Giant Congenital Capillary Hemangioma of Pericranium
—Case Report—

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Abstract

The authors report a newborn male infant with a giant congenital capillary hemangioma of the pericranium. An elastic mass, measuring 6.5 × 6.9 × 3.9 cm, was located in the parieto-occipital region. Neurological examination revealed no abnormality. Angiographically, the tumor was fed symmetrically by the bilateral superficial temporal, occipital, and middle meningeal arteries. At surgery, the encapsulated tumor appeared to have arisen from the periosteum and was removed completely. Histological diagnosis was capillary hemangioma. Capillary hemangioma is a common benign tumor in infancy and usually present as a strawberry mark or port-wine stain. However, when the tumors seat relatively deeply as in the present case, they produce little or no discoloration in the overlying skin. Angiography is then useful to differentiate capillary hemangioma from other lesions and to choose an appropriate treatment.

Key words: capillary hemangioma, juvenile hemangioma, pericranium, scalp, surgical treatment

Introduction

Hemangioma is a common benign tumor in infancy and locates frequently in the skin and mucosa, especially in the head and cervical regions.1,7,6,10,19) In almost all cases, it appears as a strawberry mark or port-wine stain which is easy to diagnose,5) whereas in some cases the tumor is seated subcutaneously and is difficult to differentiate from other mass lesions.9) We describe a case of giant congenital capillary hemangioma located in the parieto-occipital region.

Case Report

A male infant was delivered at term by cesarean section with normal birth weight (2980 gm) and Apgar score 9, but a mass lesion with the size of an adult fist was noted on the parieto-occipital region. Two weeks later, he was referred to our hospital.

On admission, his head circumference was normal (36.8 cm). There was neither mass lesion, dimple nor abnormal hair in the lumbosacral region, and the anterior fontanel was soft. Neurological examination showed no abnormality. Hematological examination including coagulation system and platelet count was normal except for anemia, and echocardiography revealed no abnormal finding.

The tumor, 6.5 × 6.9 × 3.9 cm in size, was found at a little to the right side from the midline in the parieto-occipital region, and large tortuous veins were observed under the normally colored scalp (Fig. 1). The tumor size was not changed by his head position, and it was immobile with slight compressibility. Neither fluctuation nor bruit was detected.

Plain skull x-ray films at birth showed thinning of the cranial vault beneath the tumor (Fig. 2 upper), but no further destruction or enlargement of the diploe was noted at 5 weeks after birth (Fig. 2 lower). On computed tomographic (CT) scans, the tumor was isodense and enhanced homogeneously, but no abnormal finding was detected intracranially (Fig. 3). Magnetic resonance imaging revealed iso-intensity on both T1- and T2-weighted images, and dilated tumor vessels showed as a signal void phenomenon on proton images (Fig. 4). The tumor had
Fig. 1 An elastic mass, measuring 6.5 x 6.9 x 3.9 cm, is located in the parieto-occipital region with tortuous and dilated veins. The overlying scalp appears normal.

Fig. 2 upper: Plain skull x-ray film at birth showing erosive changes of the cranial vault beneath the tumor. lower: Plain skull x-ray film after 5 weeks demonstrating no further destruction or enlargement of the diploe.

Fig. 3 Pre- (left) and postcontrast (right) CT scans showing an isodense mass with marked and homogeneous enhancement but no intracranial abnormality.

Fig. 4 Proton MR images. Tumor has no connection with intracranial tissues. Tumor vessels are shown as signal void area.

no continuity with intracranial tissues, especially the superior sagittal sinus. Intra-arterial digital subtraction angiography revealed feeding from bilateral superficial temporal arteries (STAs), occipital arteries (OAs), and middle meningeal arteries (MMAs) (Fig. 5). In particular, the branches of left MMA were spreading radially into the base of the tumor through the cranium, and enhanced it markedly and heterogeneously. No feeding artery was noted from the internal carotid arteries (ICAs), but opacification of ICAs was poorer than that of the external carotid arteries. These findings suggested that cerebral blood flow was stolen by the tumor. However, normal circulation time in the tumor and the absence of early venous filling suggested the absence of arteriovenous fistulae. The draining veins were dilated scalp veins with no connection to the superior

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At 2 months after birth, the tumor was totally removed under a diagnosis of extracranial hemangioma. The skin flap was dissected easily from the tumor after ligation and resection of the feeding arteries. The tumor was covered with a fibrous capsule which was continuous with the pericranium at the base of the tumor, and adhered tightly to the cranium.

