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CASE REPORT Open Access

Right iliopsoas kaposiform hemangioendothelioma: a case report

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Abstract

Background Kaposiform hemangioendothelioma (KHE) is a rare neoplasm of vascular origin. It may involve skin, bone, connective tissue, and parenchymal organs, and it is difficult to make a definite diagnosis in the early stage because Kasabach-Merritt phenomenon may occur in some cases.

Case presentation On June 12, 2022, a 19-month boy was admitted to our department with the chief complaint of flexion of the right hip joint for over 1 year. An exploratory laparotomy and pelvic lesion resection were conducted and the resection of the iliopsoas muscle mass was performed. The intraoperative resection of the neoplasm confirmed the pathological diagnosis of kaposiform hemangioendothelioma. The vital signs of the child were stable and recovered well after surgery, and the platelet and coagulation indices were normal.

Conclusion KHE is a rare disease in clinics, the number of confirmed cases in the world is still relatively few, and the etiology and mechanism of the disease are still unknown, which could result in misdiagnose and delayed treatment in the disease early stages. It is necessary to continue to collect relevant cases, to unify the standard treatment guidelines as soon as possible, to reduce the mortality rate, and to improve the quality of life of the cases.

Keywords Kaposiform hemangioendothelioma, Iliopsoas muscle, Connective tissue neoplasm, Case report

Background

Kaposiform hemangioendothelioma (KHE) is a rare endothelial-derived junctional angiosarcoma with potential to invade locally, which is more common in infants and children, and is rare in adults [1]. Clinical manifestations include the following: infiltrating invasion, local compression, and fatal consumptive coagulation dysfunction, Kasabach-Merritt syndrome (KMP) with thrombocytopenia [2]. Different sites of invasion result in different clinical manifestations of KHE, about 88% of KHE cases involve skin, and for the remaining 12% frequently involve retroperitoneum; it can also involve extremity's deep muscles, bones and/or joints, mediastinum, and abdominal pelvic cavity [3]. Currently, the clinical incidence rate of KHE is very low, and muscle tissue invasion is extremely rare, and relevant reports of deep muscle tissue KHE have not yet been reported in the world.

Case presentation

Chief complaint

A boy, 19 months old, was admitted to the hospital with the chief complaint of the right hip joint flexion for a year.

History of present illness

A year before admission, the boy developed right hip joint flexion which could not be straightened without obvious cause. The parents did not pay attention to it at the very beginning of the abnormality and assumed that it was caused by the right plantar hemangioma, and did not have systematic examination and treatment conducted, resulting in progressive exacerbation. Ten days prior to admission, it was found that the right hip joint flexion was further aggravated than before. The lower

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limb of the affected side was atrophied compared with the opposite, so he was sent to a hospital. The family visited our hospital for the sake of systematic diagnosis and treatment. The outpatient clinic admitted this case to our department on the diagnosis of abdominal mass.

History of past illness

The boy was born at the gestational age of 28 weeks and was admitted to our hospital with the diagnose of a preterm with very low birth weight. Systematic treatment was for 46 days and was discharged after getting better.

Physical examination

When the boy was admitted, he had lucid mind, clear speech, and no rash nor pigmentation; his pupils were equal and round, he had normal light reflex, no congestion in the pharynx, no abnormality in cardiopulmonary examination, and soft abdomen; his liver and spleen under the ribs untouched, no peristaltic wave or bowel pattern were seen, and he had normal bowel sounds. The right hip joint was flexed, could not be straightened, slight internal rotation, the leg circumference of the affected limb was smaller than the opposite side, and the muscle tone of both lower limbs was fair. About 2×2 cm of hemangioma can be seen on the right foot sole, the color faded on pressing, with no obvious pain and itching. There were no abnormalities in neurologic examination. Auxiliary examination: CT of the lower abdomen showed a right iliopsoas muscle mass, and the size was about 79×62×40 mm (A); full length bone X-ray indicated that the right femur, tibia, and fibula were thinner than the opposite, and the right lower limb was bent and rotated (B) (Fig. 1). Before the operation, blood routine examination, biochemical, and coagulation function were tested. Contraindications for operation, such as failure of systems and organs, hematological disorders, and coagulation disorders, were checked; no abnormalities were identified in the results. Preoperative assessment revealed that the abdominal neoplasm was large with speedy progress, the neoplasm boundary was not clear, and there was possibility that the neoplasm was a malignant one. It was also possible that the abundant blood vessels around the neoplasm were present and the risk of intraoperative bleeding might be high; therefore, it was necessary to prepare blood prior to surgery.

Laboratory examinations

Immunohistochemistry

The first time examination: CD31 (+), CD34 (+), Ki-67 (2%+), ERG (partial+), SMA (+), WT1 (-), VEGF (-); the second time: D2-40 (-), HHV8 (-), S-100 (-).

