

Case Report

Neonatal Biliary Obstruction Caused by Pancreatic Kaposi Hemangioendothelioma

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Abstract:

Kaposiform hemangioendothelioma (KHE), is a rare benign locally invasive proliferation of blood vessels that forms a mass. Besides the mass effect of KHE, it infiltrates adjacent tissues, it can be associated with severe thrombocytopenia, consumptive coagulopathy with high mortality rate. The manifestations of KHE are variable and range from cutaneous lesions with wide varieties of appearances to deep masses without cutaneous signs. We report a case of 3 month old male who presented as extrahepatic biliary atresia in the form of jaundice, hepatomegaly, clay colored stool and elevated liver enzymes. Dynamic Magnetic Resonance Imaging revealed pancreatic head solid mass. Operative findings and biopsy of the highly vascular lesion confirmed that it was KHE of head of pancreas. The mass was excised and hepato-portoenterostomy was successfully performed. One year later the boy is disease free with no organomegaly or any other complication. KHE of head of pancreas is a very rare cause of extrahepatic biliary obstruction early in life. It is amenable to successful surgical intervention.

Level of Evidence of Study: IV (1).

Keywords: Pancreatic Kaposiform hemangioendothelioma; extrahepatic biliary obstruction; hepato-portoenterostomy; neonatal.

Abbreviations: KHE: Kaposiform hemangioendothelioma; EOAE: evoked otoacoustic emission; NICU: Neonatal Intensive Care Unit

Introduction

Kaposiform hemangioendothelioma (KHE) is a rare benign vascular tumor that affects infants and young children. Often KHE presents as a cutaneous birthmark of torso or extremities. Although benign, it can infiltrate into adjacent tissues, with an aggressive locally growing tumor and/or a life-threatening Kasabach–Merritt phenomenon (KMP) of consumption coagulopathy (2). Although visceral involvement is very uncommon, KHE was reported to infiltrate paranasal sinuses (3), deltoid muscle, spleen, bone (4), retroperitoneal, mediastinal or intestinal involvement have been described (5). These tumors tend to be locally invasive. KHE might be multifocal in extreme rare cases, but not known to produce distant metastases (6). The aim of this work is to report that KHE presented as a head of pancreas mass complicated by external compression of common bile duct that presented as biliary obstruction early in life.

Case Presentation

A 3 months old boy presented by jaundice with dark urine and clay colored stool for 15 days. There was no history of bleeding, rash, gastrointestinal tract symptoms, renal symptoms or other system affection. There was no family history of similar condition in the family. By examination

the child weight and height percentiles were within the 50th percentile for his age. Abdominal examination revealed hepatomegaly with no splenomegaly. His laboratory investigations showed elevated total bilirubin 7.4mg/dl and direct bilirubin 3.5mg/dl, alanine aminotransferase 3.9 folds of upper level of normal, aspartate aminotransferase 4,7 folds and elevated lipase while his blood picture was normal and normal coagulation profile. He did abdominal ultrasound which revealed dilated CBD and non-visualized gall bladder. His dynamic MRI of the abdomen with contrast and MRCP showed pancreatic head solid mass with abrupt biliary obstruction at the confluence right and left hepatic ducts. (Figures 1-3). His liver needle biopsy showed major bile duct obstruction compatible with extrahepatic biliary obstruction with moderate portal fibrosis (Figure 4).