Histological examination showed vascular nests having lumina of various sizes. Tumor cells were comprised of endothelial cells with a relatively large nucleus. In some parts, there were apparent lumina, but in other parts, the lumina were filled by proliferative endothelial cells (Fig. 6 left). Silver impregnation showed irregular and annular arrangements similar to basement membrane enclosing capillary vessels (Fig. 6 right). These findings were congruous with juvenile hemangioma in the category of capillary hemangioma. Electron microscopy revealed surrounding narrow lumina, endothelial cells with a relatively large nucleus, and basement
membrane with pericytes constituting the architecture of capillary vessels (Fig. 7).

The postoperative course was favorable, and he exhibits good growth now.

Discussion

Hemangiomas are common benign tumors in infancy and childhood. Capillary hemangioma is the most common type, and the majority of them are superficial lesions found in the skin or mucosa in the oral cavity, face, scalp, and in the neck. They usually appear as strawberry marks or port-wine stains. However, giant hemangioma located subcutaneously as in the present case have rarely been reported.

In subcutaneous tumors, it is necessary to differentiate them from other mass lesions. Choux et al. studied 36 tumors of the cranial vault in children, and divided into 4 groups of bony, dural, cutaneous, and embryological origin. Six tumors were capillary hemangioma of cutaneous origin. In the other 3 groups, there were nine epidermoid cysts, six eosinophilic granulomas, and three of sinus pericranii. In the present case it was considered that the tumor had arisen from the pericranium, because it was located below the galea and the pericranium was continuous with its capsule. Ruge et al. reviewed 70 nontraumatic lumps on the head of infants and children, and observed as many as 41 dermoid tumors but only two hemangiomas.

Angiography provided valuable findings for preoperative diagnosis of the tumor nature; 1) the tumor was fed symmetrically by the bilateral STAs, OAs, and MMAs but not by the ICA, 2) the draining veins had no connection with intracranial veins such as the superior sagittal sinus, 3) poor opacification of ICAs suggested that cerebral blood flow might be stolen by the tumor, and 4) control of bleeding was considered to be most important at surgery because the MMA penetrating the cranial vault to the base of tumor would finally appear.

Capillary hemangioma on the face and scalp is frequently associated with a developmental anomaly of a major cerebral artery. Pascual-Castroviejo studied angiographically 38 child cases with hemangioma on face and scalp, and reported nine anomalies such as absence of some major cerebral artery or persistence of primitive cerebral artery. Seven of them were capillary hemangioma with an anomaly of the cerebellum or a congenital heart disease. However, the present case had no anomaly.

Giant hemangioma offers various clinical problems. The first problem is reduction of cerebral blood flow due to blood steal by the tumor, which may cause mental retardation. Other problems are congestive heart failure due to high cardiac output and thrombocytopenia known as the Kasabach-Merritt syndrome. But there is no clear definition as to what size should be called a "giant" hemangioma. Colebatch et al. described it as a tumor with a diameter of at least 5–6 cm and growing to enormous size within several months after birth, and stated that serious complications often occur in this growing period. In the present case, the observation period was relatively short after birth, but angiography showed the stealing of cerebral blood flow, although neither obvious delay of development nor heart failure and thrombocytopenia were noted. However, if the tumor is left untreated, it is quite possible that mental retardation or heart failure may occur.

Capillary hemangioma usually enlarges rapidly and achieves the largest size at about 6 months after birth, but it begins to regress gradually. It is reported that 70–95% of them spontaneously resolve after several years. Therefore, most cases are treated conservatively in expectation of spontaneous resolution. In these cases, systemic steroid therapy is used to get some regression of the hemangioma and to control complications such as thrombocytopenia. However, steroid therapy is not always effective and may induce side effects such as the Cushing syndrome, gastric ulcer, or cataract. Radiation therapy and argon laser treatment produce unsuccessful results because of scar formation.

In the present case, it was considered that early radical treatment was needed, because the abundant blood flow of the tumor posed a risk of inducing complications and it was necessary to differentiate the tumor from other mass lesions, particularly from malignant tumor. Selective embolization of the feeding arteries of this 2 months infant was too difficult. Therefore, surgical treatment was chosen for histological diagnosis and because of the resectable location of the tumor. The most important point at surgery was control of bleeding. For this purpose, it is necessary to identify the feeding arteries and draining veins accurately by preoperative angiography.

References


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