Pathological diagnosis: (right iliopsoas muscle) kaposiform hemangioendothelioma.

CD31, CD34, and Fli are positive, indicating that the tumor cells originate from vascular endothelial cells; SMA positivity suggests the presence of pericyte components within the tumor mass; Ki-67 positivity indicates a relatively low proliferative activity of the tumor cells.

Final diagnosis

Right iliopsoas kaposiform hemangioendothelioma.

Treatment

Thorough preoperative preparation was conducted and specific surgical approaches were planned, and surgical resection was performed. After the operation, the

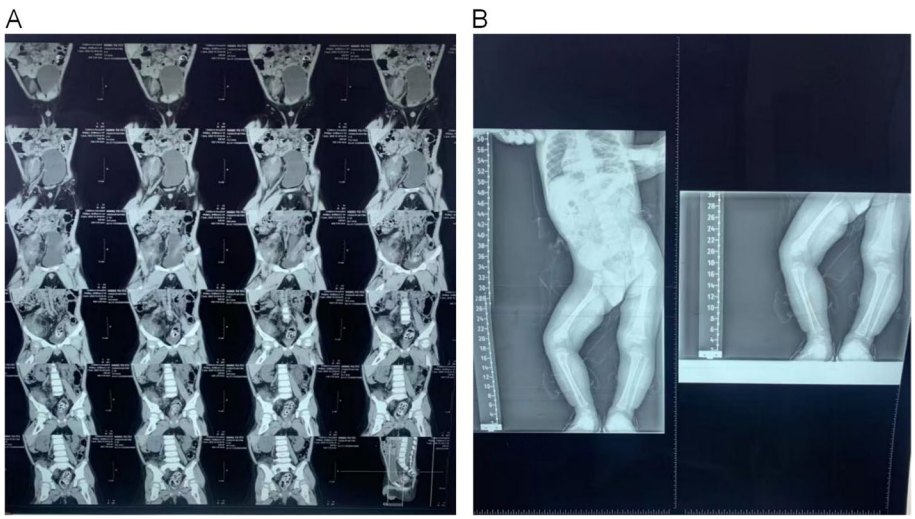

Fig. 1 CT examination and X-ray. **A** Abdominal CT: right iliopsoas muscle mass, size about 79 * 62 * 40 mm. **B** Long bone X-ray: the right femur, tibia, and fibula are thinner than the opposite side, and the right lower limb is bent and rotated

boy was transferred to the intensive care unit. There was no obvious abnormality in the coagulation function. The boy's condition improved and was successfully discharged from the hospital. Pathological examination of the neoplasm revealed a pile of gray-brown tissue, diameter 8 cm, section gray-yellow soft (C-1). Microscopic examination: (right iliopsoas muscle) vasogenic neoplastic lesions, presence of a large number of thick-walled blood vessels, infiltration of lymphocytes and plasma cell around the vessels; fiber, fat, and muscle tissue around infiltration, and interstitial mass of lymphoid and plasma cell infiltration (Fig. 2). There was formation of lymphoid vesicles, cluster proliferation of capillaries in the center of the lesion, and abundant cells and fibrous separation, showing the morphological characteristics of kaposi-type hemangioendothelioma (C-2).

Outcome and follow-up

One month after operation, there was no obvious abnormality in platelet and coagulation function, and there was no sign of recurrence according to imaging findings. The function of the affected hip is better than before. The family was advised to strengthen the rehabilitation physiotherapy after surgery and the patient was followed up regularly.

Discussion

According to relevant literature, the KHE incidence is roughly 0.7–9.1 per million cases, of which congenital onset accounts for only 7.5% [4]. The age range of onset is between 5 and 22 months [2]. In essence, it is a endothelium-derived, invasive vascular tumor with no tendency of spontaneous regression [5, 6] . It is proposed that GNA14 gene mutation resulting in the change of the mitogen-activated protein kinase (MAPK) signaling pathway may be the cause for the KHE onset [4]. The specific clinical manifestations are related to the corresponding location of tumor infiltration; the incidence of skin involvement is high with purplish red or violet tumor of hard nature, color not fading when pressed, and the demarcation from the surrounding tissues is unclear [7]. Involvement of the thoracic cavity and peritoneum can lead to pericardial effusion, pleural effusion with blood accumulation, peritoneal masses, ascites, intestinal obstruction, and jaundice; involvement of the limbs may present with characteristics of redness, swelling, heat, and pain [8]; involvement of bones and muscles may manifest as joint dysfunction or contractures, and chronic pain [7]. In the present case, the boy has a retroperitoneal iliopsoas muscle mass, which affects the joint function and causes a disorder in blood supply and malnutrition for the affected limb by compression, resulting in uneven thickness of both lower limbs. If severe thrombocytopenia, hypofibrinemia, and consumptive coagulation dysfunction occur, it is called Kasabach-Merritt phenomenon (KMP) [9]. It is reported that KMP is present in about 70% of KHE; however, the causes for the development of KMP remain unclear [3]. According to relevant reports, it may be related to the following factors: (1) abnormal proliferation of vascular endothelial cells can activate the coagulation cascade reaction and form tiny thrombus to aggravate the consumption of platelets and coagulation factors, resulting in intratumoral hemorrhage and DIC [3]. (2) Abnormal proliferation of vascular endothelial cells triggered by the special structure of the lesion causes the trapping of platelets, the exposure of the basement membrane of endothelial cells, and the formation of local turbulence of the lesion, resulting the activation, aggregation, and release of coagulation factors such as platelets and fibrin [3]. (3)

