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Case Report

Cutis Verticis Gyrata in a Newborn with Turner syndrome

Dr. Pradipprava paria[1], Dr Khurram Hayat[2], Dr Shibani Paul[3], Dr Balai Chandra Karmakar[4]
[1,2]MD (Pediatrics), RMO-cum-Clinical tutor
[3]Senior Resident, [4]Associate Professor, Dept. of Pediatric medicine, R G Kar Medical College, Kolkata, West Bengal, India

***Corresponding author**
Dr. Pradipprava Paria
Email: drpradip83@gmail.com

Abstract: Cutis verticis gyrata (CVG) is hypertrophy and folding of the skin of the scalp presenting as gyrate or cerebriform pattern. It may be primary (essential or non-essential) or secondary to a number of diseases. Chromosomal anomalies also may be seen. Treatment is symptomatic. Sometimes, surgical repair needed. We here present a case of cutis verticis gyrata in a newborn with Turner syndrome.
Keywords: Cutis Verticis Gyrata, scalp dermatoses, Turner syndrome

INTRODUCTION:

CVG is a descriptive term where folds & deep furrows formed from thickened skin of the scalp which resembles the surface of the brain [1]. It was first cited by Alibert in 1837. In 1953, Polan & Butterworth classified CVG into primary & secondary form which was later modified by Garden & Robinson into primary essential CVG for cases in which no other abnormality was found, primary nonessential CVG in association with neuropsychiatric or opthalmologic abnormalities & secondary CVG occurring due to a number of localized or systemic inflammatory or neoplastic diseases [2]. Chromose abnormalities have been observed in some cases.

In this case report we describe the rare case of a female neonate with Turner syndrome having secondary CVG and its diagnostic & therapeutic aspects.

CASE REPORT

A 3 day old female baby was referred to our hospital for evaluation of congenital anomalies. She was born to 28 year old multigravida mother who had uneventful antenatal history. The infants parents were non consanguinous. Birth history detailed her 37 weeks gestation with 3.3 kg birth weight. She was born by normal vaginal delivery. Physical examination revealed 7 x 6.5 cm symmetric skin fold in anterior posterior direction with relative alopecia in central and parieto-occipital region of scalp which could not be corrected by pressure or traction. No local inflammatory signs or hyperpigmentation could be found (fig 1). Head circumference was 35 cm with normal sutural opening. Other findings were low set ear, hypertelorism, low hair line and extra skin fold at nape of the neck. There was wide spaced nipple with bilateral congenital talipes equinoverus(CTEV) and lymhedema of both feets (fig 2 & 3) . Systemic examination was nomal except a soft systolic murmur over precordium, subsequently diagnosed as PDA by echocardiography. Abdominal and cranial utrrasonography did not reveal any abnormality. Occular examination was also normal. According to her phenotypic characteristics we send blood for chromosomal analysis and its revealed 45 XO, suggestive of Turner syndrome. Other laboratory work up including complete haemogram, biochemistry panel including alkaline phosphatase, BUN, free T4, TSH all were within normal limits. Cranial CT scan revealed normal cranial contents with thickening of the skin and subcutaneous fat. Skin biopsy could not be performed as her parents did not give consent due to religious constrain.

She was diagnosed to have secondary form of CVG .The benign process of the disease was explained to the parents & she was discharged & advised for follow up in our clinic mainly due to its' syndromic association.

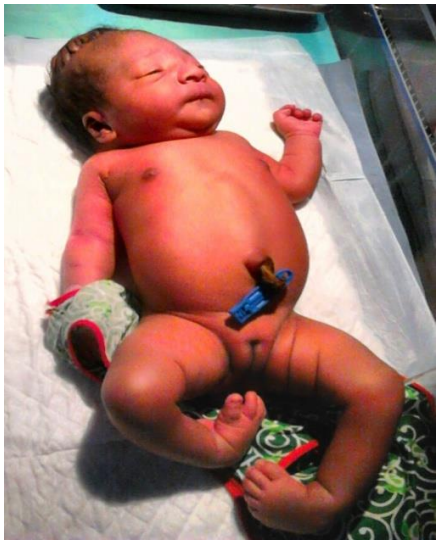

Fig-1: showing girl neonate with low set ear, hypertelorism, wide spaced nipple, bilateral CTEV with congenital lymphedema