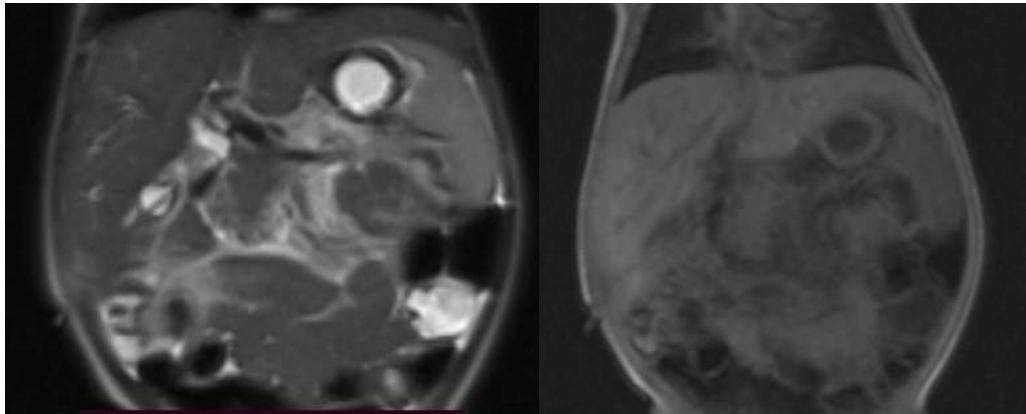


Figure 1. (a) Coronal T2 HASTE sequence and (b) coronal T1 inphase sequence reveals a well defined pancreatic head focal lesion eliciting in T1 and T2 signal intensity, the second part of duodenum is seen splayed over it, with no clear plane of cleavage.

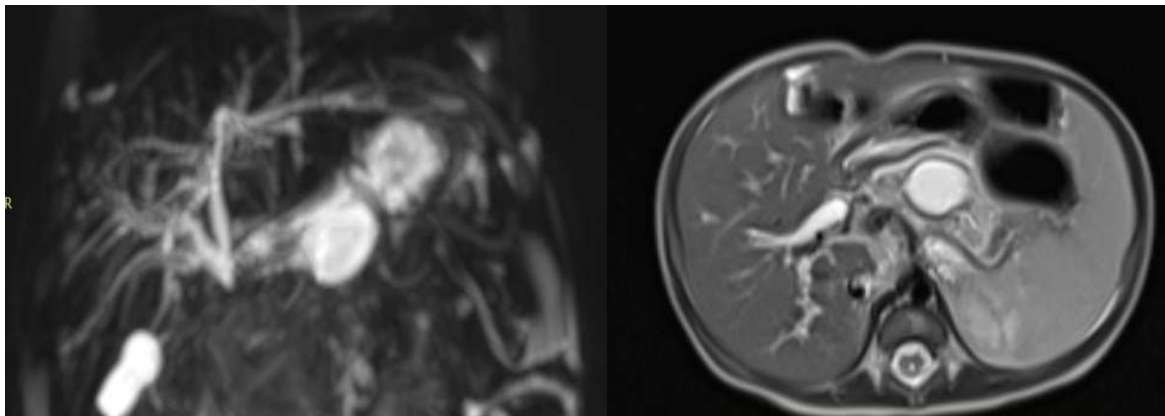


Figure 2. (a) 3D MRCP showing moderate dilatation of the intrahepatic biliary radicles with abrupt termination at the confluence of the right and left hepatic ducts. (b) axial T2 HASTE respiratory triggered sequence reveals a thin-walled cystic lesion inseparable from the body of pancreas, associated with mild dilation of the main pancreatic duct likely representing a pseudopancreatic cyst.

Abdominal exploration was performed, we started by mobilization of the duodenum and exposed the whole parts of pancreas by opening the lesser sac. The second part of the duodenum was found stretched over the tumor, that was involving the head, neck, body of pancreas. The tail of pancreas contained multiple hard nodules from the lesion as well. Dissection over superior mesenteric vein at the neck of pancreas was done as it was encased by the lesion. The lesion was extending to the supra duodenal part of common bile duct, so it was decided to limit the surgery to palliative drainage. Biopsy from pancreatic head mass and mesenteric lymph node showed mainly pancreatic tissue with preserved lobular architecture demonstrating developing acini and ducts with mainly detected islets of Langerhans.