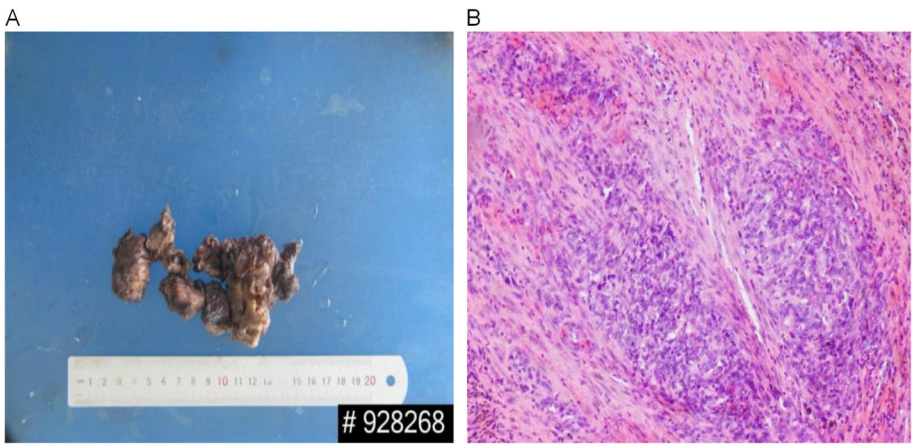

Fig. 2 Specimen of muscle mass and light microscope image. **A** Excision of part of the specimen. Gray-brown tissue, about 8 cm long, gray-yellow cut plane, and soft in quality. **B** Light microscope image. There was infiltration of lymphocytes and plasmacytes, capillaries in the center of the lesion

VEGFR3-PI3K-AKT-mTOR pathway is involved in the pathogenesis of KMP [3].

Pathological characteristics of KHE are as follows: under low magnification, KHE presents infiltrative growth with multi-nodular or lobular lesion to form hyperplasia areas and dilated lymphatic vessels around the lesion [10]. Under high-magnification microscopy, the spindle cells within the tumor are lined in bundles or vortex-like, and there are the fissure blood vessels inside [11]. Interlaced vascular structures with hemosiderin deposition or microthrombus are present. Transition with spindle cells are epithelioid tumor cells, which may have characteristic glomerular structure; it is easy to see lipofuscin deposition and vacuole formation [10]. Interstitial substances often contain collagen-like substances with vitreous denaturation [11]. If it is accompanied by KMP, a large number of red blood cells can be seen in the tumor, even extravasation [10]. Immunohistochemistry: tumor somatic cells contain spindle cells and epithelioid cells. CD31, CD34, and Fli are strongly positive, mainly showing that tumor cells are of vascular endothelial cell origin; SMA positive, indicating that the tumor contains pericellular components; and Ki67 positive, suggesting that the proliferative activity of tumor cells is low [6]. The pathological report of the present case is consistent with the pathological characteristics of KHE, and the immunohistochemical aspects are also consistent with the presentations of KHE, so the diagnosis can be definitive.

As far as KHE is concerned, the treatment strategies include surgery, arterial embolization, radiotherapy, and pharmaceutical treatments [3]. According to the relevant literature, a ladder therapy is proposed. (1) The mass is solitary or well-defined; it can be removed after vascular embolism and ligation; (2) those without surgical indications, first-line drugs such as glucocorticoids are preferred; the second-line drug therapy is administered when the first-line therapy is ineffective, such as interferon, immunosuppressant, or combined use; (3) radiotherapy can be considered when both of the above two therapies are ineffective [12]. Surgical treatment is the fastest treatment for the patients who already have symptoms of compression. Arterial embolism generally uses anhydrous alcohol and sclerosing agents to block the blood supply to the tumor, often combined with surgical treatment, which can shrink the tumor to a certain extent, and temporary relief of abnormal blood vessels caused by thrombocytopenia, coagulation consumption, and other manifestations [13]. The precondition is that the important blood supply vessels to the tumor are well defined and the side effects of related drugs are known. Radiotherapy has been widely used to inhibit the progress of the tumor, but considering the greater side effects for children, it is rarely used in clinical practice. For pharmaceutical treatment, it includes systemic use of glucocorticoid, vincristine, interferon, antiplatelet drugs, propranolol, and sirolimus [14]. In recent years, the use of sirolimus in the treatment of KHE has been reported more than before. Some literature suggests that sirolimus is still effective in patients with steroid resistance and vincristine inefficacy or in association with KMP, and the effect is long-lasting, but it is necessary to provide in future clinical big data to further support the therapeutic effect of different drugs [15].