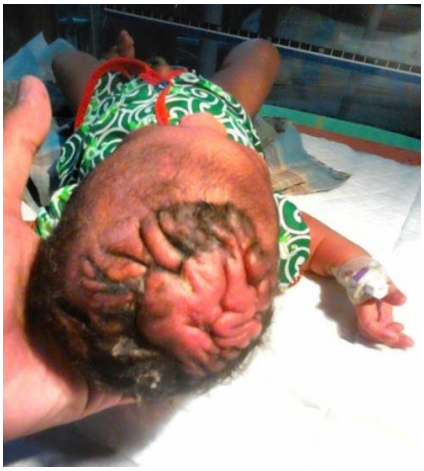

Fig-2: showing folds and deep furrows in parieto-occipital region of scalp. There is relative alopecia over the folds and normal hair growth in furrows

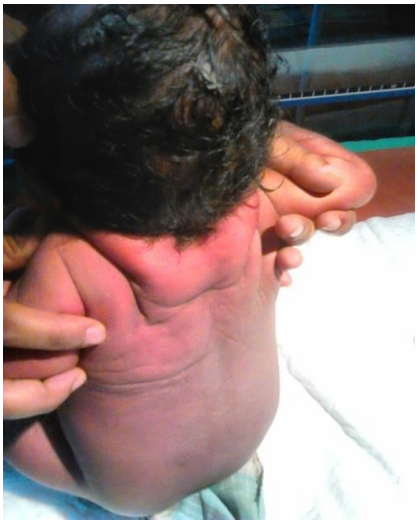

Fig-3: Showing abundant skin in nape of the neck and low posterior hair line

DISCUSSION

Prevalence of 1 in 100000 in men and 0.26 per lakhs in women. Other terms include pachydermie vorticelle, cutis sulcata, bulldog scalp & cutis capitus strata [3]. The etiology of this condition is varied and 2 groups have been described, with primary and secondary variants. The primary form can be divided in essential and non-essential form [2].

In primary essential CVG, usually, the clinical picture starts during or after puberty, occurring especially after 30 years of age. No other associated anomalies are found and histological findings of biopsied skin shows hypertrophy of sebaceous structure without any evidence of collagen thickening [4]. Most cases of primary essential CVG seems to be sporadic, although autosomal recessive & dominant inheritance with variable expression have been described. Primary non essential CVG occurs often with neuropsychiatric conditions such as seizure disorder & mental retardations. IQ is rarely above 35. There is also increased frequency of ocular abnormalities in primary non essential form. Secondary CVG occurs as a result of systemic illness or local process of the scalp such as eczema, folliculitis, pemphigus, local neoplasms including congenital cerebriform intradermal nevus, neurofibroma or hamartoma. It may also be associated with endocrine disorders such as acromegaly, cretinism & insulin resistance and various other conditions like idiopathic hypertrophic osteopathy, amyloidosis, syphilis, leukemia, acanthosis nigricans, tuberous sclerosis, neurofibromatosis, Ehlers-Danlos syndrome, and trauma [5, 6].

Neonatal CVG has been reported in Turner & Noonan Syndrome in context of congenital lymphedema, where resolution of lymphedema leaves redundant skin [7]. CVG also occurs in genetically inherited condition such as pachydermo periosteosis.

Althuogh CVG is disfiguring it has benign prognosis except secondary CVG with cerebriform inradernal nevus which can degenerate into malignant melanoma & should be managed with complete excision. In majority of cases treatment is symptomatic. Patient should be educated in scalp hygiene to avoid accumulation of skin debris & secretions in the furrows causing odour & itching [8]. Plastic surgery may be performed for esthetic purpose. Surgical techniques available are simple resection of the lesion, placement of skin expanders or skin grafting [9]. In cases of total resection of the lesion, latissimus dorsi or myocutaneous flap is used for covering the affected area.

CONCLUSION:

Through this case report we propose that CVG may be considered in newborn though it is a very rare entity. And congenital CVG must be evaluated by chromosomal analysis for its syndromic association.

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