

**Case study**

Multifocal kaposiform hemangioendothelioma in multiple visceral organs: an autopsy of 9-day-old female baby



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Summary Kaposiform hemangioendothelioma is a vascular tumor categorized as intermediate malignancy. We experienced an autopsy of a female baby with kaposiform hemangioendothelioma with Kasabach-Merritt syndrome. She died of systemic bleeding tendency following disseminated intravascular coagulation at the age of 9 days. At autopsy, a huge main tumor, histologically kaposiform hemangioendothelioma, was discovered in the mediastinum between the right chest cavity and pericardium. Furthermore, kaposiform hemangioendothelioma with the same histology involved the lungs, heart, liver, subserosa of cardiac part of the stomach, retroperitoneum around the right adrenal gland, broad ligament of the uterus, and muscular tissue around the thyroid. To date, a few previously reported cases of multifocal kaposiform hemangioendothelioma have demonstrated locally aggressive distributions mainly in bone and soft tissues. The present case with extensive distribution including visceral organs implies that kaposiform hemangioendothelioma may have higher potential to spread than considered before.

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1. Introduction

Kaposiform hemangioendothelioma is a rare neoplasm, in which a Kaposi sarcoma-like fascicular spindle cell growth pattern is histologically observed [1]. This tumor usually occurs in skin or deep soft tissue of infants or young children. Kaposiform hemangioendothelioma is often complicated by Kasabach-Merritt syndrome, a condition of severe thrombo-

cytopenia and consumptive coagulopathy with life-threatening hemorrhage. Kaposiform hemangioendothelioma cases complicated by Kasabach-Merritt syndrome usually have poor prognoses [2]. Kaposiform hemangioendothelioma is categorized as an intermediate (locally aggressive) tumor in the WHO classification of tumors [3].

A few cases with synchronous occurrence of multifocal kaposiform hemangioendothelioma have been reported (Table) [4–6]. However, kaposiform hemangioendothelioma cases with multiple visceral organ involvement except regional lymph nodes have not been reported yet [7,8]. We experienced a 9-day-old female baby autopsy case of multifocal kaposiform

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hemangioendothelioma. This case may be regarded as one of these multifocal kaposiform hemangioendotheliomas, but this case had more extensive distributions including multiple organs than previously reported cases in which locally aggressive distributions were described.

2. Clinical history

During the pregnancy, a female baby had ascites at the 32nd week of fetal life and pleural effusions at the 34th week; and her hydrops fetalis progressed. She was born by cesarean delivery at the 34th week and 2nd day of fetal life because of progression of systemic edema. She was born under the condition of mild neonatal asphyxia; Apgar score was 5/7. The baby was medicated under the control of artificial respiration because of her severe respiratory failure. A 3-cm tumor mass was discovered in her mediastinum by imaging study. She had no obvious congenital heart diseases. It was suggested by computed tomography angiography that the mediastinal tumor mass was a hemangioma fed by intrathoracic artery. She had severe disseminated intravascular coagulation since the beginning of her hospitalization, and she was diagnosed with Kasabach-Merritt syndrome because of the hemangioma.

She was treated with prednisolone from 3 days old and sometimes underwent blood transfusion or disseminated intravascular coagulation treatment. Her edema progressed because of increasing vascular hyperpermeability. Hemorrhage in the right chest cavity occurred at 6 days old; and therefore, she was treated with drainage and massive blood transfusion. Subsequently, a large blood clot in the right chest cavity was discovered; and severe ventilatory failure and anuria progressed. In spite of ascetic drainage and peritoneal dialysis, her edema and circulatory failure progressed; and she died at 9 days old. The autopsy was performed 3 hours after death.

3. Pathological findings

The cadaver (3.9 kg, 45 cm) showed generalized purpura and edema. Initial thoracotomy disclosed a 5 × 4 × 3-cm lobulated brownish tumor in the mediastinum between the

right lung and the heart, which adjoined the diaphragm and the pericardium. Right chest cavity contained a large-volume hematoma that pressed the right lung (Fig. 1A). Cut surface of the tumor was spongy solid and brownish to whitish (Fig. 1B). Multiple grayish-white hemorrhagic nodules were observed on visceral pleura of predominantly right but bilateral lungs (Fig. 1C). Histopathological examination revealed that the mediastinal tumor was cellular and intermingled by hemorrhage. Cellular area of this tumor was mainly composed of spindle cells growing in irregular bundle pattern, forming Kaposi sarcoma-like appearances with abundant slit-like vascular cavities (Fig. 1D). The tumor partially contained glomeruloid structures (Fig. 1E) and capillary hemangioma-like structures with clear vascular cavities (Fig. 1F). Necrotic changes were scant.

Immunohistochemistry revealed that the tumor cells showed strong positivity for endothelial markers CD34 and CD31. Many smooth muscle actin (SMA)-positive spindle cells, presumably pericytes, were present around the vascular spaces (Fig. 2A, B, C) [7]. Many slitlike microvascular cavities contained CD61-positive platelet coagulations and fibrin thrombus (Fig. 2D) [7]. The tumor was negative for human herpesvirus 8, a hallmark of Kaposi sarcoma [9]. The tumor was also negative for GLUT-1; therefore, it was unlikely to be juvenile hemangioma [10]. Thus, this tumor was pathologically diagnosed as kaposiform hemangioendothelioma.

In addition to the main tumor in the mediastinum, she had multifocal microscopic kaposiform hemangioendothelioma lesions in other organs. In bilateral lungs, up to 4-mm (mainly 1-2 mm) kaposiform hemangioendothelioma nodules scattered predominantly just below the pleura (Fig. 3A). Pleural membranes also had lymphangiectasis. Occasionally, alveoli had edema and hemorrhage. The liver contained several kaposiform hemangioendothelioma nodules beneath the serosa, up to 4 mm in diameter (Fig. 3B). Besides the tumors, hepatocytes had necrotic changes mainly around the central venous areas.

In the heart, multiple kaposiform hemangioendothelioma nodules, up to 1 mm in diameter, were identified inside the myocardium of the right atrium (Fig. 3C). The cardiac cavity contained 7 mL of hemorrhagic fluid. No congenital heart malformations were identified.

Table Multifocal kaposiform hemangioendothelioma

Year	First author	Age	Sex	Involvement	Kasabach-Merritt
1999	Gianotti [4]	1 wk	Not written	Forehead, right ear, left leg, right arm, left hand, trunk	–
2006	Deraedt [5]	3 y	M	Skin, thyroid, lymph node (neck)	+
2010	Veening [6]	1 y	M	Multiple bone lesions (right femur, left humerus), liver, right neck	+
2013	Nakaya (present case)	9 d	F	Mediastinum, lung, liver, heart (right atrium), stomach/esophagus, retroperitoneum, mesometrium, muscle	+

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