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FULL PAPER

Clinical and imaging features of Kaposiform Hemangioendothelioma

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Objective: Kaposiform hemangioendothelioma (KHE) is a unique locally aggressive vascular tumor with poor prognosis. The aim of this study is to assess the clinical and imaging features of KHE, and to compare the differences between solitary and diffusive infiltrative subtype further.

Methods: The clinical and radiological findings of a cohort of 25 cases with histologically proven KHE, between June 2011 and June 2016, were reviewed retrospectively. 7 solitary and 18 diffusive infiltrative subtypes KHE were included. The differences of clinical and imaging features between these two subtypes were compared statistically by Wilcoxon rank sum test and Fisher exact test.

Results: The median age was 4 months old. 20 cases (80%) were accompanied by Kasabach-Merritt phenomenon (KMP). Most KHE located in trunk and/or extremity. The masses showed inhomogeneous echogenicity and were rich in vascularity on ultrasound; showed isoattenuation relative to muscle on unenhanced CT, isointense ($n = 15$) or slightly hyperintense ($n = 7$) T_1 weighted imaging (T_1 WI) signal relative to muscle, mainly heterogeneous

hyperintense or slightly hyperintense with speckled hypointense ($n = 17$) T_2 WI signal (77%) relative to muscle, and notable ($n = 15$) and moderate ($n = 3$) enhancement. Feeding and draining vessels were revealed in 15 cases. Five masses with DWI showed slightly restricted diffusivity, with average apparent diffusion coefficient value of $(1.28 \pm 0.09) \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$. Necrosis and hemorrhage were also found. Compared with solitary ones, diffusive infiltrative KHE were larger, more commonly accompanied by KMP and reticular lymphedema, and more frequently located in trunk and/or extremity.

Conclusion: Five masses with DWI showed slightly restricted diffusivity. A hypervascular mass accompanied by KMP and reticular lymphedema, with speckled hypointense signal T_2 WI signal, especially in pediatric patients, is highly suggestive of the diagnosis of KHE.

Advances in knowledge: Speckled hypointense signal T_2 WI signal, and notable enhancement were unique features of KHE. KHE showed slightly restricted diffusivity on DWI, commonly accompanied by KMP and reticular lymphedema.

INTRODUCTION

Kaposiform hemangioendothelioma (KHE) is rare, and classified as a locally aggressive vascular tumor by International Society for the Study of Vascular Anomalies.¹ It was composed of spindle endothelial cells, and immunopositivity for both vascular and lymphatic endothelial markers, absence of mitosis and nuclear atypia. Numerous KHE in a broad range of locations have been reported (e.g. mediastinum, retroperitoneum, extremity, trunk, gut wall, and even long bone).²⁻⁶

KHE is composed of both vascular and lymphatic components, characterized by infiltrative spindle endothelial nodules. This vascular tumor occurs mostly in children,²⁻⁶ and exceedingly rarely in adult.^{7,8} KHE is usually

accompanied by Kasabach-Merritt phenomenon (KMP), characterized by severe thrombocytopenia (or with hypofibrinogenemia, raised D dimers), even microangiopathic anemia in the presence of a rapidly growing tumor.^{2,3} Its prognosis is usually poor due to hemorrhage and infiltration of vital organ, and the mortality rate of about 10–30%.^{2,5,9} Therefore, a timely diagnosis and treatment of KHE is crucial.

Ultrasound is the modality of choice for small and superficial vascular tumor.⁹ CT is valuable in assessing bony involvement.¹⁰ MRI has most value in evaluating the extent of vascular anomalies and differentiate between vascular tumors and vascular malformations.^{9,11}

Majority of literatures about KHE are case reports, and scarcely do they focus on the CT and MR features of this vascular tumor¹² or on the differences between different subtypes. Thus, we intend to review and characterize the clinical, ultrasound, CT and MR features of 25 children with pathology-proven KHE retrospectively, and compare the clinical and radiologic differences between different subtypes further.

METHODS AND MATERIALS

This retrospective analysis was approved by our institutional review board, and informed consent was waived. A total of 32 cases of KHE from our institution (Children's Hospital of Fudan University and Fudan University Shanghai Cancer Center) were collected, between June 2011 and June 2016. Seven were excluded for incomplete clinical date, or lacking in the radiological exam.

The demographic and clinical findings of KHE cases The clinical features of 25 cases with histologically proven KHE, were reviewed and analyzed; including gender, age at presentation, clinical history, symptoms, anatomic locations, extent, and laboratory data.

