Letter to the Editor

Two cases of hemangioma with Kasabach-Merritt phenomenon diagnosed via ultrasound

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Introduction

Kasabach-Merritt phenomenon (KMP) is associated with kaposiform hemangioendothelioma and plexus hemangioma. KMP is characterized by thrombocytopenia, extremely low fibrinogen levels, and high fibrin degradation (1,2). These hemangiomas occur rarely in infants and occur more commonly in children. They are mainly located in the soft tissue surface and visible in the muscle layer and adipose tissues. Lesions are usually dark red with high skin tension and temperature and do not recede with age. When hemangiomas occur in the visceral organs (e.g., liver), hepatic hemangioendothelioma appears with hypoechoic nodules and peripheral isoecho on ultrasound, peripheral annular enhancement, and internal radial arrangement on contrast (3). Due to its rapid growth, severe thrombocytopenia and consumptive coagulation disorder, namely, KMP, may occur and even be life-threatening. In the 2 cases reported here, diagnoses were made according to the corresponding ultrasound characteristics with laboratory results, which were confirmed to be consistent with pathological results. Multimodal ultrasound can provide a comprehensive understanding of the condition and is an accurate basis for the clinical formulation of a treatment plan.

Case report

Case 1—kaposiform hemangioendothelioma

A 3-month-old infant was admitted to Gansu Provincial Maternity and Child-care Hospital and referred to several hospitals before consultation, with signs and symptoms all suggesting hemangiomas; however, no related examinations were performed. Laboratory and related tests indicated low platelet and fibrinogen levels and high fibrin degradation products {fibrinogen degradation products: 34.5 ug/mL [0–5]; Fibrinogen: 0.71 g/L [2–6]}. The patient was referred to our department for an ultrasound examination. The skin of the patient's elbow appeared to be dark red, had elevated skin tension and temperature, and was accompanied by a pulsating sensation. Ultrasound examination with a Philips EPIQ 7 ultrasound system and a 12 to 5-MHz probe showed a thickened elbow skin layer (3.2 mm), enhanced fat layer echo, echo unevenness, and a solid-mixed echo mass 65 mm × 58 mm × 52 mm in size with unclear borders in the deep muscle fiber layers (*Figure 1A*). Rich dendritic blood flow signals were found on color Doppler flow imaging (CDFI), and tortuous feeding arteries were visible around the mass communicating with the ulnar artery (*Figure 1B*). PW recorded the arterial spectrum, which showed high-speed and low-resistance types. Elastography

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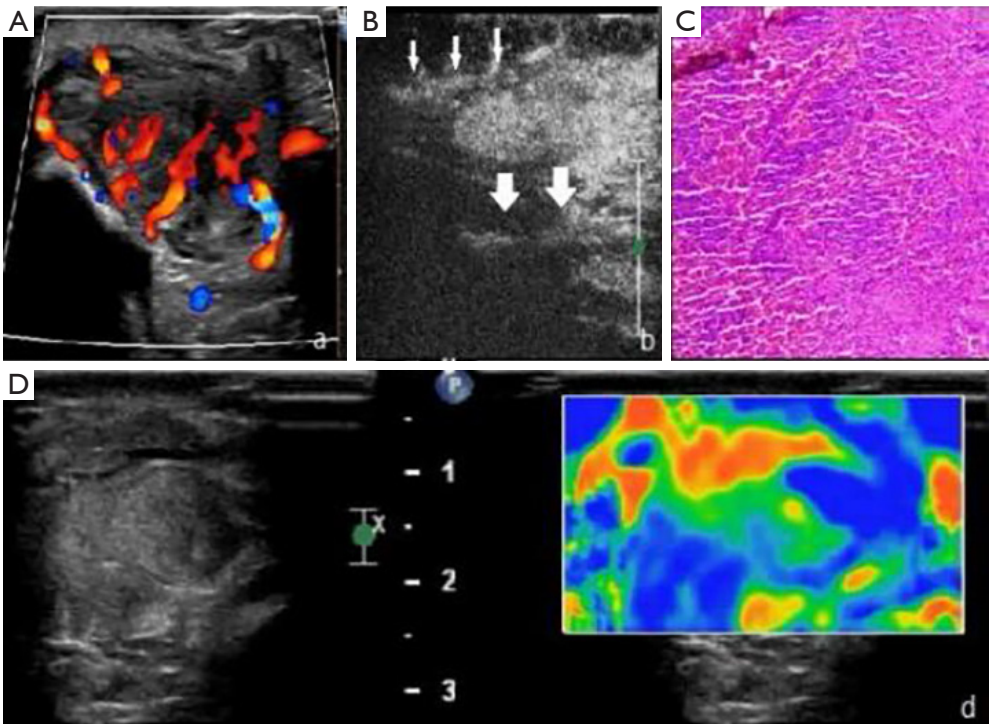

