Infantile Hemangioma of the Eyelid

Report of a Case

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SUMMARY

A case of infantile hemangioma of 3 month-old female was reported. The tumor was in the subcutaneous tissue of the right upper eyelid, and small daughter nests were scattered in the surrounding connective tissue. Massive bleeding occurred during the surgical operation.

Histologically, it looked like a capillary hemangioma, but cytoplasm of proliferating cells was plump and swollen instead of flattened. In most parts, the tumor cells were arranged in solid sheets with relatively few blood vessels of capillary size. Findings by silver impregnation showed that tumor cells were derived from the endothelial cell. Histogenesis and classification of this tumor were discussed.

INTRODUCTION

Infantile hemangioma are most commonly found at birth or soon after birth. Clinical behavior of this tumor can not be predicted from its pathological features[1-14].

It is generally accepted that many of hemangiomas are congenital tumor and have hamartomatous characteristics[1-17]. This concept is also suggested by the clinicopathological features.

CASE REPORT

The patient was 3 month-old female born at full term and the delivery was normal. Her birth weight was 3,550 gm. After 2 months of normal development, a small swelling of right upper eyelid was noticed. This a swelling grew gradually and she was brought to Ehime Rosai Hospital for an examination.

Familial History: Not remarkable.

Clinical Findings:

Development was normal and no malformation was found. A small round cystic mass about 2 cm. in diameter was palpable along supermedial aspect of right upper eyelid. A sharply demarcated and movable tumor was situated in the subcutaneous tissue. Puncture of this tumor yielded fresh blood. Massive bleeding about 100 cc. occurred during the surgical operation.

Histological Findings:

Tumor was composed of lobules which were separated from each other by fibrous septa. Each consisted of varying number of small capillaries and proliferating endothelial cells in sheet-like arrangement. For the most parts, cellular component was dominant compared to the vascular channels. Cytoplasm of endothelial cells was plump and swollen, and its nucleus was large in size and round or ovoid in shape. Mitotic figures were relatively frequent.

Small tumor nests were found in the muscular layer and around peripheral nerve fibers. Silver impregnation showed that the proliferating cells were inside the reticulin sheath. Therefore, these tumor cells were probably derived from endothelial cell.

In solid tumor cell nest, small vascular channels filled with tumor cells were supported by the reticulin sheath.

DISCUSSION

In reference to the previous reports, infantile hemangioma has some characteristic features just like other vascular tumors[1-8),11-13)]. Clinically this tumor is most commonly found at birth or soon afterwards. Therefore, it is called "infantile"[2)] or "juvenile"[6)] hemangioma. In some cases, it persists to older childhood or young adulthood[2-15)]. The cause is not known in the majority of instances, but many of them are congenital[2)] and about three-fourths are present at birth.

Sex difference is not identifiable. It grows slowly and expands deeply. In some instances, it may regress spontaneously as a result of organization following thrombosis or necrosis[2),8)]. No report has been published which concerned about its systemic development like some other hemangiomas. But multicentric development of this tumor is not rarely encountered[2-5),7-12),24),25)]. Therefore, some investigators called this "benign metastasizing hemangioma"[2),6),7)] or "endothelioma-in-situ"[23].

Infantile hemangioma should be distinguished from malignant hemangioendothelioma, but angiosarcoma with low grade malignancy may be included[11),26)].

The commonest sites are the scalp, face (especially forehead and eyelid), and extremities.[2-16)]. When the tumor occurs in the orbita, it provokes various clinical symptoms by the expansion[2),4),5),7)]. It has not been reported that this tumor demonstrated some clinical complications, for example, thrombocytopenia, purpura, erythrocytopenia, and production of certain hormones. These were rarely reported in some other types of vascular tumors, such as capillary hemangioma and hemangiopericytoma [18-22)].

Gross apearance of infantile hemangioma resembles that of capillary hemangioma but much more pale and more firm. When cystic change occurs, it looks like other types of tumors derived from skin adnexae. Fibrous tumors with rich vascular component are also misdiagnosed as infantile hemangioma[7)].

Histologically, the tumor is composed of lobules of massively proliferating endothelial cells in which various numbers of small capillaries are recognized. For most parts, tumor cells are arranged in solid sheets with small amount of vascular channels[1-12)]. Cytoplasms of proliferating endothelial cells are plump and swollen, and their nuclei are large and hyperchromatic, and round or ovoid in shape. It is quite similar to that of a primitive endothelial cell except for lack of nucleolus. Mitotic figures are relatively frequent. Silver reticulin impregnation plays an important role in distinguishing this from tumors of perithelial origin. The proliferating cells of infantile hemangioma are always situated inside reticulin sheath and are never surrounded individually by reticulin fiber. As a result of this, the origin of this tumor is considered to be endothelial cell of blood vessel[2),4),11),17)]. Many designations have been given for this tumor: simple, hyperplastic, hypertrophic, juvenile, cellular, and angioblastic hemangioma [2-13)]. Domonkos called this "juvenile benign hemangioma" to distinguish it from malignant angioendothelioma[6)]. It seems to be generally accepted that a more proper term is "benign hemangioendothelioma" designed by

Table. 1. Tumors of Endothelium and Perithelium
(from "Human Pathology")

A. Benign Endothelial Tumors (vascular hamartomas)
  1. Angioma
    a. Hemangioma
      (1) Infantile hemangioma
      (2) Capillary and cavernous hemangiomas: nevus flammeus; nevus vasculosus; cavernous hemangioma; angioma racemosum; nevus araneus; angiokeratoma-angiokeratoma corpis diffusum; senile hemangioma; familial hemorrhagic teleangiectasia
    b. Lymphangioma
      (1) Capillary and cavernous lymphangiomas: lymphangioma circumscriptum; lymphangioma cavernosum; lymphangioma pyogenicum
  1. Glomus tumor
  3. Kaposi's sarcoma
  4. Diffuse angiomatosis

