan revealed a

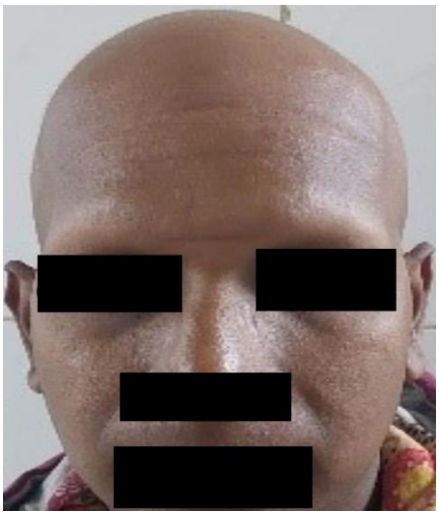

Figure 4: Alopecia universalis congenita in the father of the patient.

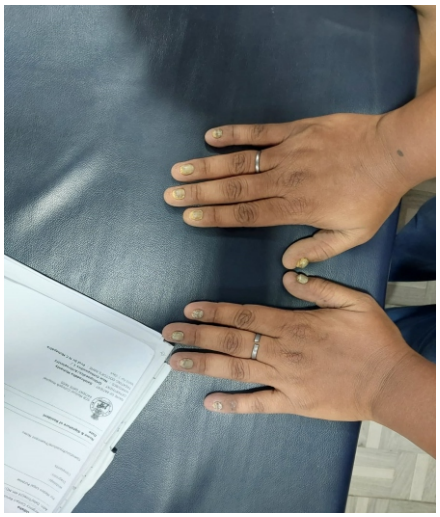

Figure 5: Nail dystrophy of all 10 upper limb digits of the father of the patient.

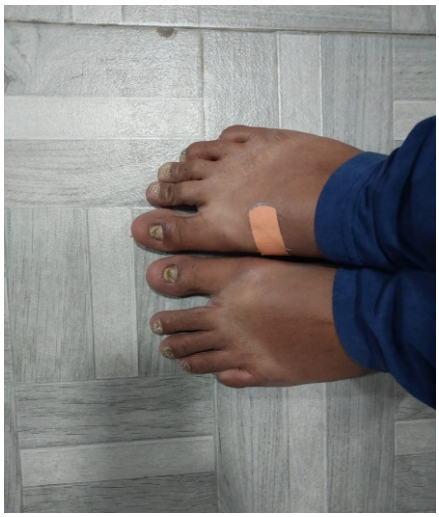

Figure 6: Nail dystrophy of all 10 lower limb toes of the father of the patient.

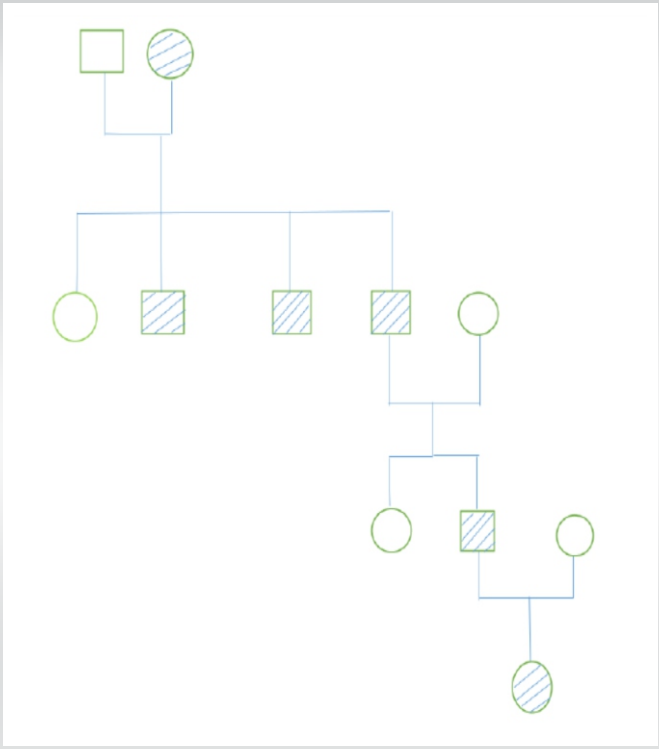

Figure 7: Pedigree analysis - Inheritance pattern of Trachyonychia and alopecia universalis congenita in patient's families across 4 generations.

