

Case Report

Agenesis of the corpus callosum and septum pellucidum together with a multiple layered duramater

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Introduction

Absence of the septum pellucidum is an unusual anomaly, which occurs in 2 to 3 individuals per 100000 people in the general population [1]. This abnormality is seen as a component of more generalized abnormalities of brain development including de Morsier syndrome, agenesis of the corpus callosum, cerebral hypoplasia, Chiari malformations, holoprosencephaly, lissencephaly and familial posencephaly [2]. We report here agenesis of the corpus callosum and septum pellucidum in a male cadaver. In addition, the endosteal layer of duramater covering the left cerebral hemisphere was formed from 5 layers. The combination of these findings has not, to our knowledge, been reported previously.

Case report

During the removal of the brain in a 55-year-old male cadaver, a multiple layered dura-mater which was covering the left cerebral hemisphere was observed. In this cadaver, the endosteal layer of dura-mater was formed from 5 layers and there was a normal meningeal layer over the left cerebral hemisphere (Fig. 1). Tissue specimens were taken for light microscopy from each of the endosteal layers of dura-mater and these samples were compared with the endosteal layer of a control cadaver. For light microscopic examination, the tissue samples were processed according to routine paraffin-embedding technique. Then, sections about 5µm in thickness were cut, they were stained with hematoxylin and eosin and photographed using a Nikon Optiphot (Japan) light microscope. In the light microscopic examination of the tissue specimens, all of these 5 layers were found to be the endosteal layer of duramater (Fig. 2) and they did not show any difference from the endosteal layer of the control

Abstract

During the removal of the brain in a 55-year-old male cadaver, a multiple layered duramater which was covering the left cerebral hemisphere was observed. In this cadaver, the endosteal layer of duramater was formed from 5 layers and there was a normal meningeal layer over the left cerebral hemisphere. Tissue specimens were taken for light microscopy from each of the endosteal layers and all of these 5 layers were found to be the endosteal layer of dura-mater. Following the mid-sagittal section of the brain, agenesis of the corpus callosum and septum pellucidum were also observed in this cadaver. In addition to all these findings, the gross anatomic appearance of the telencephalon was very abnormal, when compared with the normal brains. Agenesis of the corpus callosum and septum pellucidum are very rare abnormalities and they are usually associated with neurological disorders. However, a multiple layered duramater has not been reported previously. In conclusion, we believe that the combination of these three abnormalities is a unique entity and they might have been formed depending upon early embryological developmental abnormalities.

Key words: corpus callosum, septum pellucidum, dura-mater, variation, agenesis

cadaver. In this cadaver, the meningeal layer of left cerebral hemisphere and the endosteal and meningeal layers of the right cerebral hemisphere were normal in every aspect.

Following the mid-sagittal section of the brain, agenesis of the corpus callosum and septum pellucidum were also observed in this cadaver. In addition to all these findings, the gross anatomic appearance of the telencephalon was very abnormal, when compared with the normal brains, however holoprosencephaly was not present in our case. The frontal lobe was very large and the occipital lobe was diminished in this cadaver (Fig. 3).

The protocol for this research project has been approved by the Ethics Committee of Hacettepe University, within which the work was undertaken and it conforms to the provisions of Declaration of Helsinki in 1995 (as revised in Edinburgh 2000).

Discussion

Rakic and Yakovlev [3] studied the embryological development of septum pellucidum and corpus callosum. According to these authors, the leaves of the septum form as a result of cavitation of the medial inferior commissural plate during the formation of corpus callosum. The commissural plate is a deep midline structure at the rostral end of the neural tube and it derives from the primitive lamina terminalis. The corpus callosum, anterior commissure, hippocampal commissure develop from the commissural plate. Rakic and Yakovlev proposed that after the corpus callosum forms in a certain place, the remaining walls of the commissural plate in this area form the leaves of the septum pellucidum, while the cavitory center becomes the cavum septi pellucidi. Then, in most people, the leaves fuse starting posteriorly and progressing rostrally after birth.

The meningeal layers originate from neural crest in regions rostral to the mesencephalon. It may generally be the case that those skull bones which are formed from neural crest, e.g. the base of the skull rostral to the sella turcica, frontal, parietal and squamous temporal bones, overlie meninges which are also formed from crest cells. The meninges may be divided in development into the pachymeninx (dura-mater) and leptomeninx (arachnoid layer, subarachnoid space with arachnoid cells and fibres, and pia-mater). All meningeal layers are derived from loose mesenchyme, which surrounds the developing neural tube, termed meninx primitiva or primary meninx. There is a very close relationship, during development, between the mesenchyme from which the cranial dura-mater is formed and that which is chondrified and ossified, or ossified directly, to form the skull, and these layers are only clearly differentiated as the venous sinuses develop. The relationship between the developing skull and the underlying dura-mater continues during postnatal life while the bones of the calvaria are still growing [4].

