PRIMARY NON-ESSENTIAL CUTIS VERTICIS GYRATA: REPORT OF A CASE

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ABSTRACT

Cutis verticis gyrata (CVG) is a rare transformation of the sca characterized y ridges and furro s resem ing surface of the rain It has rimary and secondary ty es Patients ith rimary CVG have norma s in and does not have an under ying atho ogica rocess It is further su divided into non-essentia CVG and essentia CVG ased on the resence or a sence of o htha mo ogica a - norma ities or neuro sychiatric ro ems such as menta retardation, cere ra a sy, schizo hrenia or e i e sy The secondary CVG is a ays a manifestation of an under ying inf ammatory, neo astic or a systemic disorder e re ort here a case of cere ra a sy in a 16-year-o d oy ho resented ith fourteen months history of rogressive deformity of head and face and as diagnosed as rimary non-essentia CVG

Key Words: Cutis verticis gyrata, Cere ra a sy, Non-essentia cutis verticis gyrata

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INTRODUCTION

Cutis verticis gyrata (CVG) is a unique condition of sca deformation characterized y fo ding and furro - ing of sca in a attern that resem es the crests and gyri of cere ra corte It is more common in ma es com- ared to fema es ith an a ro imate reva ence of 1 in 100,000 in ma es and 0 026 in 100,000 in fema es[1] It has rimary and secondary ty es Patients ith ri- mary CVG have norma s in and is further su divided into non-essentia CVG and essentia CVGs ased on the resence or a sence of neuro sychiatric ro ems such as menta retardation, cere ra a sy (CP), schizo hrenia or e i e sy[2] In secondary CVG, the sca has atho ogic changes secondary to either oca inf ammation, neo- asia or a systemic i ness e resent here a case ho resented ith sym toms of rogressive head and face deformity and as found to have rimary non-essentia CVG

CASE REPORT

e resent here case of a 16-year-o d oy from rura Pun a ho resented ith fourteen months history of rogressive deformity of head and face According to the arents, the atient had mi d ea ness in right arm and eg since infancy ut had im roved a ot after ta - ing some her a medicines The atient had no history of fits, visua ro ems, menta retardation or a sychi- atric disorder There as no history of recent drug in- ta e The fami y history as ositive as the e der rother had simi ar head and facia deformities

On e amination, he had asting of right thenar mus- c es (Figure 1A) Right foot as s ight y inverted on free hanging (Figure 1B) A rominent a domina hernia and a groove over um ar s ine as evident (Figure 1C and 1) e had a norma o er e ce t for right interossei and a ductor o icis revis that had a o er of -4 The dee tendon ref e es ere norma , ho ever, the Ba ins i sign as ositive on the right side e had mi d acne over u er chest and ac (Figure 2A and 2B) On sca e amination, he had mu ti e horizonta non-re- duci e furro s over the sca (Figure 2C and 2) There ere no s in esions over the sca The chi d as as ed for a head shave to have a c earer sca vie ut he re- fused is l eva uation revea ed order ine deficiency

is com ete ood count, rena , thyroid and iver function tests ere norma The serum gro th hormone eve s ere ithin the norma range The -ray s u did not sho any a norma ity The com uterized tomogra-

Figure 1: (A) wasting of right thenar muscles, (B) slightly inverted right foot on free hanging, (C) a prominent abdominal hernia (D) a groove over lumbar spine

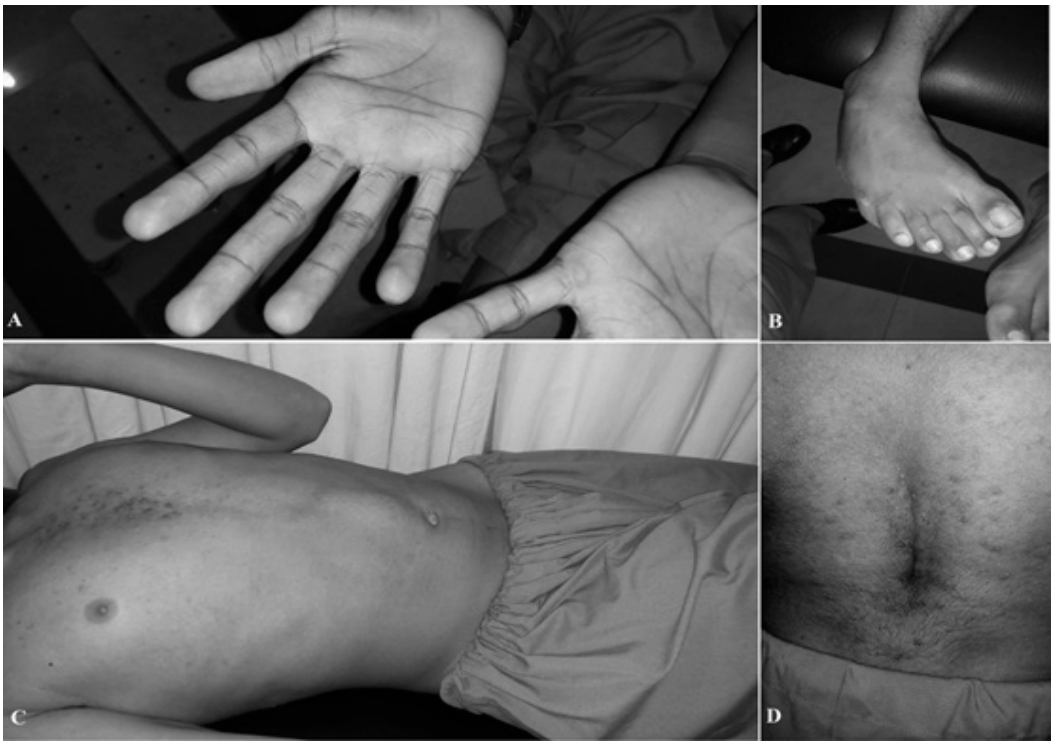
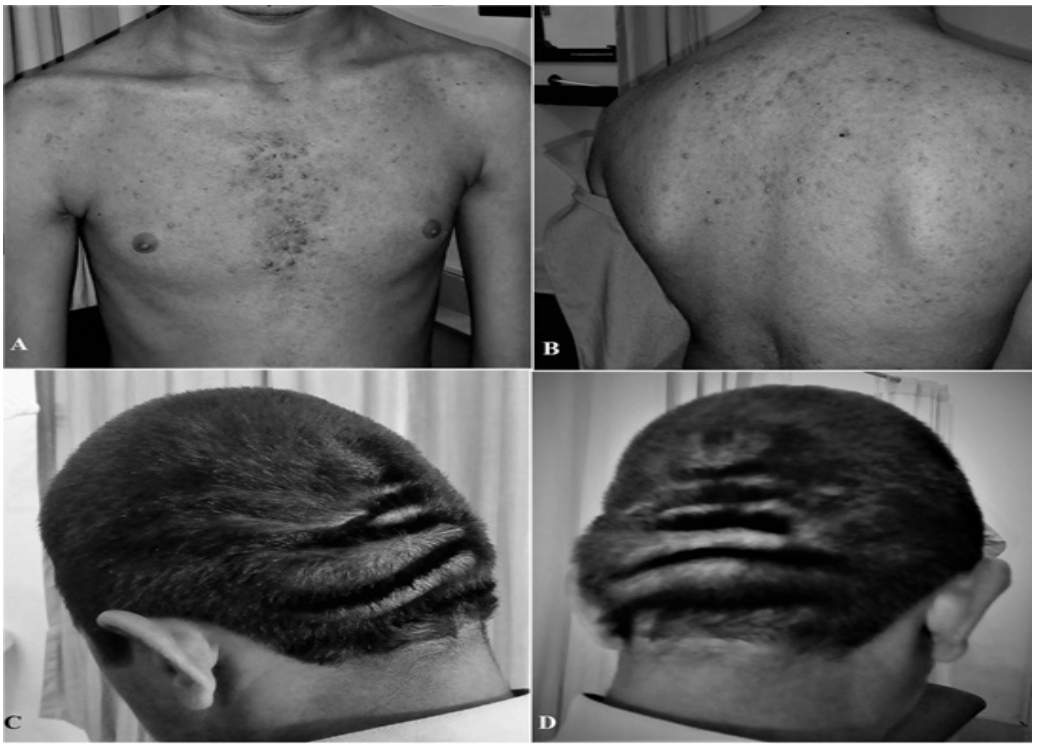


Figure 2: (A and B) mild acne over upper chest and back, (C and D) multiple horizontal non-reducible furrows over the scalp



hy (CT) scan as norma e ce t for sca convo utions The nerve conduction studies for right arm and eg ere norma e ce t for moderate median nerve com ression at right rist The sca s in io sy as refused y the atient and his arents

Thus, the atient as diagnosed as a case of rimary non-essentia CVG The ea ness in right arm and foot inversion ere ro a y sequa ae of right sided hemi- egic CP due to hich he as assumed to e suffering from non-essentia ty e of rimary CVG The atient as suggested surgica resection of the esions that his arents refused ecause of financia ro ems

DISCUSSION

Primary CVG is a rare condition, unique in a ear- ance due to rogressive fo ding and furro ing of the sca simu ating the cortica convo utions The usua age of onset of rimary CVG is after u erty and efore 30 years of age[2] Essentia rimary CVG is not associated ith other co mor id conditions and is more reva ent in men3 Most of the cases are s oradic, ut autosoma recessive and autosoma dominant inheritance have a so een seen[4,] On the other hand, the non-essen- tia form occurs in association ith various neuro ogica conditions i e microce ha y, menta retardation and e i e sy[3] or o htha mo ogica changes such as cataract, stra ismus or indness[3] Secondary CVG manifests ith an under ying atho ogy and may e ist since irth[2] The atho ogy may e the resu t of a oca inf amma- tory (eczema, soriasis, fo icu itis, im etigo, erysi e as, ato ic dermatitis and acne cong o ata)[4,6,] or a neo as- tic rocess or systemic i ness (eu emia, neurofi roma, my edema[2,4,] Secondary CVG has a so een re orted fo o ing treatments ith vemurafeni and ho e- rain radiothera y