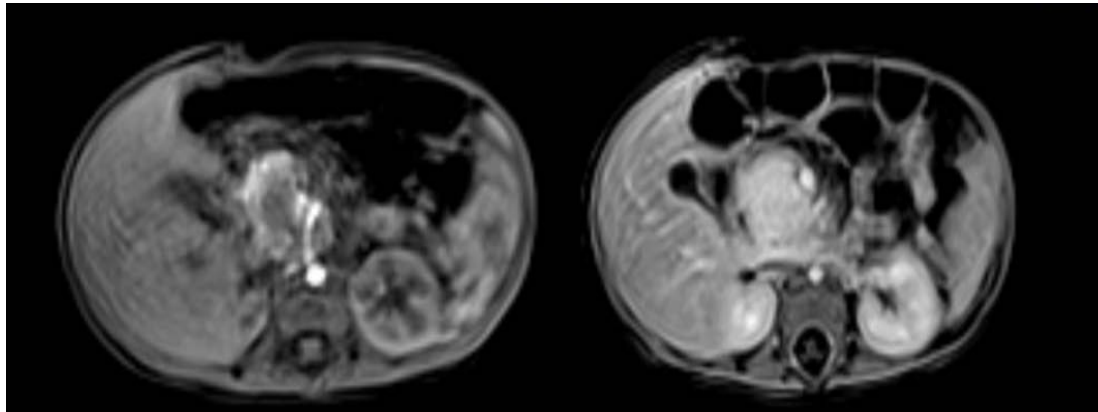


Figure 3. (a) early arterial and (b) early porto-venous phases of dynamic contrast enhanced sequence revealed early peripheral arterial enhancement of the pancreatic head focal lesion with progressive rather homogenous enhancement on porto-venous phase.

