Macerated Fetus *in Utero* Relating to Intracranial Angioma : Report of 2 Cases

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Abstract

Two macerated fetuses *in utero* are presented. The mothers experienced no abnormalities during pregnancy. Both fetuses were found postmortem to have intracranial angiomas. In case 1, the angioma in the choroid plexus of the right lateral ventricle was associated with intraventricular and subarchnoid hemorrhages. There was additional angiomatosis in the leptomeninges and substance of the brain. In case 2, there was leptomeningeal angiomatosis with diffuse subarchnoid hemorrhage. It is suggested that spontaneous rupture of the angioma of the choroid plexus in case 1 and of the leptomeninges in case 2 resulted in immediate death of the fetuses with subsequent maceration *in utero*. Asymptomatic angioma of the fourth ventricular choroid plexus is also described in case 2 because of its extreme rarity.

Intrapartum and neonatal morbidities have considerably decreased in recent years because of progression in obstetrics and neonatology. Stillbirth, however, has not yet been comparably reduced as revealed by its occurrence in about 1 per cent of pregnancies[1]. The etiology of fetal death, therefore, should be scrutinized not only for achieving reduction in perinatal mortality but also for counseling distressed parents who experienced fetal death in their subsequent pregnancies. Unfortunately, the definite cause of intrauterine fetal death (IUFD) still remains difficult to be evaluated. Such uncertain cause of IUFD has reported to range from 20 to 57 per cent of cases[2]. Fetal malformation was detected in 13 per cent of 253 macerated stillbirths[3]. The most common major congenital abnormalities include CNS anomalies, congenital heart defects, and renal malformations[3]. We report herein 2 cases of fetal maceration which is thought to be related to ruptured angiomas (hemangiomas, congenital vascular anomalies, or arteriovenous malformations) of the choroid plexus as well as of the leptomeninges and brain with intraventricular and subarachnoid hemorrhage. To our knowledge, fetal maceration in association with rupture of intracranial angiomas has not yet been described.

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CASE REPORT

Case 1

A 27-year-old woman who had a 38-week pregnancy with normal antenatal history presented with absence of fetal movement for 2 days. Subsequently, she developed labor pain with chocolate-colored vaginal discharge. There was no history of abdominal trauma. Aside from the absent fetal heart sound, other physical examinations disclosed no abnormalities. After 5 hours of labor pain, a macerated feus was spontaneously delivered.

An autopsy disclosed a 1,680-g macerated male fetus in which the epidermis was easily peeled off from the dermis. The placenta and the umbilical cord were normal. A 270-g soft and friable brain showed diffuse subarachnoid hemorrhage. Numerous blood vessels were conglomerated in the leptomeninges. All cerebral ventricles also contained blood, especially the right lateral one in which its choroid plexus was hemorrhagic. The choroid plexuses of the third and the fourth ventricles were not examined. There was no choroid plexus in the left lateral ventricle and it was presumably agenetic. The substance of brain was severely hyperemic but was not hemorrhagic.

Microscopically, multiple sections of the brain demonstrated many thin-walled blood vessels of various sizes in the leptomeninges, in the choroid plexus from the right lateral ventricle which was extensively hemorrhagic (Fig. 1), and in the substance of the brain to represent the angiomatosis. The pathologic impression was that of bleeding angioma of the choroid plexus of the right lateral ventricle with extension of blood into the subarachnoid space.

Case 2

A 30-year-old pregnant women regularly attended the antenatal clinic 13 times without detection of any abnormality. There was no history of trauma to the abdomen. At 41 weeks of gestation, she delivered a stillborn female weighing 3,100 g.

Postmortem examination revealed a macerated fetus in which its epidermis could be peeled off easily from the cutis. A 400-g brain, however, showed diffuse recent subarachnoid hemorrhage