Figure 1 Ultrasound image and pathological diagram of case 1. (A) Rich dendritic blood flow signals were found on color Doppler flow imaging. (B) Contrast-enhanced ultrasound showed large feeding arteries (thick arrows) and peripheral tortuous blood vessels (thin arrows). (C) Pathology: the tumor tissue was nodular, consisting of the capillaries, epithelioid cells, and spindle cells (hematoxylin and eosin, 10×). (D) Elastography: the tissue was harder in the hyperechoic regions than that in the hypoechoic tube-like regions.

indicated that the whole mass consisted of a strong echogenic area with hardness higher than that of the low-echo and tubular structure areas (*Figure 1C*). Contrast-enhanced ultrasound showed that the size of the mass was significantly larger than that in 2-dimensional imaging; 2 large tortuous feeding arteries originating from the ulnar artery and peripheral blood vessels of the mass were also affected. Ultrasound results indicated either Kaposiform hemangioendothelioma or plexiform hemangioma. Ultrasound-guided biopsy was performed, confirming the diagnosis of Kaposiform hemangioendothelioma pathologically. Immunohistochemical results were as follows: CD34+ (vascular), CD31+, smooth muscle actin (SMA) partial+, Ki-67 proliferation index 20%, and D2-40– (*Figure 1D*). According to the clinical guidelines, oral glucocorticoids and sirolimus were recommended, and monthly follow-up blood routine reexamination was prescribed. The second blood routine examination showed that the blood platelet level had returned to normal levels. After more than 1 year of follow-up, the tumor gradually decreased.

Case 2—plexiform hemangioma:

A 6-month-old infant was admitted to the Department of Plastic Surgery of Gansu Provincial Maternity and Child-care Hospital because of a thigh lump and no obvious discomfort, but when the lump was squeezed, the patient cried. Laboratory tests showed low platelet and fibrinogen levels and high fibrin degradation products {fibrinogen degradation products: 23 ug/mL [0–5]; fibrinogen: 1.9 g/L [2–6]}. The patient was referred to our department for an ultrasound examination. The skin in the left middle thigh was slightly bluish. Ultrasound examination with a Philips EPIQ 7 ultrasound system and a 12 to 5-MHz probe showed a 2.8-mm thick left-middle thigh-skin layer and a nodule 23 mm × 18 mm × 8 mm in size with a strong-echo and uneven internal echo in the fatty tissue anterior to the muscle fiber layer (*Figure 2A*). The nodule had clear borders, and no tube-like structures were observed within it. Slightly rich dendritic blood flow signals appeared on CDFI (*Figure 2B*). A feeding artery communicated with the femoral artery through the muscular layer, and PW

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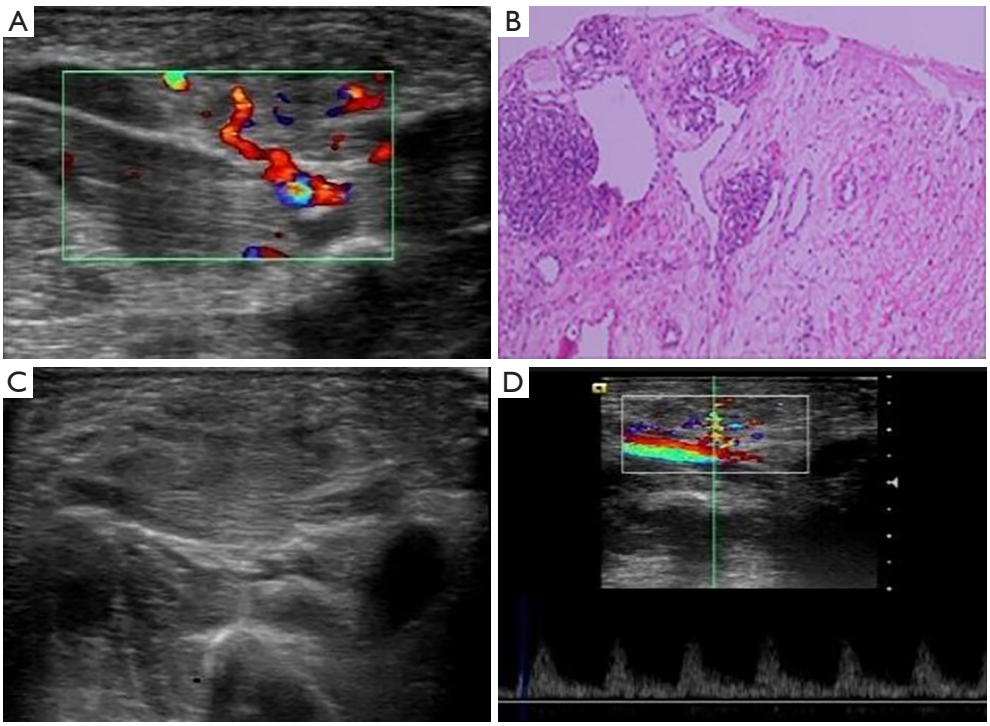

Figure 2 Ultrasound image and pathological diagram of case 2. (A) Rich dendritic blood flow signals were found on color Doppler flow imaging. (B) Pathology: fibrous connective tissue and focal epithelioid angiomatous nodules were found in the tumor tissue (hematoxylin and eosin, 10×). (C) The nodule was located in the fatty layer, and its boundary was fairly clear. (D) Spectral Doppler showed high speed and low resistance.