The locations of masses were divided into head and neck region, trunk, and extremity sections. The extent of masses were defined as superficial (located within deep fascia) or deep masses (invaded beyond the layer of deep fascia). The masses were divided into solitary (focal with well- or ill-defined margin) and diffusive infiltrative (ill-defined with infiltrative growth pattern) subtype.

KMP was diagnosed when the platelet count was less than $100 \times 10^9 \text{ l}^{-1}$, with or without hypofibrinogenemia (normal range 2–4 g l^{-1}) and elevated D-dimer level.

Imaging studies

15 children underwent ultrasound exam. 13 patients underwent CT scans, 7 with enhancement. 22 patients were performed MR exams, 18 with enhanced MR and 5 with diffusion weighted MR. Nine cases had both CT and MR scans.

The CT scans were performed with tube voltage of 80–120 kVp (kilovoltage), tube current of 40–50 mAs (milliampere second), and slice thickness of 5 mm. Enhanced CT was performed 50–70 s after contrast injection of OMNIPAQUE (1–2 ml per kg body weight, Beijing BeiLu Pharmaceutical Co., Ltd).

MR scans were performed with 1.5 T superconducting MR system (Signa Twin Speed Exite, GE Medical Systems, Milwaukee, WI), with 4–5 mm slice thickness and 0.5–2.0 mm gap spacing. Axial, coronal or sagittal T_1 weighted imaging (T_1 WI with or without fat saturation) was performed, and pre- and/or post-contrast medium (OMNISCAN, 0.1 mmol per kg body weight, Beijing BeiLu Pharmaceutical Co., Ltd); axial, coronal or sagittal T_2 weighted imaging with and without fat saturation, was also performed. Five patients underwent diffusion-weighted MRI (DW-MRI, b value: 0, 400 and 800 s mm^{-2}). Sedation was performed if necessary. The location, size, shape, extent, internal architecture, homogeneity, and margin of all the lesions were independently evaluated by two

radiologists with more than 5 years experiences in radiological diagnosis.

The greatest diameter was measured and documented. Homogeneous and heterogeneous were visually evaluated and documented based on the texture of the masses. The reticular lymphedema was recorded when reticular stranding in subcutaneous fat and/or deep tissue was seen. The cutaneous thickness, compared with normal skin, was recorded.

The density and signal intensity of KHE were documented as hypo-, iso- and hyper- respectively relative to muscle. The degree of enhancement was compared with those of muscle and vessels. Enhancement was defined as slight when comparable to that of muscle, intermediate (enhanced degree between muscle and vessel), and notable if it resembles that of vessels. Feeding and draining vessels were documented when flow void on T_2 WI and lined notable enhancement were revealed on enhanced CT and MR. All the images were rereviewed by two radiologists unblinded to each other's review, and recorded by consensus.

Pathological investigations

All the patients underwent surgical excision ($n = 6$) or biopsy ($n = 19$). The pathological sections underwent hematoxylin–eosin staining, with slice thickness of 4 μm . The immunohistochemical staining results of CD31, CD34, GLUT-1 (glucose transporter protein-1) and D2-40, were all documented. The light microscopy and immunohistochemical staining features were also revealed and analyzed. KHE was recorded, when masses were positive both for vascular endothelial markers CD34/CD31, and lymphatic endothelial marker D2-40, but negative for GLUT-1 staining.

Statistical analysis

The Wilcoxon rank sum test was used to compare nonparametric continuous data between solitary and diffusive infiltrative subtype KHE, and the Fisher exact test to compare proportions of these two groups. The software SPSS 17.0 (SPSS inc., Chicago, IL) was employed for statistical analysis.

RESULTS

Demographics

The age at presentation ranged from 19 days to 9 years old, and the median age was 4 months old, with the sex ratio 13:12 (female:male). 19 cases (78%) occurred in the first year of life, 21 (84%) less than 2 years old, and 23 (92%) under 5 years old.