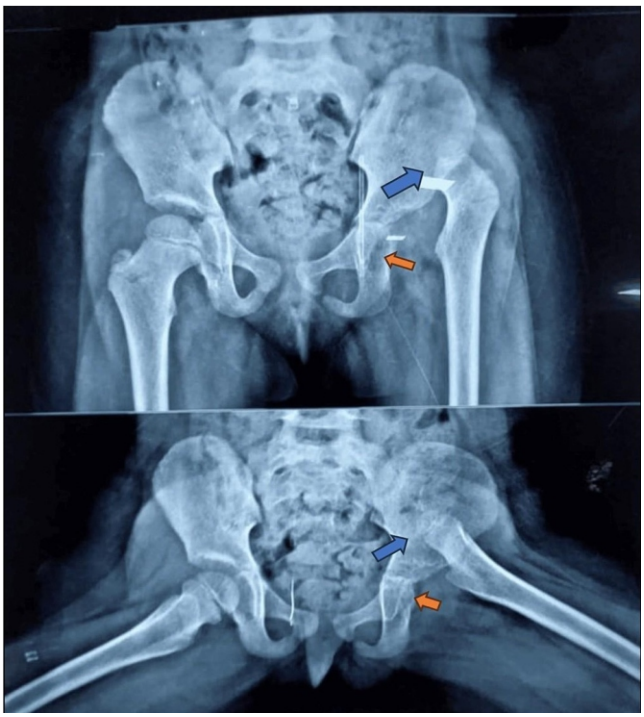

Figure 8: X-ray dislocated left hip of the patient. Blue arrow - Displaced and hypoplastic femoral epiphysis. Orange arrow – Dysplastic and shallow acetabulum.

shallow acetabulum, a superolaterally displaced femur head, and a small hypoplastic femoral epiphysis.

With the parents' consent and after a proper pre-anesthetic check-up, the patient's left hip was planned for open reduction of the femur head with pembertons osteotomy of acetabulum and varus derotation osteotomy of the proximal femur.

Across the iliofemoral Smith-Peterson approach, the iliac apophysis was divided into two halves and dissected laterally. The origins of gluteus medius and gluteus minimus were elevated subperiosteally. The dislocated head was approached after opening the left hip joint capsule. The fibrous tissue, hypertrophied ligamentum teres, and pulvinar fat from the acetabulum were excised. The head was reduced into the native acetabulum and subtrochanteric osteotomy was done. Subtrochanteric varus derotation osteotomy was done and fixed with a 3.5mm recon plate. A shortening of 1cm was done. Pembertons osteotomy was done, to increase the acetabular roof coverage, with a triangular graft from the osteotomised femur bone. The hip was immobilized in a hip spica cast for 8 weeks.

Discussion

DDH refers to a variety of conditions related to hip development [10]. The condition is idiopathic in most of the cases. The left hip is affected in 60% of idiopathic cases [8]. DDH is known to be associated with various neuromuscular syndromes, such as spina bifida and cerebral palsy. Teratologic dysplasia may be seen with Arthrogyposis, Down syndrome, and Ehlers-Danlos syndrome [9]. Our patient is likely a case of idiopathic DDH and a rare epidermal syndrome consisting of Alopecia universalis and Trachyonychia. There is only a single previous case report, describing two siblings with similar cutaneous symptoms [5]. However, this is the first case report showing the inheritance of the cutaneous symptoms (or syndrome) across 4 generations of a family.

Conclusion

DDH can be idiopathic or associated with a syndrome. Some of the syndromes with DDH having cutaneous manifestations include Ehlers-Danlos syndrome and Arthrogyposis multiplex congenita. This case report shows the occurrence of Idiopathic DDH in a female child with a rare epidermal syndrome consisting of ALUNC and Trachyonychia with an autosomal dominant inheritance pattern. This seems to be the first case report showing such a clinical association. Moreover, highlights the necessity of further research into the etiology of DDH and its possible association with other identified and unidentified syndromes.

Clinical Message

This case report highlights the importance of multidisciplinary approach to patient care. Research into the etiopathogenesis of DDH and its association with other syndromes is an ongoing endeavour.Clinicians must be vigilant of these associated syndromes while dealing with patients with DDH and ensure holistic care for their patients. Some of these syndromes may have a genetic preponderance and hence genetic analysis of these patients is of paramount importance.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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Consent: The authors confirm that informed consent was obtained from the patient for publication of this case report

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