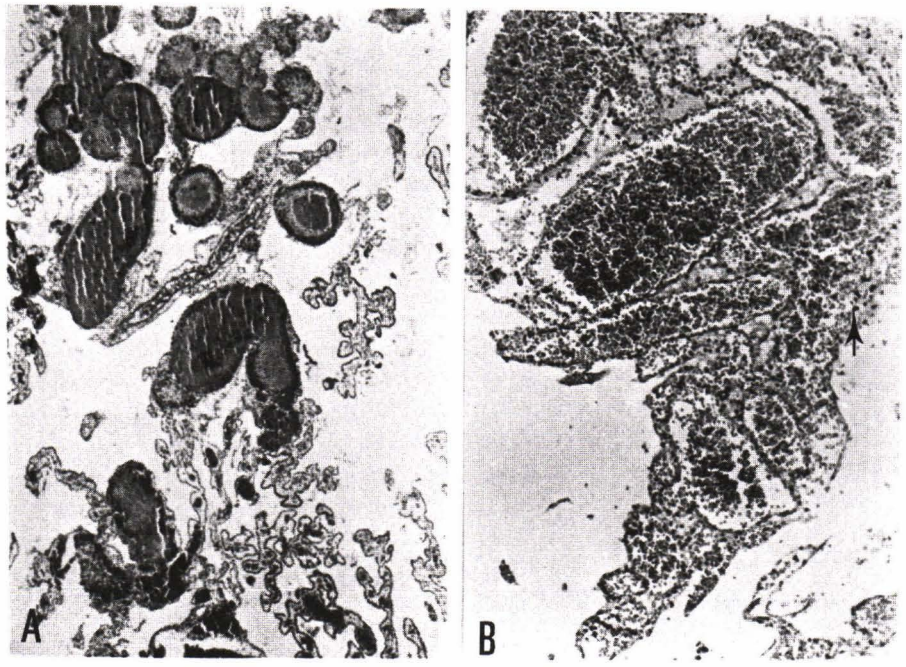

Fig. 1. (case 1). Angioma of choroid plexus from right lateral ventricle. (A). Numerous congested blood vessels form angioma in the choroidal matrix. Note many villi of the choroid plexus (Hematoxylin and eosin, x 50). (B). Same choroid plexus as in A but different region to show several thin-walled blood vessels comprising angioma in choroidal stroma. The arrow indicates area of free blood corpuscles in the latter representing hemorrhage (Hematoxylin and eosin, x 100).

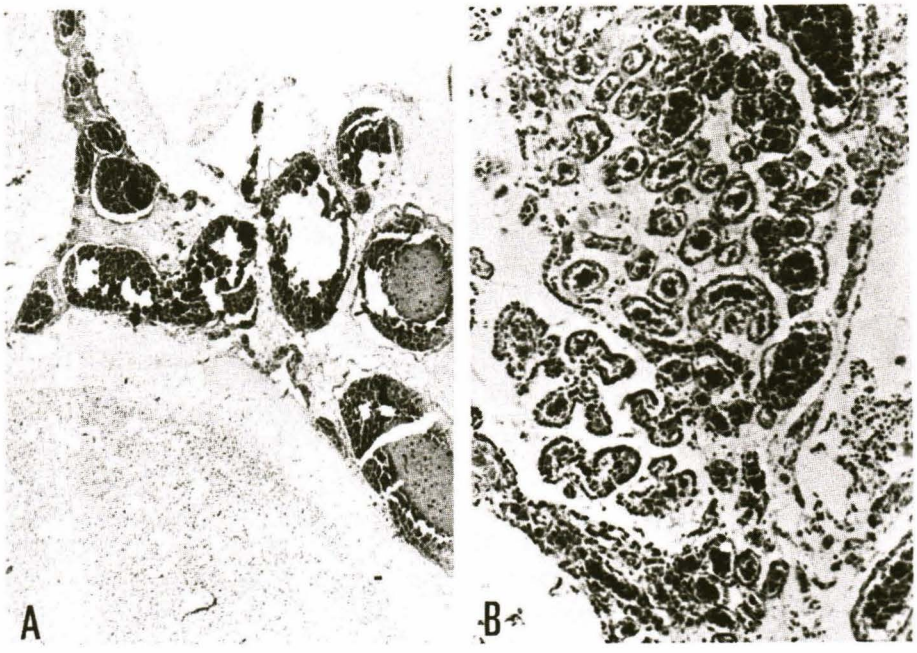

Fig. 2. (case 2). Leptomeningeal and choroidal angiomas. (A). The leptomeninges contain many blood vessels forming angioma (Hematoxylin and eosin, x 50). (B). The choroid plexus from the fourth ventricle shows stromal angioma consisting of plentiful blood vessels of various sizes (Hematoxylin and eosin, x 50).

and severely congested substance. Blood, however, was absent within various cerebral ventricles.

Microscopic studies demonstrated angiomatosis of the leptomeninges as represented by conglomeration of numerous arteries, veins, and capillaries (Fig. 2A). Similar conglomeration of small blood vessels forming angioma was also noted in the stroma of the choroid plexus of the fourth ventricle (Fig. 2B) but blood was absent in this ventricle. No angioma was observed in the brain substance. The pathologic impression, then, was that of generalized subarachnoid hemorrhage presumably from rupture of leptomeningeal angiomatosis.