Symptoms and anatomic locations

17 of 25 children complained of immovable enlarging mass. 10 of them accompanied by red cutaneous markings. 20 children (80%) were found with thrombocytopenia, 9 with the elevation of D-dimer level, and decrease of fibrinogen. The patients under 1 year old were complicated with thrombocytopenia. 10 of them were found with anemia, 1 with heart failure. Other cases manifested swelling of the left hand, enlargement of testicle, limit range of articular motion, and hematochezia, respectively. These symptoms lasted for several days to months. Seven masses located in the cervicofacial region, one in the left ischium, one

Figure 1. A 21-months-old male having KHE(m) in the left ischium. (a–c) Coronal T_1 WI, axial and coronal FS T_2 WI show isointense signal relative to muscle on T_1 WI, and heterogeneous hyperintense signal relative to muscle on T_2 WI with surround edema(e). FS, fat-suppressed; KHE, kaposiform hemangioendothelioma; T_1 WI, T_1 weighted imaging; T_2 WI, T_2 weighted imaging.

in the right testicle, eight in the extremity, one intra-abdomen cavity, two intra-abdomen cavity and retroperitoneum, two thoracoabdominal wall, one waist and back, and two involved both the trunk and extremity. 7 solitary and 18 diffusive infiltrate subtype KHE were included. Except for two solitary KHE were well-defined, the others were all ill-defined.

The greatest diameter ranged from 2 to 20 cm, and mean diameter was 9 cm; 19 masses involved both superficial and deep tissue, and the other masses located in deep tissue.

Imaging findings

One intratesticular solitary KHE was homogeneous, the other masses were all heterogeneous. One of the seven solitary subtype KHE located in the left ischium (Figure 1), and other five in cervicofacial regions (Figure 2). Two were well-defined, and the other five were ill-defined.

3 diffusive KHE located in intra-abdominal and/or retroperitoneal cavity (Figure 3), 13 in the trunk and/or extremity (Figure 4), and 2 in cervicofacial regions. One of diffusive infiltrative KHE was found without definite solid regions.

The masses showed inhomogeneous echogenicity, and were rich in vascularity (Table 1). The KHE parenchyma (13 cases) showed isoattenuation relative to muscle on unenhanced CT [valued at 38.9 ± 3.8 Hounsfield unit], and 7 cases with enhanced CT showed notable enhancement (147 ± 17.1 Hounsfield unit). One in the left ischium showed ill-defined osteolytic destruction, with atrophy of adjacent muscle and internal calcifications. Three cases were found with erosion or remodeling of adjacent bone.

On MR images, 15 cases showed isointense signal, 7 showed slightly hyperintense signal on T_1 WI relative to muscle (Figures 1a,

Figure 2. A 1-month-old male having well-defined solitary mass (white arrow) in the left cervical region. (a) Coronal T_1 WI showed mainly isointense signal relative to muscle, with focal hyperintense signal hemorrhage. (b) Axial T_2 WI showed heterogeneous hyperintense signal relative to muscle. (c, d) Coronal- and axial-enhanced T_1 WI showed heterogeneous notable enhancement with peripheral feeding and draining vessels (v). T_1 WI, T_1 weighted imaging; T_2 WI, T_2 weighted imaging.

2a and 3a). On T_2 WI, 16 cases showed heterogeneous hyperintense signal (Figures 1b–c and 2b) and 5 showed slightly hyperintense signal, 17 of them (77%) with speckled hypointense signal relative to muscle (Figures 3b–c and 4a). Notable ($n = 15$) (Figure 2c–d) and moderate ($n = 3$) (Figures 3d and 4b) enhancement were found. Obvious flow voids on T_2 WI, or linear enhanced vessels, representing feeding or draining vessels were revealed in 15 cases (Figure 2d).

Necrosis ($n = 4$), hemorrhage ($n = 2$) (Figure 2a) and little ascites ($n = 3$) were also detected. The cutaneous thickness compared with the normal skin, was detected in 15 cases. 13 masses in the trunk and extremity were imbedded in reticular lymphedema (reticular stranding in subcutaneous fat and deep soft tissue). Three intraabdominal and/or retroperitoneal KHE showed diffusive infiltrative mass extending into surrounding vital organ, abdominal spaces, with obscure boundary of omentum and mesentery (Figure 3).

Five patients underwent diffusion weighted MRI (Figure 4c). The masses show slightly heterogeneous hyperintense signal, and the mean apparent diffusion coefficient (ADC) value was $(1.28 \pm 0.09) \times 10^{-3} \text{ mm}^2 \text{ s}^{-1}$.

Distant metastases were not found.