Table 1 Summary of manifestations of 2 cases

Case	Skin thickness (mm)	Anteroposterior diameter (mm)	Border	Blood flow	Spectrum	Feeding artery	Pathology
1	3.2	52	Unclear	Rich	High speed and low resistance	Ulnar artery	Kaposiform hemangioendothelioma
2	2.8	8	Clear	Slightly rich	High speed and low resistance	Femoral artery	Plexiform hemangioma

recorded an arterial spectrum, showing high speed and low resistance (*Figure 2C*). Ultrasound results indicated either kaposiform hemangioendothelioma or plexiform hemangioma. Based on the previous experience in diagnosing kaposiform hemangioendothelioma, ultrasound elastography was skipped and biopsy under ultrasound guidance was directly performed. The diagnosis of the plexiform hemangioma was confirmed pathologically. Immunohistochemical results were as follow: CK5/6–, CD3+, D2-40–, and P63– (*Figure 2D*). According to clinical guidelines, oral glucocorticoids and sirolimus were recommended, and a monthly follow-up blood routine was prescribed. The first blood routine reexamination showed that the blood platelet level had returned to normal levels (*Table 1*). After more than 1 year of follow-up, the tumor gradually decreased.

All procedures described in this study were performed in accordance with the ethical standards of the Ethics Committee of Gansu Provincial Maternal and Child-care Hospital (Gansu Province Central Hospital) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from patients' parents for publication

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of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

KMP was diagnosed for both cases. By combining ultrasound examination and laboratory test results, reliable diagnoses could be reached at an early stage, promoting positive effects on the patients' prognoses. KMP is a rare clinical phenomenon. It is now internationally recognized (4) that KMP specifically refers to coagulation dysfunctions associated with kaposiform hemangioendothelioma and plexiform hemangioma (5). Our plastic surgery department has treated a total of 5 children to date. KMP is caused by coagulation in and around the tumor blood vessels, resulting in severe thrombocytopenia, low fibrinogen levels, and high fibrinogen degradation products. The disease usually occurs in the trunk, limbs, and retroperitoneal areas in infants and children, occasionally originating from abdominal organs. Its onset and progression are rather rapid. Severe complications may occur and even be life-threatening without appropriate diagnosis and active treatment (6,7).

Specific ultrasound characteristics for kaposiform hemangioendothelioma and plexiform hemangioma have not been established. Therefore, they can be diagnosed with both ultrasound and laboratory test results. However, for hemangioma with KMP, a difference in the ultrasound imaging results of the 2 hemangiomas was observed. According to previous studies (8), kaposiform hemangioendotheliomas are usually characterized by uneven internal echo, unclear borders, an anteroposterior diameter of >2 cm, internal tube-like echoes, muscular layer invasions, dendritic blood flow signals usually detected on color Doppler, and high-speed low-resistance blood flow appearing on spectral Doppler. In contrast, plexiform hemangioma is a benign hemangioma with relatively uniform echo, clear boundaries, no muscle layer invasion, a thickness of <1 cm, and low or average color flow. Kaposiform hemangioendothelioma with a thickness of >2 cm is typically accompanied by KMP (9). This was applicable in case 1, with elastography showing that the tissue was softer in hypoechoic tube-like regions and harder in hyperechoic regions; contrast-enhanced ultrasound showed that the size of the kaposiform hemangioendothelioma was significantly larger than that in the 2-dimensional imaging, and the peripheral blood vessels were tortuous. Both are obvious signs of infiltration, which is consistent with the growth characteristics of kaposiform hemangioendothelioma. Additionally, hemangioma is a benign tumor caused by vasodilation or dysplasia. Ultrasonographic findings are varied, with the shape usually being round or oval. Capillary hemangioma and mixed hemangioma usually show iso-echo or low echo, while arteriovenous hemangioma shows high echo, with a periphery that is often hypoechoic, and the blood flow signal in hemangioma is usually low resistance. The internal blood flow signal of arteriovenous hemangioma is similar to that of the peripheral artery and typically has high resistance (10).

In conclusion, ultrasound can accurately locate and evaluate the size, shape, and internal characteristics of lesions, elastography and contrast-enhanced ultrasound can further evaluate tissue softness and hardness as well as specific internal blood flow signals; ultrasound-guided tissue puncture can ensure the integrity and accuracy of sampling; and the above ultrasonic characteristics, combined laboratory test results, can largely increase the accuracy of diagnosing kaposiform hemangioendothelioma and plexiform hemangioma with KMP.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-23-81/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the Ethics Committee of Gansu Provincial Maternal and Child-care Hospital (Gansu Province Central Hospital) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from patients' parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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