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Nevus comedonicus syndrome: An unusual case report

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Abstract
A rare syndrome characterized by the association of typically unilateral, closely arranged, linear, slightly elevated, multiple, nevus comedonicus lesions located usually on the face, neck, trunk and both limbs with extracutaneous presentation within the variety of syndactyly, polydactyly, valgus deformity in lower limb and scoliosis. Our case presented with additional findings of bilateral congenital inguinal hernia with atrophied right testis and retractile left testis, which has not been mentioned before.

Keywords: Nevus comedonicus, syndactyly, polydactyly, congenital hernia

Introduction
The nevus was described in 1895 under the term "comedo nevus," the standard lesion is an overgrowth of clumps of papules, usually with a central black firm center. These are morphologically blackheads, later evolving into inflammatory acne.
An uncommon organoid nevus of epithelial origin, nevus comedonicus consists of linear plaques of plugged follicles that simulate comedones; they will be present at birth or may appear during childhood. The horny plugs represent keratinous debris within dilated, malformed pilosebaceous follicles. The lesions are most frequently unilateral and should develop at any site. Rarely, they're associated with other congenital malformations, extracutaneous manifestations which may include central system (CNS), skeletal, opthalmological, neurological, and spinal abnormality. We herein report the case of a 37 year old male with nevus comedonicus syndrome with classical unilateral black comedones present in his right half of body along with skeletal and spinal anomalies, there was rare gonadal lesions in addition to a congenital bilateral inguinal hernia.

Case Report
A 37year old male came to the OPD with the complain of congenital left inguinal swelling which was causing him discomfort from last 4 months, he was operated for right inguinal hernia when he was 7 years old. He also gave the history of black papules over his right 1/2 of body and supernumerary and webbed few fingers of his upper and lower limbs. On examination closely arranged, linear, slightly elevated, multiple, nevus comedonicus lesions with a central, dark, firm, hyperkeratotic plug were located on the right half of the body (face, neck, trunk and both limbs) of the patient. The other findings present within the patient was syndactyly of left hand ring and small finger, right foot great toe and second toe, left foot 2^{nd},3^{rd} and 4^{th} toe, with six toe in left foot, had got excision of his supernumerary finger of his right hand, had valgus deformity of his leg bones, had scoliosis. There was a retractile left testis with left inguinal herniation which was reducible and had positive cough impulse; scar of previous right inguinal hernia surgery was evident with comparatively small right testis. He didn't present with any neurological deficit nor did he have any ophthalmic complain.
He was investigated and his ECG and Echo were normal, all his blood investigations, chest X-ray and urine reports were normal. Ultrasound confirmed a left inguinal hernia with a 21mm defect. Spine x-ray confirmed scoliosis with few lumbar osteophytes. Left hernioplasty with left orchidopaxy was done and for his dermal lesions he was referred to dermatologist.