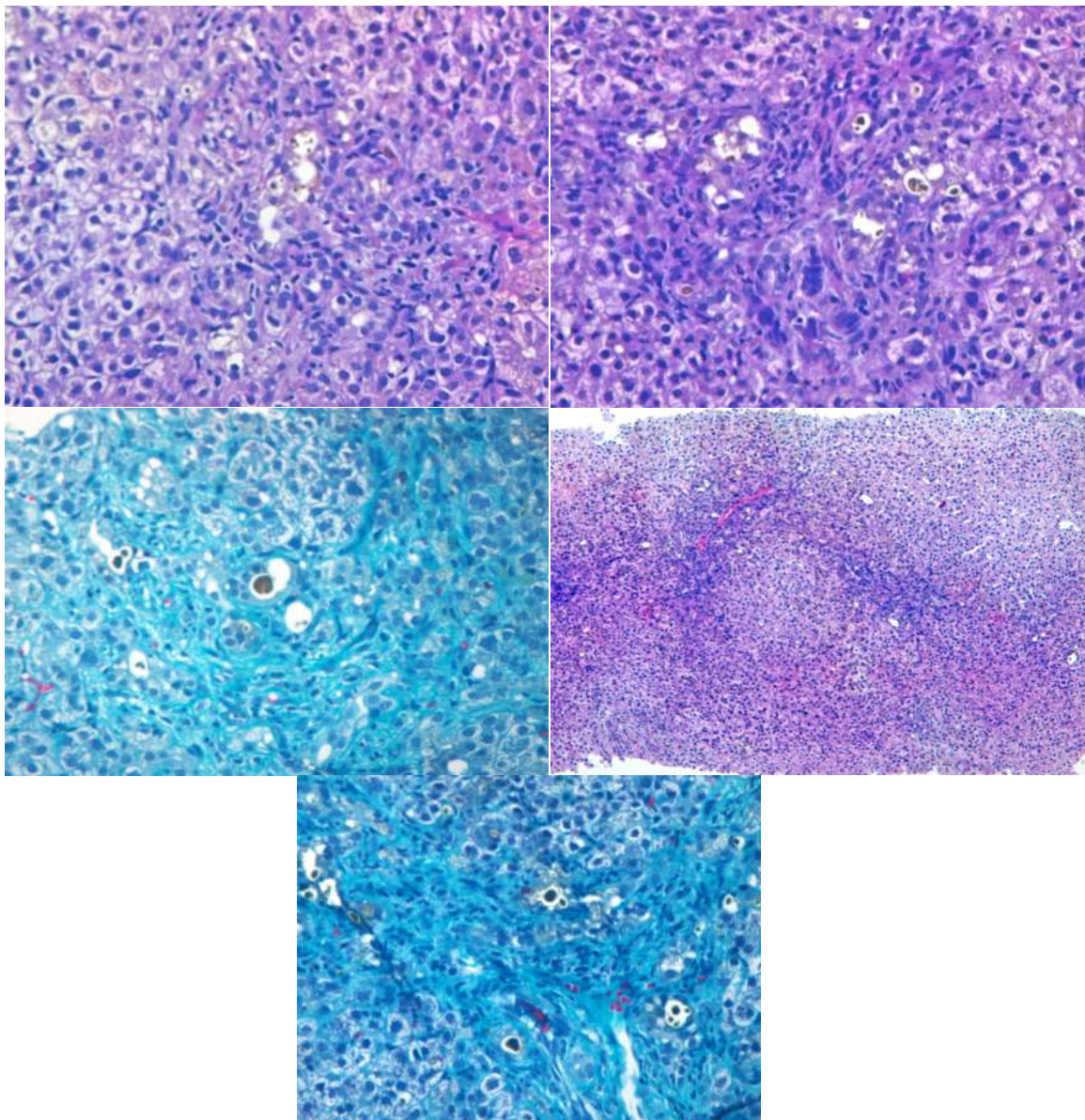


Figure 4. Liver biopsy of the child with Pancreatic Head Kaposihemangioendothelioma. The central area of each photo includes a portal area with proliferated bile ducts showing luminal bile plugs. Original magnification X400 of all except (a), (b) and (d) are haematoxylin and eosin stained while (c) and (e) are trichome stained.

The intervening stroma showed fibrosis, few collections of lymphocytes, hemorrhage, and hemosiderin with crushing artifacts. Few foci showed proliferating spindle cells that have plump hyperchromatic nuclei and eosinophilic cytoplasm, displaying mild to moderate cellular pleomorphism, with rare mitosis. Tumor cells were arranged in lobulated pattern with observed slits containing red blood cells. The mesenteric lymph node biopsy revealed preserved nodal architecture, patent sub capsular and medullary sinuses, and reactive lymphoid follicles. The inter-follicular zone showed reactive lymphocytes, histocytes and proliferating capillaries. The capsule was fibrotic with observed extra nodal attached pancreatic tissue. Pancreatic head mass biopsy was compatible with minimally invasive Kaposiform hemangioendothelioma. One week later, during the second operation retrograde cholecystectomy was performed to reach the dilated common hepatic ducts followed by "ante colic " gastro jejunostomy, hepatico jejunostomy and jejunojejunostomy. A hepato-renal drain was inserted and followed by closure of abdominal wall in layers (Figure 5).