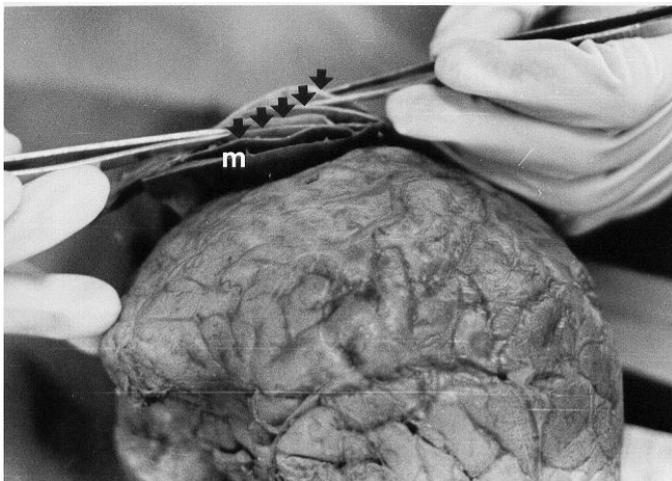


Figure 1 Photograph showing the endosteal layer of duramater formed from 5 layers (arrows) covering the left cerebral hemisphere of the cadaver. m: Meningeal layer of duramater

Agnesis of the corpus callosum and septum pellucidum are very rare anomalies found in the literature. Pineda et al. [5] reported agnesis of the corpus callosum and septum pellucidum together with hypothermia and apneic spells in two siblings. Both children died after a few months of life. In a review of more than 2000 MR images of the brain, Barkovich and Norman [6] found 2 patients with agnesis of the corpus callosum and absence of the septum pellucidum. Sener [7] described septo-optic dysplasia (de Morsier's syndrome) in a 6-months-old boy, which is associated with total callosal absence. Lahat et al. [8] described an uncommon association of infantile spasm, septo-optic dysplasia, agnesis of the splenium of corpus callosum and an interhemispheric cyst in a 2-months-old female. Partial agnesis of the corpus callosum has been found in 2 (1.4%) of 140 schizophrenic patients studied with MRI in an ongoing research program [9]. Aldur et al. [10] described an unreported anatomical variation of septum pellucidum. In this abnormality, the two laminae of the septum pellucidum were fused together and there was a hole located 1 cm anterior to its apex.

It is likely that morphological differences in brain structure are also correlated with significant underlying changes that affect function. This hypothesis is supported by the observation that hypoplasia of the corpus callosum is also associated with an increased incidence of neurological abnormalities [11-12].

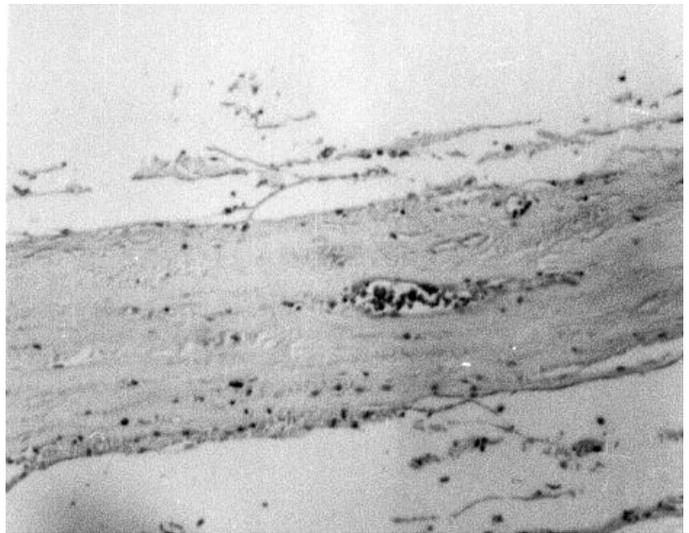


Figure 2 Photomicrograph showing the histologic appearance of one of the abnormal endosteal layers of duramater. All the other endosteal layers had the same histologic appearance (Hematoxylin and eosin, original magnification X 4).

As seen from the studies found in the literature, agnesis of the corpus callosum and septum pellucidum are very rare abnormalities and they are usually associated with neurological disorders. However, a multiple layered dura-mater has not been reported previously. In conclusion, we believe that the combination of these three abnormalities is a unique entity and they might have been formed depending upon early embryological developmental abnormalities.



Figure 3 Photograph showing the agnesis of corpus callosum and septum pellucidum. In this cadaver, the gross anatomic appearance of the telencephalon was abnormal, when compared with the normal brains.

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