

Section Editor John J. Millichap, MD

Giuliano da Paz Oliveira, MD Nara Lívia Rezende Soares, MD Ricardo Lira Araújo, MD Lívia Almeida Dutra, MD, PhD José Luiz Pedroso, MD, PhD Orlando G.P. Barsottini, MD, PhD

Correspondence to Dr. Barsottini: orlandobarsottini@gmail.com

Teaching Neuro*Images*: Clinical and neuroimaging features in Gorlin-Goltz syndrome

Figure 1 Patient's face



Patient with Gorlin-Goltz syndrome who presented with multiple nevi and basal cell carcinomas.

An 18-year-old man presented with neuropsychomotor development delay since birth associated with multiple nevi and basal cell carcinomas (BCCs), which appeared in the third year of life (figure 1). Family history was unremarkable. Brain CT showed tentorium and falx cerebri calcification, associated with sphenoid hypoplasia (figure 2). Brain MRI disclosed a jaw odontogenic cyst (figure 2). Gorlin-Goltz syndrome (GGS) was diagnosed based on clinical and neuroimaging features.

GGS (OMIM:109400) is an unusual autosomal dominant disorder caused by mutations in one of the 3 following genes: *PTCH1*, *PTCH2*, or *SUFU*.¹ Around 5%–10% of patients with GGS develop

medulloblastoma or meningiomas.² GGS is characterized by multiple BCC, jaw cyst, palmar and plantar pits, cerebral sickle calcification, and skeletal malformation.²

AUTHOR CONTRIBUTIONS

Dr. Oliveira: project conception, project organization, project execution, writing of the first draft, review and critique. Dr. Soares: project conception, project organization, project execution, writing of the first draft, review and critique. Dr. Araújo: project conception, project organization, project execution, review and critique. Dr. Dutra: project organization, project execution, writing of the first draft, review and critique. Dr. Pedroso: project conception, project organization, project execution, writing of the first draft, review and critique. Dr. Barsottini: project organization, project execution, writing of the first draft, review and critique.

Download teaching slides: Neurology.org

> From the Department of Neurology and Neurosurgery (G.d.P.O., L.A.D., J.L.P., O.G.P.B.), Division of General Neurology and Ataxias, Universidade Federal de São Paulo; Centro Universitário de Saúde Ciências Humanas e Tecnológicas do Piauí (N.L.R.S.); and FACID Devry (R.L.A.), Teresina, Piauí, Brazil.

> Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.



Brain CT scan shows cerebellar tent (A) and falx cerebri calcification (B). There is also lower wing hypoplasia of the sphenoid (C). Coronal T1-weighted brain MRI discloses jaw odontogenic cyst (D).

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES

- John AM, Schwartz RA. Basal cell nevus syndrome: an update on genetics and treatment. Br J Dermatol 2016;174:68–76.
- Fini G, Belli E, Mici E, et al. Nevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome): case report. G Chir 2013;34:176–179.

© 2017 American Academy of Neurology. Unauthorized reproduction of this article is prohibited.

Neurology®

Teaching NeuroImages: Clinical and neuroimaging features in Gorlin-Goltz syndrome Giuliano da Paz Oliveira, Nara Lívia Rezende Soares, Ricardo Lira Araújo, et al. *Neurology* 2017;88;e53-e54 DOI 10.1212/WNL.00000000003618

Updated Information & Services	including high resolution figures, can be found at: http://www.neurology.org/content/88/7/e53.full.html
Supplementary Material	Supplementary material can be found at: http://www.neurology.org/content/suppl/2017/02/13/WNL.000000000 0003618.DC1
References	This article cites 2 articles, 0 of which you can access for free at: http://www.neurology.org/content/88/7/e53.full.html##ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Genetics http://www.neurology.org//cgi/collection/all_genetics CT http://www.neurology.org//cgi/collection/ct Mental retardation http://www.neurology.org//cgi/collection/mental_retardation MRI http://www.neurology.org//cgi/collection/mri Other neurocutaneous disorders http://www.neurology.org//cgi/collection/other_neurocutaneous_disord ers
Permissions & Licensing	Information about reproducing this article in parts (figures,tables) or in its entirety can be found online at: http://www.neurology.org/misc/about.xhtml#permissions
Reprints	Information about ordering reprints can be found online: http://www.neurology.org/misc/addir.xhtml#reprintsus

This information is current as of February 13, 2017

Neurology [®] is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright [©] 2017 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