Table. 2. Classification of Orbital Vascular Tumors
(from "Orbital Tumors")

A. Capillary Hemangioma
  Ocular adnexal types
  Nevus flammeus (port-wine stain)
  Strawberry mark
  Sturge-Weber syndrome (encephalotrigeminal angiomatosis)
B. Cavernous Hemangioma
C. Hemangioendothelioma
  1) Benign (infantile, angioblastic, hypertrophic, hyperplastic, cellular, and juvenile hemangioma)
  2) Malignant (malignant angioendothelioma, angiosarcoma)
D. Hemangiopericytoma
  1) Benign (Pericytoma, Perithelioma)
  2) Malignant
E. Vascular Leiomyoma (venous hemangioma)
F. Vascular Malformations
  1) Orbital aneurysm (arterial)
  2) Orbital varix (venous)
  3) Arteriovenous fistula
  4) Arteriovenous aneurysm
G. Lymphangioma

Stout (1943)[1]. We employed the term "infantile hemangioma" because we believe that this designation indicates its clinical and pathological behaviors much more precisely than other terms.

Classification of vascular tumor after Morehead[5] is cited in Table 1, and that of orbital vascular tumors by Henderson is also given[2] in Table 2.

The question whether vascular tumors are true neoplasm is still unanswered. Many of them are considered to be vascular malformations or hamartomatous tumors, but some of them evidently possess true neoplastic characteristics[1-8),11-13]. Willis had discussed comprehensively on these important and difficult points[8]. Clinical aspects of vascular tumors do not always correspond with pathological findings. It is suggested that infantile hemangiomas possess these natures predominantly. Therefore, this tumor is often misdiagnosed as a malignant tumor[2),7),8)]. Management of the infantile hemangioma is a controversial problem like other hemangiomas in infancy and childhood. Among many clinicians, it is believed that no treatment is required because some of the tumors disappear spontaneously with the growth of the child. Several treatments have been proposed injection of sclerosing agents, systemic administration of corticosteroids, radiotherapy, and surgical removal[2),27),28)].

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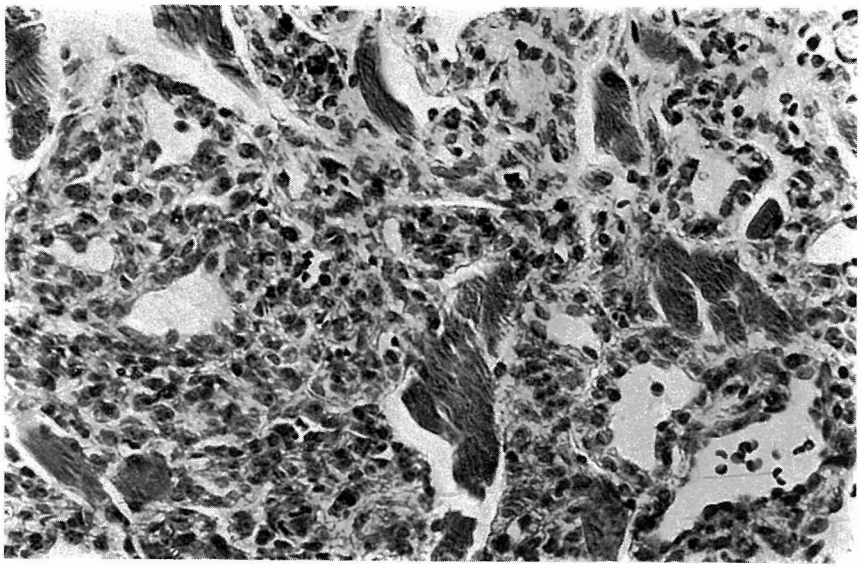

Fig. 1. Infantile hemangioma involved in the muscular fibers. (H.E. ×400)

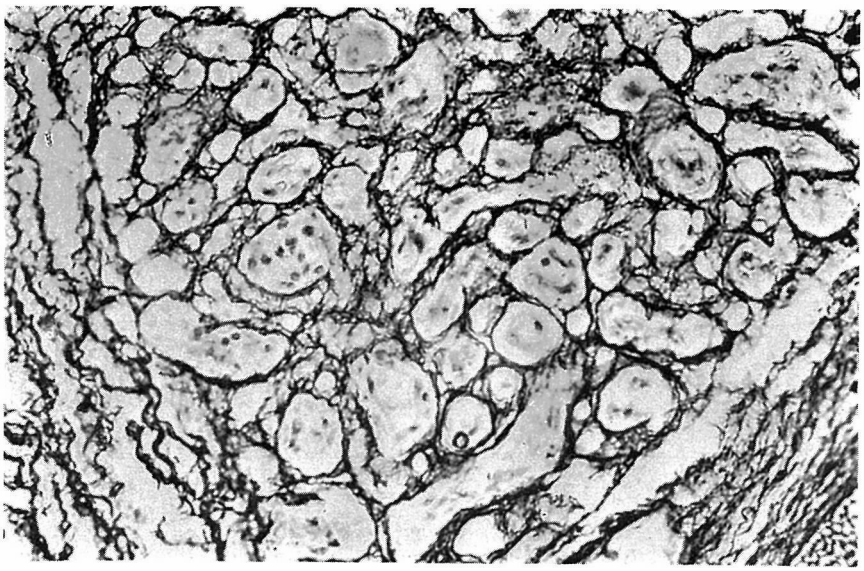

Fig. 2. Silver reticulin impregnation indicates that proliferating cells are within the capillary sheath.