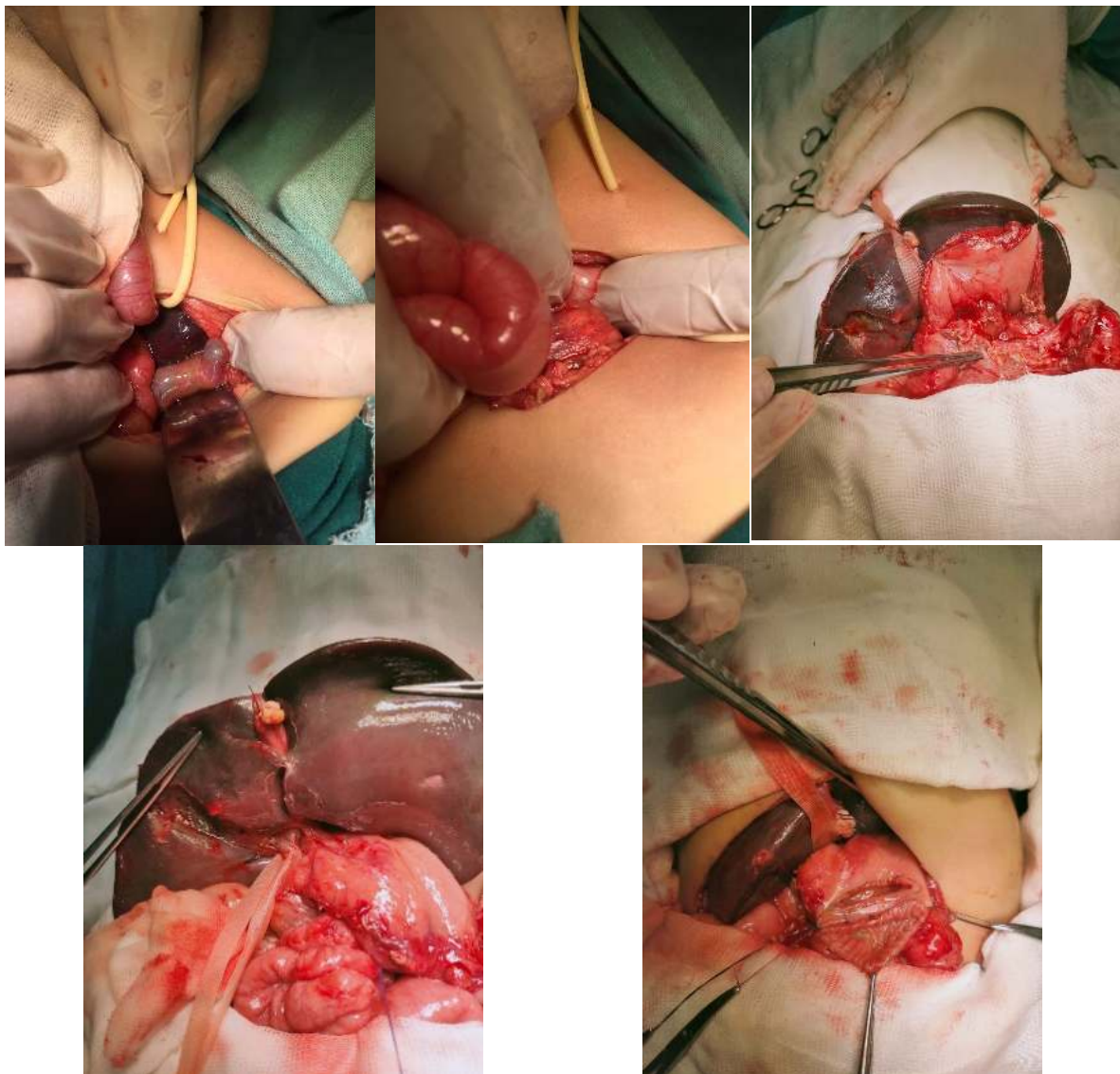


Figure 5. (a) Exploration of hepatic biliary tree. (b) Large head of pancreas mass. (c) tumour resection in the second operation. (d) gastrojejunostomy. (e) hepaticojejunostomy.

Discussion

Kaposiform hemangioendothelioma is a rare, benign locally aggressive vascular tumor that mostly occurs during infancy or early childhood (2). KHE usually develops as a solitary, ill-defined, red-to-purple plaque in the skin or deep soft tissue of the head, neck, trunk, or extremities (7). It might be associated with Kasabach-Merritt phenomenon (KMP), with potentially life-threatening thrombocytopenic coagulopathy (8). KHE of head of pancreas is



extremely rare in the children, yet it was reported as young as 1.5 months old (9), 4 months (10), 8 months (2) and in a 9 year old boy (11). Our case presented at 3 month of life by biliary obstruction not associated by coagulopathy which allowed surgical intervention and removal of the mass. The mass was amenable to resection. We did not resort to sirolimus therapy in our studied infant. Sirolimus was reported to be effective in controlling residual disease and in the Kasabach-Merritt phenomenon. the optimal treatment for KHE is not established and variable responses to different treatment strategies, including vincristine, corticosteroids, propranolol, interferon, and radiation therapy, have been reported. (12), but none of them was encountered in our studied case. The boy followed up for one year and did not develop multifocal disease, recurrence, Kasabach-Merritt phenomenon, bone–joint pain or changes (13), or any of the known complications of KHE.

Different gene mutations were reported to be associated with the development of KHE, but are not unanimous. Hence KHE is believed to have a spectrum of presentations (14). Unfortunately molecular studies were not performed.

Conclusion

Pancreatic KHE of head of pancreas is a very rare cause of extrahepatic biliary obstruction in early life. Diagnosis was made only by dynamic MRI and MRCP. The patient had palliative gastric and biliary drainage as the tumor was irresectable. The patient showed marked clinical and laboratory improvement in the following follow-up to date. It is amenable to successful surgical intervention.

Author Contributions: All authors searched medical literature, databases, conceptualized, conducted the case review and reviewed the final manuscript. All authors have read and agreed to the published version of the manuscript.

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CONFLICT OF INTEREST

The authors declare no conflict of interest in connection with the reported study. Authors declare veracity of information.