Pathological findings

During excision or biopsy, several masses were found just below the skin. On light microscopy, infiltrating nodules were composed of spindle endothelial cells (Figure 5a–b). The fibrosis,

Figure 3. A 3-month-old male having diffusive infiltrative KHE(m) located in intra-abdominal and retroperitoneal space with infiltrative growth pattern (invasion of pancreas, porta of liver and spleen(s), vessels of retroperitoneum), and ascites. (a) Axial T_1 WI showed mainly slightly hyperintense signal relative to muscle. (b, c) Axial T_2 WI and FS T_2 WI showed heterogeneous slightly hyperintense signal with speckled hypointense signal relative to muscle (*white arrow*), obscure boundary of mesentery and adipose tissue. (d) Axial-enhanced T_1 WI showed heterogeneous moderate enhancement. FS, fat-suppressed; KHE, kaposiform hemangioendothelioma; T_1 WI, T_1 weighted imaging; T_2 WI, T_2 weighted imaging.

abnormal lymphatic channels, and slit-like vascular channels containing erythrocytes, microthrombi, and deposition of hemosiderin were also detected, which lead to the heterogeneity and hypervascularity of KHE. Immunohistochemical staining were positive both for vascular endothelial markers CD34 (Figure 5b)/

Figure 4. A 2-month-old male having diffusive infiltrative KHE located in left thoracic wall (*white arrow*). (a) Coronal FS T_2 WI showed mainly heterogeneous hyperintense, with speckled hypointense signal relative to muscle. (b) Axial-enhanced T_1 WI showed heterogeneous moderate enhancement. (c) DWI (800 s mm^{-2}) show heterogeneous hyperintense signal relative to muscle. FS, fat-suppressed; KHE, kaposiform hemangioendothelioma; T_1 WI, T_1 weighted imaging; T_2 WI, T_2 weighted imaging.

CD31, lymphatic endothelial marker D2-40, but negative for GLUT-1 staining.

Differences between solitary and diffusive infiltrative subtype

In comparison to solitary masses, the diffusive infiltrative ones were more commonly accompanied by KMP (Table 2), larger than the solitary counterparts ($p < 0.05$), and more commonly located in the trunk and/or extremity.

DISCUSSION

7 solitary and 18 diffusive infiltrative subtype KHE were included in this group. All the masses invaded both the superficial and deep tissues or located in deep anatomic site. The trunk and extremity were the most frequently location. Most KHE were ill-defined, and accompanied by KMP; showed mainly hyper- or isointense signal on T_1 WI, hyperintense or slightly hyperintense with speckled hypointense signal on T_2 WI (77%), and were rich in vascularity. Compared with solitary subtype KHE, the diffusive infiltrative ones were larger, more commonly accompanied by KMP and reticular lymphedema, and more commonly located in the trunk and/or extremity. The diagnosis sometimes remains challenging for solitary KHE, especially in rare site (bone or testicle and so on). The pathological results still serve as gold-standard.

KHE was called Kaposilike hemangioma, hemangioma with Kaposilike features, Kaposi-like infantile hemangioendothelioma, or congenital hemangioendothelioma previously.^{13,14} The prevalence of KHE as reported for Massachusetts, USA was about 0.91 case per 100,000 children.² KHE can occur in any site, most of them frequently affect young children.² Two KHE located in ischium and testicle respectively, which are extremely rare.^{6,15,16} Most KHE manifested as immovable enlarging mass, with/without red cutaneous markings. These manifestations were nonspecific. 20 patients were found with KMP, a common and

Table 1. Imaging findings of KHE

	Main findings
Ultrasound	Inhomogeneous echogenicity; rich in vascularity
CT	Isoattenuation (valued at 38.9 ± 3.8 HU) on unenhanced CT
	Enhanced notably (valued at 147 ± 17.1 HU)
MRI	Isointense signal ($n = 15$), slightly hyperintense signal ($n = 7$) relative to muscle on T_1 WI
	Heterogeneous hyperintense signal ($n = 16$) relative to muscle, slightly hyperintense signal ($n = 5$), 17 of them with speckled hypointense signal on T_2 WI
	Notable ($n = 15$) and moderate ($n = 3$) enhancement
	Slightly heterogeneous hyperintense signal on DWI
Other manifestations	Feeding or draining vessels, cutaneous thickness, reticular lymphedema

HU, Hounsfield Unit; KHE, kaposiform hemangioendothelioma. Cutaneous thickness was defined compared to that of normal skin.

specific manifestation, could lead to life-threatening bleeding. The percentage of KHE with KMP is higher than literatures.^{2,3,12} The greatest diameter of most masses exceeds 5 cm,¹⁷ diffusively infiltrating surrounding tissues, which poses a challenge for treatment. Actually, most masses were unresectable, and the subsequent chemotherapy was needed. "Platelet trapping" and the turbulent blood flow hypothesis were proposed to explain the phenomenon of KMP.^{14,18–20} KHE were also found without KMP.²¹

