International Journal of Medical Science and Research
www.medicaljournals.in
Online ISSN: 2664-8768, Print ISSN: 2664-875X
Received: 01-02-2024, