Sultan Qaboos University Med J, November 2018, Vol. 18, Iss. 4, pp. e560–562, Epub. 28 Mar 19
Submitted 6 Jun 18
Revision Req. 10 Jul 18; Revision Recd. 31 Jul 18
Accepted 30 Aug 18
doi: 10.18295/squmj.2018.18.04.025

INTERESTING MEDICAL IMAGE

Primary Essential *Cutis Verticis Gyrata*

A scalp condition that may appear in various disorders

*Mónica García-Arpa,[1] Miguel A. Flores-Terry,[1] Lucía González-Ruiz,[1] Monserrat Franco-Muñoz,[1] Manolo Calvo-García[2]

المعلومات الأولية عن تثخن الفروة

حالة مرضية تصيب فروة الرأس تحدث في عدد من الأمراض

مونيكا قارسيا-أربا، مقيول فلوريس-تيري، لوسيا قونزلس-رويس، منصرات فرانكو-منوز، مانولو-كالفو-قارسيال

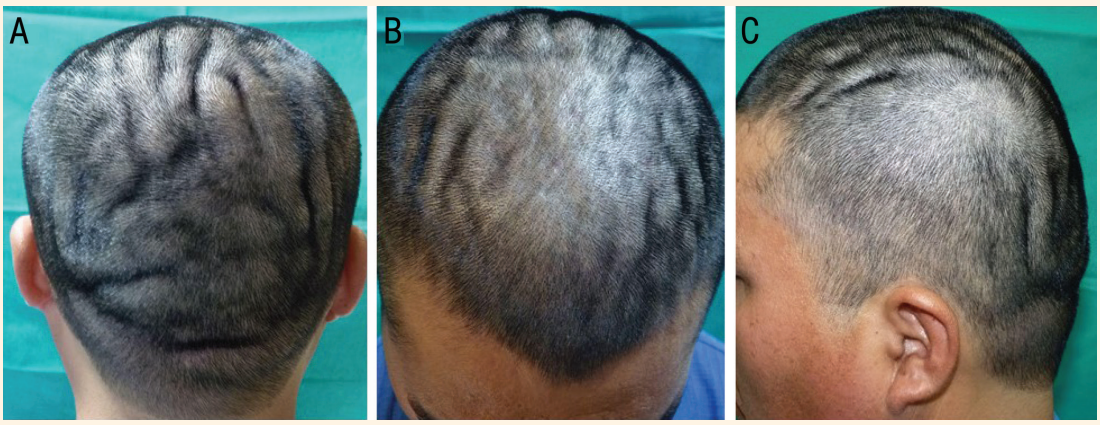

Figure 1: Photographs of the scalp of a 35-year-old male patient with intense scalp *pruritus* and nodules showing skin redundancy and deep symmetrical cerebriform folds in the frontoparietal, *vertex* and occipital areas, typical of *cutis verticis gyrata*.

A 35-year-old male patient of Bolivian origin was referred to the Dermatology Department of the Hospital General de Ciudad Real, Ciudad Real, Spain, in 2017 with intense scalp *pruritus* and nodules of one year's duration. A physical examination revealed skin redundancy with deep cerebriform folds and wrinkles in the scalp consistent with a diagnosis of *cutis verticis gyrata* (CVG) [Figure 1]. There was no reported family history of similar scalp conditions or any evidence of intellectual disabilities or neurological, psychiatric or ophthalmological manifestations. The rest of the physical examination revealed no other abnormalities, including digital clubbing.

In order to exclude secondary disorders, a cutaneous biopsy and various complementary examinations were performed. This included a complete blood count, biochemistry testing, electrophoresis and urine analysis and testing for immunoglobulins, serum thyroid-stimulating hormone, free thyroxine, growth hormone, insulin-like growth factor 1, insulin and insulin resistance index. In addition, the patient underwent serological screening for the hepatitis virus, HIV and syphilis, as well as karyotyping, hand X-raying and an ophthalmological examination. There was no evidence of any abnormalities. Magnetic resonance imaging (MRI) with volume-rendering revealed subcutaneous thickening of the scalp [Figure 2]. Intracranial abnormalities were ruled out by axial MRI [Figure 3]. Given the benign nature of the condition, the patient was discharged and localised hygiene practices were recommended.

Comment

Overall, CVG cases are classified into primary and secondary forms; primary CVG cases are sometimes further subclassified as either non-essential (i.e. those with associated neurological, psychiatric and ophthalmological manifestations) or essential (i.e. those without abnormalities or secondary disorders).[1–3] Primary essential

Departments of [1]Dermatology and [2]Radiology & Diagnostic Imaging, Hospital General Universitario de Ciudad Real, Ciudad Real, Spain
**Corresponding Author's e-mail: mgarciaa73@yahoo.es*

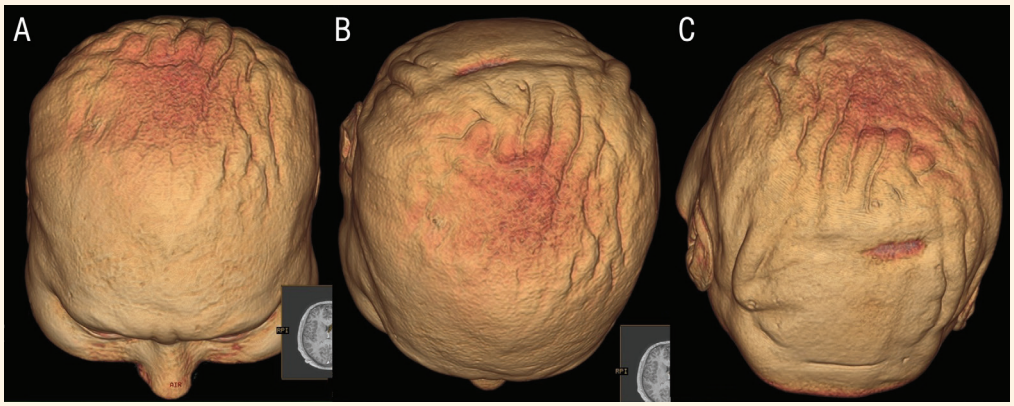

Figure 2: Volume-rendering magnetic resonance imaging of the scalp of a 35-year-old male patient showing typical features of *cutis verticis gyrata*.

CVG is a rare disorder of unknown aetiology which is five times more frequent in men.[4] Most cases are sporadic, although hereditary cases have been described.[2] The prevalence of CVG is unknown in most South American countries, including Bolivia; however, a Colombian study indicated the prevalence of CVG to be 5.9% among institutionalised patients.[5]

Primary essential CVG usually presents in puberty as symmetric scalp folds extending from the *vertex* to the *occiput* which cannot be flattened by traction or pressure.[1–3] Typically, the folds resemble the brain *cortex* and extend anteroposteriorly, although they may also occur in a horizontal direction in the occipital region. Biopsies of the folds show normal skin histopathology.[1,3] On MRI, CVG has a typical appearance due to the thickening of the skin and subcutaneous tissues, especially with three-dimensional volume rendering.[2]

Primary essential CVG must be differentiated from secondary CVG. The latter type can manifest in areas other than the scalp and has been associated with different conditions, such as acromegaly and pachydermoperiostosis.[6,7] Furthermore, CVG has been described among patients receiving external-beam whole-brain radiotherapy, vemurafenib and anabolic steroids.[8,9] Primary essential CVG may also be associated with certain genetic disorders, like fragile X syndrome.[10] Other conditions that may present with CVG include endocrine diseases (i.e. thyroid disease and diabetes mellitus), intracranial and benign scalp tumours, intracerebral aneurysms, internal malignancy, HIV-related lipodystrophy, inflammatory dermatoses and direct cutaneous infiltration in the setting of malignant haematological disease.[1,2]

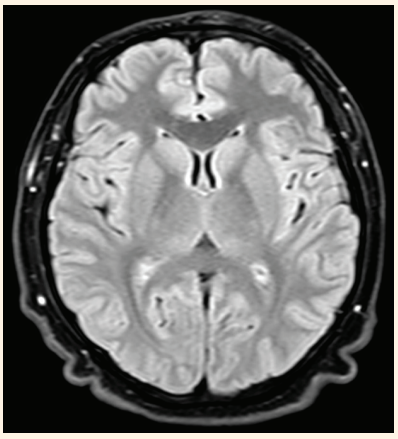

Figure 3: Axial magnetic resonance imaging scan of the brain of a 35-year-old male patient showing no evidence of intracranial abnormalities.

In the present case, the patient had a normal karyotype and did not present with any features of other disorders. Apart from the cutaneous scalp lesions, the physical examination was normal. Furthermore, the patient had no medical or family history of genetic diseases; as such, he was diagnosed with primary essential CVG. As the condition is benign, only symptomatic treatment is necessary.[2] Careful local hygiene is sometimes recommended in order to avoid secretions, unpleasant odours and *pruritus*. Surgical treatment is only necessary for aesthetic reasons and ranges from simple excision to tissue expansion and skin grafts, depending on the size and location of the lesions.[2]

References

1. Yang JJ, Sano DT, Martins SR, Tebcherani AJ, Sanchez AP. Primary essential cutis verticis gyrata: Case report. An Bras Dermatol 2014; 89:326–8. https://doi.org/10.1590/abd1806-4841.20142949.
2. Tucci A, Pezzani L, Scuvera G, Ronzoni L, Scola E, Esposito S, et al. Is cutis verticis gyrata-intellectual disability syndrome an underdiagnosed condition? A case report and review of 62 cases. Am J Med Genet A 2017; 173:638–46. https://doi.org/10.1002/ajmg.a.38054.
3. Zeljko-Penavić J, Šimić D, Jurišić K, Gunarić A. Cutis verticis gyrata: A case report. Acta Dermatovenerol Croat 2016; 24:140–3.
4. Akesson HO. Cutis verticis gyrata and mental deficiency in Sweden: II - Genetic aspects. Acta Med Scand 1965; 177:459–64.

5. Fernando Hernández J, Espejo Jiménez LM, Correa Díaz RL, de la Espriella R. Cutis verticis gyrata in adult male institutionalized population in Colombia: Case series. Acta Neurol Colomb 2015;31:404–11. https://doi.org/10.22379/2422402259.

6. Yerawar C, Bandgar T, Lila A, Shah NS. Acromegaly presenting as cutis verticis gyrata. QJM 2016; 109:423. https://doi.org/10.1093/qjmed/hcw020.

7. Sandoval AR, Flores-Robles BJ, Llanos JC, Porres S, Dardón JD, Harrison RM. Cutis verticis gyrata as a clinical manifestation of Touraine-Solente-Golé syndrome (pachydermoperiostosis). BMJ Case Rep 2013; 2013:bcr2013010047. https://doi.org/10.1136/bcr-2013-010047.

8. Harding JJ, Barker CA, Carvajal RD, Wolchok JD, Chapman PB, Lacouture ME. Cutis verticis gyrata in association with vemurafenib and whole-brain radiotherapy. J Clin Oncol 2014; 32:e54–6. https://doi.org/10.1200/JCO.2013.49.3528.

9. Sommer A, Gambichler T, Altmeyer P, Kreuter A. A case of cutis verticis gyrata, induced by misuse of anabolic substances? Clin Exp Dermatol 2005; 31:129–156. https://doi.org/10.1111/j.1365-2230.2005.01911.x.

10. Schepis C, Palazzo R, Cannavo SP, Ragusa RM, Barletta C, Spina E. Prevalence of primary cutis verticis gyrata in a psychiatric population: Association with chromosomal fragile sites. Acta Derm Venereol 1990; 70:483–6.