CT is useful in demonstrating the osteolytic destruction, skeletal erosions and remodeling. T_1 and T_2 MR weighted imaging aid in the detection of hemorrhage, hemosiderin, impressive flow voids and reticular lymphedema. MR facilitates in revealing a hyper-vascular mass as well as the feeding and draining vessels. Heterogeneous hyperintense or slightly hyperintense with speckled hypointense signal T_2 WI signal and notable enhancement are valuable features of KHE. The hypointense signal on T_2 WI we thought is valuable for the diagnosis of KHE and helpful in differential diagnosis, which due to the "deposition of hemosiderin and dense hyaline stromal response".¹⁴ On DWI, parenchyma shows slightly hyperintense signal and the ADC value is low, which is useful to distinguish from infantile hemangioma.

The light microscopy and immunohistochemical features of KHE are unique. Infiltrating nodules are composed of spindle vascular

Figure 5. Light microscopy findings and immunohistochemical staining of KHE. (a) A slice of HE staining show infiltrating nodules composed of spindle endothelial cells. (b) CD34 is positive in spindle endothelial cell. KHE, kaposiform hemangioendothelioma.

endothelial cells, and positive for both vascular and lymphatic biomarkers on immunohistochemical results. There are several hypotheses proposed to elucidate the coexistence of KHE and lymphatic abnormalities.^{14,22} Some thought lymphatic obstruction causing lymphangiectasia.²² Some believe the lymphangioma-like component is a part of tumor. Others believe that it is lymphatic endothelial growth factors produced by KHE that lead to the abnormal proliferation of adjacent lymphatics.¹⁴ The abnormal lymphatic component is a characteristic feature to differentiate KHE from other vascular abnormalities. D2-40 as a selective marker of lymphatic endothelium is positive in KHE, and especially valuable for differential diagnosis.^{23,24}

We speculate that the reticular lymphedema of KHE may be the lymphatic abnormalities and a part of this unique disease, which were not found in hemangioma. Infantile hemangiomas classically appear at around 6 weeks of age, show rapid growth pattern and then start to involute at the age of 9–12 months. They are soft and benign, and typically color the skin bright red. These features are in contrast to congenital hemangiomas which look almost identical both clinically and on imaging, but they are fully formed at birth. KHE is often present at birth, it can be extremely large forming a hard mass, with discoloration and involvement of the overlying skin and has generally a more aggressive appearance. It doesn't show the same rapid growth as infantile haemangiomas do and doesn't involute. The speckled hypointense signal on T_2 WI, slightly restricted diffusivity on DWI, accompanied by KMP and reticular lymphedema (lymphatic abnormalities) we thought are valuable findings to differentiate from infantile haemangioma and congenital haemangioma (RICH & NICH).

There are several limitations should be mentioned. This retrospective study is based on a small size sample, so the selection bias can't be avoided. Radiological findings compared with pathology point to point, is unable to perform. Since not all the patients underwent both CT and MR scans, the image data was uneven. The ADC value, correlated with the cell density and micro-capillary perfusion, the utility of DWI in the diagnosis of KHE should be valued further with larger patient cohort.

Table 2. Differences between solitary and diffusive infiltrative type Kaposiform Hemangioendothelioma ($p \leq 0.05$ was considered statistically significant, SPSS 17.0)

	GD (Mean $\pm$ SD)	Percentage (masses in trunk and/or extremity take)	Percentage (KMP)	Percentage (RL)
Solitary mass	4.06 $\pm$ 2.15 cm	29% (2/7)	43% (3/7)	29% (2/7)
Diffusive mass	9.42 $\pm$ 5.53 cm	89% (16/18)	94% (17/18)	87% (13/15)
p value	0.003	0.007	0.012	0.014

GD, Greatest diameter; KMP, Kasabach-Merritt phenomenon; RL, reticular lymphedema.

CONCLUSION

An immovable enlarging mass with ill-defined margin is the most common manifestation of KHE. It is accompanied by KMP, and reticular lymphedema or obscure boundary of omentum and mesentery. Five masses with DWI showed slightly restricted diffusivity.

When a hypervascular mass with ill-defined margin or infiltrative growth pattern, is accompanied by KMP and reticular lymphedema or obscure boundary of omentum and mesentery, with speckled hypointense signal T_2 WI signal, especially in children, a definite diagnosis could be reached.

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IRB STATEMENT

This retrospective analysis was approved by our institutional review board.

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