Our case showed no signs of thrombocytopenia and coagulation dysfunction during the entire treatment process, and the joint restriction improved after surgical treatment, and the current regular follow-up and re-examination showed that the surgery is successful. There is no recurrence, and follow-up is to be continued.

Conclusion

KHE is a rare disease in clinics; the number of confirmed cases in the world is still relatively few; the etiology and mechanism of the disease are still unknown, which will result in misdiagnose or delayed treatment in the disease early stage. It is necessary to continue to collect relevant cases, unify the standard treatment guidelines as soon as possible, reduce the mortality rate, and improve the quality of life of the cases.

Abbreviations

KHE	Kaposiform hemangioendothelioma
KMP	Kasabach-Merritt syndrome
MAPK	Mitogen-activated protein kinase

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Not applicable.

Authors' contributions

YFZ conceptualized the study and revised and reviewed the manuscript. NR, YG, NS, WQ, and WHG contributed to the draft of the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

Not applicable.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent for publication was verbally obtained orally from the patient for publication of this report and any accompanying images.

Competing interests

The authors declare no competing interests.

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References

1. Luo C, Gao ZP, Dong Z, Lou Y (2021) Kaposiform hemangioendothelioma of bone: a case report with a literature review. Clin Ped Sur 20(2):145–152
2. Dai SY, Peng SH, Chen SY, Ji Y (2020) Recent advances in researches and treatments of kaposiform hemangioendothelioma. J Clin Ped Sur 19(6):544–547
3. Azma R, Alavi S, Khoddami M, Arzanian MT (2014) Nourmohammad A, Esteghamati S. Multifo-cal kaposiform hemangioendothelioma of soft tissue with bilateral pulmonary involvement in an adolescent Korean J Pediatr 57(11):500–504
4. Jin J, Chen X, Zhang J (2005) A case of Kaposiform hemangioendothelioma. J Clin Dermatol 34(1):35–37
5. Li JY, Cai YJ, Sun HL (2021) A case of congenital intracranial hemangioendothelioma in an infant and literature review. J Chin Neurosurg 37(11):1170–1172
6. Guan JW, Wu LG (2018) Report of one case of abdominal hemangioendotheliama and literature review. J Qiqihar Medical University 39(19):2355–2357
7. Xiong Z, Tang SP, Shi W (2014) One case of hemangioendotheliama in the left thigh of a newborn. J Clin Ped Sur 13(6):570–571
8. Xiong Z, Tang S, Shi W, Cao J, Jiang X, Song J, Guibing Fu, Sun Ke, Jianglong Xu, Liu S (2014) Kaposiform hemangioendothelioma in the left thigh of a neonate. Lin chuang xiao er wai ke za zhi 13(6):570–571
9. Li KL, Yao W, Qin ZP et al (2019) Chinese expert consensus on diagnosis and treatment of Kasabach-Merritt phenomenon. China J Oral Maxillofac Surg 3:97–105
10. Ma J, Shi QL, Zhou XJ, Wu SJ, Jiang SJ, Zhang ZJ, Zhou HB (2007) Clinicopathologic features of kaposiform hemangioendothelioma of bone. J Clin Exp Pathol 23(2):189–193
11. Xue M, Liu XJ, Chen T, Tai MZ, Yao SB, Qin ZP (2020) Clinicopathologcal characteristics of Kaposiform hemangioendothelioma. Chin J Cancer Prev Treat 27(13):1253–1258
12. Zhu J, Li C, Liu B, Chen SL, Tian GL (2014) Diagnosis and treatment of Kaposi hemangioendothelioma ofhand. Chin J Bone Joint 3(3):233–237
13. Ferrer Puchol MD, La Parra Casado C, Cervera Araez A et al (2017) Role of selective intra-arterial embolization in benign liver tu-mors. Radiol 59(5):414–421
14. Xu ZQ, Yang XN, Qi ZL (2019) Research progress of sirolimus in Kaposiform hemangioendothelioma treatment. Chin J Aesthet Med 28(8):155–157
15. Wang L, Zhang L, Kong XZ, Yang QL, Bu XJ, Yang SJ (2011) Kaposiform hemangioendothelioma in spleen: a clinicopathological observation. Chin Medical Guide 11(31):248–250

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