Esta ishing the a ro riate diagnosis is aramount in management iagnosis of rimary CVG is c inica and does not require a oratory tests In secondary cas- es, the resentation and the associated disease deter- mine the a oratory tests need to e done Magnetic resonance imaging and CT scan are essentia in CVG resenting at irth or hen associated ith neuro og- ic and or o htha mo ogic a norma ities in ru ing out concomitant structura a norma ities of rain[9] It is rec- ommended to o tain a histo atho ogica ana ysis of the affected area the icture is norma in rimary ty e hereas in the secondary form it de ends on the un- der ying disease

There is on y sing e re ort of CVG revious y from Pa istan It as a 60-year-o d ady ith s in esions due to the effects of infi trating intraducta reast carcino- ma[2] Our case as the first re ort of rimary non-essen- tia CVG from Pa istan ho had associated effects of CP

Primary CVG carries a etter rognosis, a though surgica intervention is required to ha t the disease ro- gression and further fo o -u s are mandatory[10] In sec- ondary cases, the nature of under ying rocess deter- mines the treatment Management in rimary essentia CVG invo ves maintenance of s in hygiene and cosme- sis a ong ith sycho ogica interventions[11-14] Surgica resection of the esions is often requested for sycho- ogica or esthetic reasons Choices of treatment in CVG de end on size of the esion, the under ying cause and the atient s requests or references Surgica treat- ment o tions inc ude rimary re air, seria e cision s in grafting and oca f a seria tissue e ansion Patients ho refuse to undergo any surgica interven- tion need [3-6] months fo o u to assess rogression of the esions There are no s ecific medicines avai a e for the treatment of CVG

CONCLUSION

Primary non- essentia CVG is an e treme y rare con- dition The diagnosis can on y e made after e c uding secondary associations through a ro riate hysica eva uation and investigations The mainstay of treat- ment is surgery that arge y de ends on the atient s choice

REFERENCES

1 Po an S, Butter orth T Cutis verticisgyrata A re- vie ith re ort of seven ne cases Am Ment efic 19 3 :613 31

2 Rahman A, Mahmood A Cutis verticisgyrata secondary to infi trating ducta carcinoma reast Co Physicians Surg Pa 2012 22:120-2

3 iven G, Tanus T, Raimer SS Cutis Verticis Gyrata Int ermato 1991 30: 10-2

4 Larsen F, Bircha N Cutis verticisgyrata: three cases ith different aetio ogies that demonstrate the c assification system Austra as ermato 200 4 :91-4

Rad ans i N, A meida M , Pitanguy I Primary essentia cutis verticis gyrata--a case re ort P ast Reconstr Aes- thet Surg 2009 62:e430-3

6 U rich , Fran e I, Go nic Cutis verticisgyrata second- ary to acne sc eroticansca itis Eur Acad ermato Vene- reo 2004 1 :499- 02

R cz E, ornse , Csi s M, o os M, Sa acz P, r - ti S arier s disease associated ith cutis verticisgyrata, hy er ro actinaemia and de ressive disorder Actat erm Venereo 2006 6: 9-60

arding , Bar er CA, Carva a R , o cho , Cha man PB, Lacouture ME Cutis verticisgyrata in association ith vemurafeni and ho e- rain radiothera y C in Onco 2014 32:e 4-6

9 O amoto , Ito , To iguchi S, Ishi a a , Furusa a T, Sa ai MRI in essentia rimary cutis verticisgyrata Neuroradio ogy 2001 43: 41-4

10 Lame as AM, Tau P , Si ver L, resse A Congenita Primary Essentia Cutis Verticis Gyrata E asty 2016 16:ic13

11 Beier P, neser U, orch RE Surgica treatment of facia cutis verticisgyrata ith direct e cision Cutan Med Surg 200 11:4-

12 Mishra A, Tehrani , ancoc , uncan C Management of rimary cutis verticisgyrata ith tissue e ansion and hair ine o ering forehead asty P ast Reconstr Aesthet Surg 2010 63:1060-1

13 su Y , Chang Y , Su L , su YL Using nove su cision technique for the treatment of rimary essentia cutis verticisgyrata Int ermato 2009 4 :30 -9

14 hao , Li , ang , Guo , Lang Y, Peng L et a Treating cutis verticisgyrata using s in e ansion method Ce Biochem Bio hys 2012 62:3 3-6