

Cutis Verticis Gyrata

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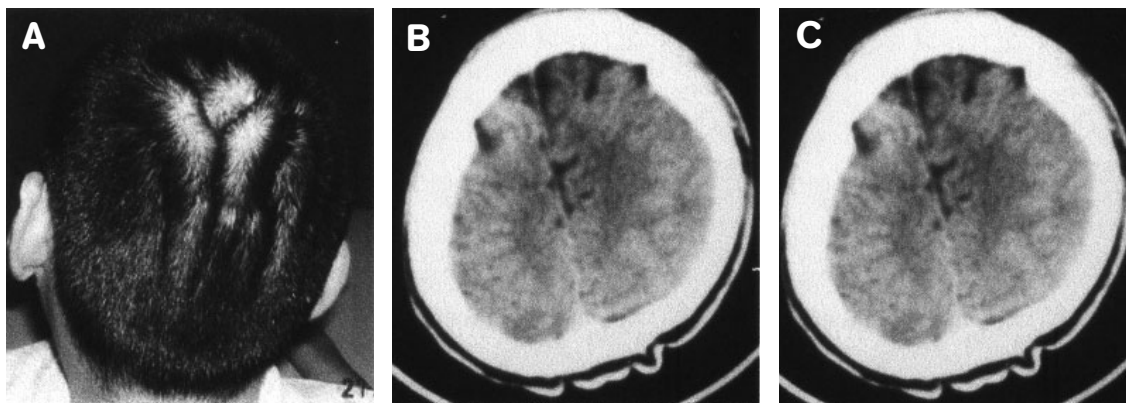


Figure. Appearance of the scalp and computed tomography of the head. Folds and furrows over vertex and upper occipital regions of the scalp (A). CT of head shows abnormal folding of scalp in the posterior parietal areas with intact underlying skull and brain structures (B-C).

A 34-year-old man was found to have several soft skin folds on the scalp during a routine health examination. Because of the difficulty during delivery, the patient was born with a vacuum extractor at full term. Psychomotor delay was subsequently noted. He was diagnosed to have quadriplegic cerebral palsy and mental deficiency in early childhood. The folds on the scalp developed gradually in the second decade and became stable thereafter. There is no history of seizure. The physical examination of the patient at 34 years of age revealed the following information. His body weight was 35.6 kg, with a body height of 151 cm, and a head circumference of 49 cm. Soft skin folds and furrows running in an anteroposterior direction were present in the vertex and upper occipital regions. Bilateral divergent, alternative strabismus was noted. He was severely dysarthric with incomprehensible speech, but his verbal

comprehension was relatively preserved. He was cooperative and could follow simple verbal commands and understood the concepts of size and color. Athetosis and spasticity were both present in his upper extremities. His legs have had been in flexion contractures for many years. Routine blood and urine tests were normal. Prolactin, thyroid hormone (T4), thyroid stimulation hormone, electrolytes, and blood sugar (fasting and postprandial) were normal. A computed tomographic (CT) scan of the head confirmed the abnormalities in the scalp tissues. Focal hypodensity in bilateral frontal parasagittal regions with widening of cortical sulci over corresponding anterior frontal areas were found, presumably due to old insult.

Cutis verticis gyrata (CVG) is a rare condition characterized by the formation of furrows and folds on the scalp^(1,2). This neurocutaneous disorder is rarely men-

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tioned in the literature of neurology, but is often recognized by dermatologist. The CVG is classified into two major categories. Primary CVG has no specific recognized cause and is frequently associated with neurological manifestations; secondary CVG is due to infiltration or inflammation of the scalp and may be associated with endocrine or neoplastic diseases⁽¹⁾. For about 20% of patients with primary CVG, there are no other abnormalities (also called essential primary CVG) whereas the majority are associated with neurological manifestations, such as epilepsy, cerebral palsy, mental subnormality, microcephaly, hereditary neuralgic amyotrophy, or ocular abnormalities⁽¹⁾. Although CT scan of the brain shows normal findings in a few cases, cerebral dysplasia and brainstem-cerebellar atrophy in patient with primary CVG were reported⁽²⁻³⁾.

Infrequent recognition of CVG may be due to subtle and gradually development of the scalp abnormality in a static medical condition and the difficulty of detection in patients with long hair. Although the etiology of the scalp abnormality is unclear, patients may have associated neurological manifestations.

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