Subependymal Nodules

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Image Description

We present the incidental MRI findings of a then 17-year-old German girl who presented with migraine-type headache. Contrast enhanced 2D spin-echo T1-weighted multiplanar reconstructions show multiple small nodules, isointense to gray matter, along the ependymal surfaces of the pars centralis of both lateral ventricles as indicated by arrows (images from September 12th 2015 are with courtesy of the Central Department of Radiology, Peine Hospital, Peine). These nodules are heterotopia of the gray matter and consist of normal neurons and glial cells but without laminar organization (Figure 1). Characteristically, they protrude into the ventricular lumen, whereby the ventricular walls get an irregular shape. Their atypical localization is due to accumulations of neurons that have had their migration to the cerebral cortex prematurely arrested [1,2]. Our patient did not have any other CNS malformations and the heterotopia is thus referred to as isolated subependymal gray matter heterotopia. Accordingly, there is an association with an X-linked (filamin 1 gene localized to Xq28) and non-X-linked (ADP-ribosylation factor guanine exchange factor 2 gene) inheritance pattern [3]. Although it is a rare condition, subependymal gray matter heterotopia is the most common identified type of heterotopia and associated with seizures in the second decade of life, affecting females more frequently than males [2]. There is no causal therapy available and management of seizures is currently the focus of treatment.

Figure 1: Subependymal nodules.

References