References

1. S. Tenny, M. Varacallo, *Evidence Based Medicine*. (StatPearls Publishing; Treasure Island (FL), 2020; <https://www.ncbi.nlm.nih.gov/books/NBK470182/>).
2. D. Mathew, N. Mahomed, Pancreatic kaposiform hemangioendothelioma complicated by Kasabach–Merritt phenomenon: A rare entity. *S. Afr. j. radiol.* **23** (2019), doi:10.4102/sajr.v23i1.1760.
3. B. L. K. Wong, R. C. Dwivedi, L. Masterson, F. Riffat, A. Marker, P. Jani, Kaposiform hemangioendothelioma of paranasal sinus: Kaposiform Hemangioendothelioma of Paranasal Sinus. *The Laryngoscope.* **124**, 2103–2106 (2014).
4. H. B. Dokhi, B. Yahya, F. Alwadi, A. Arafat, M. Almahdi, Kaposiform hemangioendothelioma of the temporal bone in a child. *Journal of Pediatric Surgery Case Reports.* **80**, 102244 (2022).
5. S. Inoue, A. Odaka, Y. Muta, S. Maruta, Y. Takeuchi, Y. Beck, T. Yamashita, Small bowel intussusception secondary to intestinal submucosal cyst in an infant. *Journal of Pediatric Surgery Case Reports.* **80**, 102248 (2022).
6. R. Azma, S. Alavi, M. Khoddami, M. T. Arzanian, A. Nourmohammad, S. Esteghamati, Multifocal kaposiform hemangioendothelioma of soft tissue with bilateral pulmonary involvement in an adolescent. *Korean J Pediatr.* **57**, 500–504 (2014).
7. Q. Liu, L. Jiang, D. Wu, Y. Kan, F. Fu, D. Zhang, Y. Gong, Y. Wang, C. Dong, L. Kong, Clinicopathological features of Kaposiform hemangioendothelioma. *Int J Clin Exp Pathol.* **8**, 13711–13718 (2015).
8. P. Mahajan, J. Margolin, I. Iacobas, Kasabach-Merritt Phenomenon: Classic Presentation and Management Options. *Clinical Medicine Insights: Blood Disorders.* **10**, 1179545X1769984 (2017).



9. Y. J. Ryu, Y. H. Choi, J.-E. Cheon, W. S. Kim, I.-O. Kim, J. E. Park, Y. J. Kim, Imaging findings of Kaposiform Hemangioendothelioma in children. *European Journal of Radiology*. **86**, 198–205 (2017).
10. P. Triana, M. Dore, V. Nuñez, J. Jimenez, M. Miguel, M. Díaz, J. Ricardo, A. Andres, M. Lopez Santamaria, J. Lopez-Gutierrez, Pancreatic Kaposiform Hemangioendothelioma Not Responding to Sirolimus. *European J Pediatr Surg Rep*. **05**, e32–e35 (2017).
11. B. Kaj-Carbaidwala, A. Bernieh, A. E. Russi, A. G. Miethke, M. Abu-El-Haija, A. T. Trout, A. Gupta, K. W. Ricci, J. D. Nathan, Total pancreatectomy with islet autotransplantation in a child with pancreatic kaposiform hemangioendothelioma. *Journal of Pediatric Surgery Case Reports*. **81**, 102266 (2022).
12. Z. Wang, W. Yao, H. Sun, K. Dong, Y. Ma, L. Chen, S. Zheng, K. Li, Sirolimus therapy for kaposiform hemangioendothelioma with long-term follow-up. *J Dermatol*. **46**, 956–961 (2019).
13. Y. Ji, K. Yang, S. Chen, S. Peng, G. Lu, X. Liu, Musculoskeletal complication in kaposiform hemangioendothelioma without Kasabach-Merritt phenomenon: clinical characteristics and management. *Cancer Manag Res*. **10**, 3325–3331 (2018).
14. R. W. Ten Broek, C. Koelsche, A. Eijkelenboom, T. Mentzel, D. Creytens, C. Vokuhl, J. M. van Gorp, Y. M. Versleijen-Jonkers, C. J. van der Vleuten, P. Kemmeren, E. van de Geer, A. von Deimling, U. Flucke, Kaposiform hemangioendothelioma and tufted angioma – (epi)genetic analysis including genome-wide methylation profiling. *Annals of Diagnostic Pathology*. **44**, 151434 (2020).



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