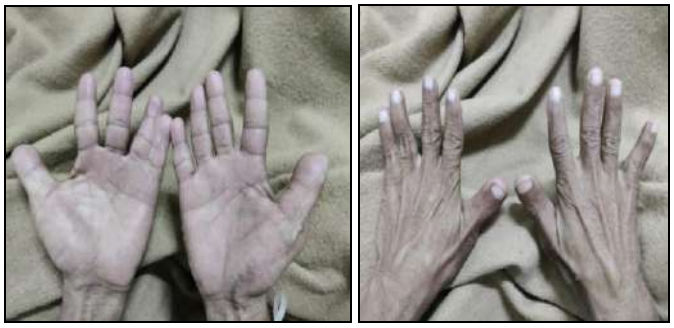

Fig 1: The Hands: Syndactyly left hand

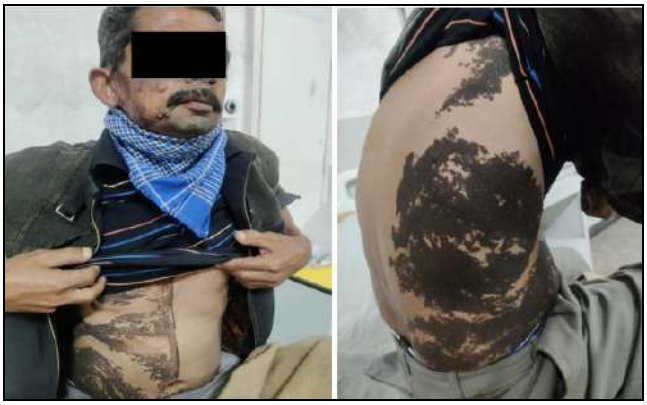

Fig 2: The Body: Nevus comedonicus Right half

Fig 3: The Foot: Polydactyly and syndactyly

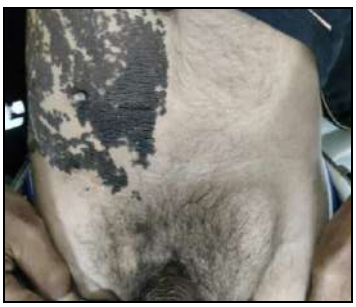

Fig 4: The Left inguinal Hernia

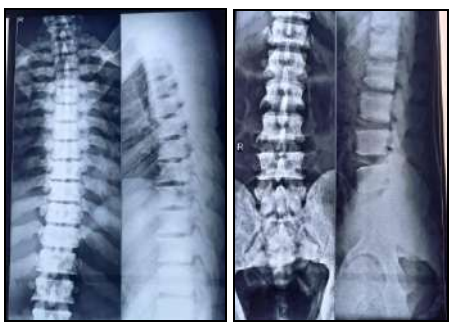

Fig 5: Spine X-ray: Scoliosis

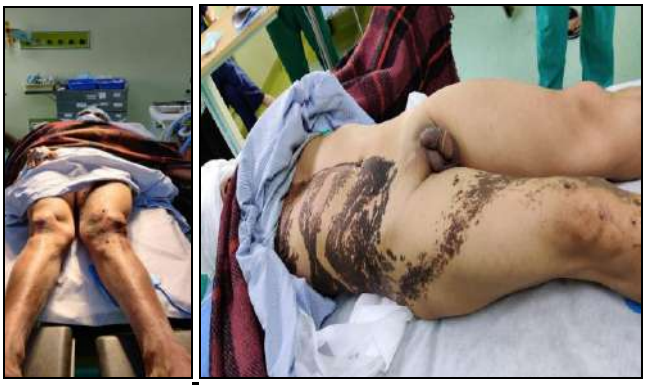

Fig 6: Valgus deformity and gonadal changes

Discussion

Nevus comedonicus could be a rare problem with an estimated occurrence of 1 case in every 45,000–100,000 individuals [1-5] Nevus comedonicus (NC) is an uncommon variant of adnexal hamartoma, clinically appearing as confluent clusters of open comedones [6]. Some view it as a morphologic variant of epidermal nevus [7]. NC may be a rare skin condition belonging to the spectrum of the epidermal nevi syndrome. Usually involving the face and neck area, it can present in unilateral, bilateral, linear, interrupted, segmental, or blaschkoid pattern. Nevus comedonicus is among those nevoid conditions to follow the so-called lines of Blaschko, which are linear patterns defined by mapping the cutaneous lesions themselves. Blaschko's lines don't conform to lines of cutaneous distribution such as Langer's lines, Voight's lines or any known lines of nervous, vascular or lymphatic structures within the skin. Explanations for the genesis of those lines include genetic mosaicism and yet undefined influences exerted by underlying mesenchymal tissues on the epidermis [9]. When it's related to non-cutaneous findings like skeletal, CNS, or ocular abnormalities, it is termed as nevus comedonicus syndrome [8]. In around 1/2 of the cases, it manifests shortly after birth with the foremost of the cases presenting before the age of 10. Our case presented with this syndrome since birth and therefore the lesions kept on growing to the current state.

Extracutaneous manifestations related to NC include electroencephalographic (EEG) abnormalities and seizures, dysgenesis of corpus callosum, microcephaly, ipsilateral cataract, and corneal changes. Skeletal anomalies have also been related to NC including hemivertebrae, scoliosis, polysyndactyly or clinodactyly, oligodontia, and absence of the fifth ray of a hand (Engber, 1978; Patrizi *et al*., 1998; Vidaurri-de la Cruz *et al*., 2004). It can also accompany the Klippel-Trénaunay and Sturge-Weber syndromes [3, 4]. Our case too presented with extracutaneous manifestations within the type of skeletal and spinal anomalies, and visceral lesions in the form of congenital hernia and testicular anomaly which stands apart this case with previously reported nevus comedonicus syndrome cases.

Recently, Happle has used the term "the epidermal naevus syndromes" to incorporate a minimum of six separate disorders including comedo naevus syndrome additionally to those with extracutaneous manifestations with verrucous epidermal nevus and nevus sebaceous [10].

The diagnosis is predominantly clinical, with biopsy only indicated in rare cases, especially in group 2. The histopathologic exam evidences an epidermis with dilated follicular openings, stuffed with horny material. The sebaceous glands and hair follicles are usually absent or rudimentary [5].

Conclusion

The reported case may be a rare syndrome with a novel right 1/2 of body from face to toe having classical cutaneous manifestation of nevus comidonicus with unusual skeletal congenital anomalies of syndactyly, polydactyly, valgus deformity and scoliosis and visceral involvement with bilateral congenital hernia with testicular anomalies and these last two anomalies aren't mentioned previously in any literature that adds to the associations that have been reported with NC syndrome. Furthermore, the pattern and distribution of NC is additionally unaccustomed to the literature.

Acknowledgments

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest

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