Unilateral absence of cerebellar hemisphere: a case report

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Abstract We describe a 38-year-old woman with absence of right cerebellar hemisphere incidentally discovered by MR imaging. No cerebellar abnormality was detected on neurological examination. Tissue probably representing dysgenic cerebellar tissue with no corticomedullary differentiation was present, connected to the right superior cerebellar peduncle. Ipsilateral enlargement of the pons and cerebral peduncle were additional findings. Although the terms “aplasia” or “agenesis” have been used to describe this entity, intrauterine destruction is the presumed pathogenetic mechanism in our case, and therefore these terms have been avoided. Asymmetry of pons and mesencephalon may be related to compensatory reorganisation or to the impairment of sequential development of nuclei and neural tracts.

Keywords Cerebellar aplasia · Cerebellar agenesis

Introduction

Complete or subtotal absence of cerebellar hemispheres is rarely reported in the literature [1, 2, 3]. The terms “aplasia” and “agenesis” have been used to describe this entity; however, some authors point to a destructive process as a pathogenetic mechanism [1, 2]. We wish to report MR findings in a case of unilateral absence of cerebellar hemisphere, together with a brief discussion of embryological development of the cerebellum and calvaria in order to identify the possible pathogenetic mechanism.

Case report

A 38-year-old housewife was admitted to Sivas City Hospital (Turkey) for occipital headache of more than 10 years’ duration. The pain was episodic; however, the neurological examination, history and laboratory tests suggested no definitive cause, and she was then referred to Erciyes University Hospitals for MR examination. MR revealed a hypoplastic right posterior fossa with thickened calvaria. The right cerebellar hemisphere was absent, and the CSF-filled right posterior fossa was continuous with the fourth ventricle (Fig. 1a, d). Tissue probably representing dysgenic cerebellar tissue with no corticomedullary differentiation (approximately 18×15×10 mm in size) was present, connected to the right superior cerebellar peduncle (Fig. 1b). Although the superior cerebellar peduncles were symmetrical in appearance, the right middle and inferior cerebellar peduncles were small compared with the left side. Interestingly, an asymmetrical enlargement of pons and mesencephalon was noted on the same site (Fig. 1b, c). The vermis was well formed, and tentorium intact. There was no contrast enhancement in the aforementioned structures. After MR examination, the patient’s neurological examination was repeated with special emphasis on cerebellar tests but no abnormality was detected. She had no intellectual deficit, despite the presence of a depressive mood.

Discussion

The cerebellum is divided into three lobes: anterior, posterior and flocculonodular [1, 4]. The primary fissure separates the anterior lobe, the most rostral portion, from the posterior lobe. The posterolateral fissure separates the posterior lobe from the most caudally located flocculonodular lobe. Each lobe consists of specific
lobules of vermis, plus the corresponding parts of the hemispheres. Embryologically, sequential growth of cerebellar parts occurs in a certain order, forming the basis of a phylogenetic classification of the cerebellum. Initially, the flocculonodular lobule (also designated as the archicerebellum) forms. The paleocerebellum, which consists of flocculi of hemispheres plus the entire vermis, forms next, and hemispheric portions of anterior and posterior lobes, i.e., the neocerebellum, forms last. Thus, the growth of the cerebellar hemispheres lags behind that of the vermis by 30–60 days [1]. Neocerebellar aplasia or hypoplasia may occur due to primary nonformation of the cerebellar anlage early in embryogenesis. The cortical defect in neocerebellar aplasia is usually bilateral and roughly symmetrical, whereas the vermis is small and flocculi are present [1]. Our patient had one-sided absence of cerebellar hemisphere plus a vermis of normal size. Although the terms "aplasia" and "agenesis" have been used to describe this entity in the literature [1, 2, 3, 5], an intrauterine destructive event is a highly probable pathogenetic mechanism in our case. The thickened calvaria due to the small posterior fossa also support this thesis: normal embryological development of membranous skull requires distension of the underlying neural mass. Skull develops from centers in each cranial plate. As neural tissue expands, collagen bundles are drawn out from these centers in a radial fashion, leading to a uniform expansion of skull. As the