DISCUSSION

To our knowledge, angioma of the choroid plexus has been reported in 96 patients[4-16]. The most common location is the lateral ventricle [10,12,15]. Most patients presented with symptoms of primary intraventricular hemorrhage *viz* headache, nausea, vomiting, meningeal signs, and varying degree of altered consciousness[15]. However, the choroidal angioma can be found as an incidental postmortem finding, as in the fourth ventricle in our case 2[5].

In case 1, we suggest that diffuse subarachnoid hemorrhage was related to rupture of the angioma of choroid plexus in the right lateral ventricle because intraventricular hemorrhage was also detected. Moreover, pathologic examination depicted hemorrhagic foci in the choroid plexus. Antepartum intracranial hemorrhage causing fetal death before labor is uncommon[17]. Only 6 per cent of stillbirths were found postmortem to have intraventricular hemorrhage[18]. To our knowledge, our case 1 is the first report of maceration in association with rupture of angioma of the choroid plexus.

In case 2, we regard rupture of the leptomeningeal angioma to have caused subarachnoid hemorrhage. Blood was not found in the ventricles. Intrauterine fetal death from rupture of the leptomeningeal angioma has been reported in nonmacerated fetus[19].

It is suggested that rupture of the intracranial angiomas in both fetuses ruptured spontaneously. Bleeding from such angiomatous rupture resulted in immediate fetal death with subsequent

maceration *in utero*. The term maceration is employed to describe autolytic alterations that ensue in dead fetuses retained *in utero*. The earliest evidence of maceration can be recognized by slipping of the skin e.g. separation of the epidermis from the dermis which was noted in both fetuses reported herein[3].

Angioma of the fourth ventricular choroid plexus is extremely rare[14]. To our knowledge, only a 54-year-old women who presented with intraventricular hemorrhage from a cryptic angioma of the choroid plexus in the fourth ventricle has been documented[14].

Definite etiology of stillbirth has been found in only 50 per cent of cases[3]. Careful postmortem examination may discover more causes. Regarding the choroidal angioma, it may be overlooked because of subtle gross appearance. Even in microscopic examination, the lesion may escape detection because it may be completely destroyed from massive outpouring of blood during the bleeding episode[15]. The choroid plexus, nevertheless, must be examined in all cases of intraventricular and even subarachnoid hemorrhages when such common cause as hypertension or aneurysm is not found[5,15]. Routine examination of the choroid plexus may increase the incidence of these developmental vascular malformations, especially as a cause of primary intraventricular hemorrhage.

SUMMARY

Two macerated fetuses were presented. Spontaneous rupture of an angioma of the choroid plexus in the right lateral ventricle (case 1) and of angioma of the leptomeninges (case 2) is suggested to have caused of immediate intrauterine death of the fetuses with subsequent maceration. Asymptomatic angioma of choroid plexus in the fourth ventricle is also described in case 2 because of its extreme rarity.

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ทารกตายเปื่อยในครรภ์ที่สัมพันธ์กับแองจิโอมาภายในโพรงกะโหลกศีรษะ : รายงานการตรวจศพ 2 ราย

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ได้รายงานทารกที่ตายในครรภ์ 2 ราย ซึ่งมารดาไม่เคยมีความผิดปรกติในระหว่างตั้งครรภ์ ตรวจศพพบมีแองจิโอมาภายในกะโหลกศีรษะทั้ง 2 ราย โดยรายที่ 1 แองจิโอมาอยู่ในคอรอยด์เพลกซัสของเวนตริเคิลข้างขวา ร่วมไปกับเลือดออกทั้งในเวนตริเคิลและช่องใต้อแรคนอยด์ รายที่ 2 มีแองจิโอมาเกิดทั่ว ๆ ไปในเลปโตเมนิงจีส์ ร่วมไปกับการตกเลือดในช่องใต้ชั้นอแรคนอยด์ ผู้รายงานเชื่อว่าการแตกของแองจิโอมาของคอรอยด์เพลกซัสและของเลปโตเมนิงจีส์ เป็นสาเหตุการตายทันทีของทารก ในรายที่ 1 และ 2 ตามลำดับ และนำไปสู่การเปื่อยของศพภายในครรภ์ของมารดาในเวลาต่อมา อนึ่งยังพบแองจิโอมาในคอรอยด์เพลกซัสของช่องที่ 4 ของสมอง ด้วย 1 ราย มีขนาดเล็กและไม่แตก

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