Angiographic Features of Alobar Holoprosencephaly

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Summary. The purpose of this report is to present two cases of alobar holoprosencephaly and to review reports from the literature dealing especially with angiographic features of this malformation. — From the features of main cerebral arteries in the present cases, alobar holoprosencephaly may be divided into two types: the azygos artery type which has an unpaired anterior cerebral artery with bilateral middle cerebral arteries, and the undifferentiated artery type with a single undulated main trunk supplying the whole cerebrum. These two types represent the different stages of the malformation in embryonal life.

Signes angiographiques de l'holoprosencéphalie alobaire
Résumé. Les auteurs présentent deux cas de holoprosencéphalie alobaire et font une revue de la littérature traitant plus spécialement des signes angiographiques de cette malformation. Les signes angiographiques des deux cas présentés permettent de diviser l'holoprosencéphalie alobaire en deux types: le type comprenant une artère azygos avec artères cérébrales antérieure impaire et cérébrale moyenne bilatérale et le type non différencié comprenant un seul tronc principal ondulé alimentant tout le cerveau. Ces deux variétés représentent les différentes étapes de la malformation au cours de la vie embryonnaire.

Angiographische Merkmale der alobären Holoprosencephalie

Holoprosencephaly is a brain malformation in which the forebrain (prosencephalon) remains undivided at the time of differentiation from three brain vesicles into five vesicles. Experimental teratology has proved that this kind of malformation does occur between the 21st and 25th days of embryonal life.

According to Kurlander et al., (1966), there are 3 types of holoprosencephaly, alobar, semilobar and lobar. In no instance has the absence of the olfactory apparatus been noted. The first two types generally show facial anomalies such as hypotelorism and cleft palate and hare lip, which suggest the presence of intra-craniual malformation. In the lobar type, however, no such anomalies are necessarily present.

Holoprosencephalies are usually the microcephalic, while there are some which have hypertensive hydrocephalus. Recently, all suspected hypertensive hydrocephalic cases have been investigated by systemic angiography, and many kinds of cerebral malformation have been demonstrated incidentally. Although Zingesser et al., (1966) made a thorough study of the clinical and neuroradiological aspects of holoprosencephaly, the literature contains few similar observations. We feel, therefore, the presentation of an additional two cases is warranted.

Case Reports

Case 1. A Japanese male infant was examined by us when 2 days old. He had a median cleft lip and palate with associated feeding difficulties. He expired at 5 months of age with acute pneumonia.

The pregnancy had been uneventful. He was born at 38 weeks gestation, and breathed and cried immediately. His weight was 2800 g. The occipitofrontal circumference measured 29.9 cm. The anterior and posterior fontanelles were palpable but not tense.

Once a week, in hospital, he had episodes of high fever without apparent causes. Primary reflexes obtainable during the first 4 months did not disappear. Laboratory values obtained were: Na, 122 mEq/l, Cl, 94 mEq/l, and 17-OHS, 0.07 mg/day. The fundi showed severe anomalies: absence of the papilla and retina on the left side and of the retina on the right. The EEG showed burst-like episodes of high-amplitude spike and delta waves, followed by flat waves.

Left branchial angiography was performed at the age of 4 months. The external carotid artery was larger than the internal carotid in diameter. At the posterior part of the sella turcica, the internal carotid artery was markedly elongated and continued to the normal cavernous portion (Fig. 1A). The supraclinoid portion of the internal carotid is also tremendously elongated. The anterior choroidal artery is stretched backward and its distal portion shows choroidal plexus blush. The vertebral and basilar arteries seem almost normal and the posterior inferior cerebellar artery is well developed. The distal portion of the internal carotid (Fig. 1B, numbered 4—9) is hard to designate properly because of its pronounced anomaly. The posterior communicating artery is not visualized. The apex of the basilar artery is also elongated upward and backward. The initial segment of the posterior cerebral artery divides directly into fine posterior choroidal arteries. Cortical branches originating from No. 4 point of the main trunk delinate the posterior margin of the cerebrum. The avascular area behind them corresponds to the dorsal sac (Fig. 1C). At least four marginal veins drain into the superior sinus. (Fig. 1D). The thalamostriate and basilar veins can be seen. The vein of Galen is relatively depressed downward.
Fig. 1. Case 1. Alobar holoprosencephaly of a 4 month old boy. Lateral views (A—D) and AP-views (E—H, see next page) of right brachial angiography. IC = internal carotid artery, EC = external carotid artery, V = vertebral artery, B = basilar artery, ACH = anterior choroidal artery, M = middle cerebral artery probably, 1—9 = distal portion of internal carotid artery supplying the whole cerebrum. P = posterior choroidal artery. PCH = posterior choroidal artery. DC = avascular area corresponding to dorsal sac. ACV = so called DC area cerebrovasculosa. IC = internal cerebral vein. G = vein of Galen, CH = Confluence of Herophilus. The main trunk from the internal carotid artery undulates and supplies the whole cerebrum. This is the undifferentiated artery type.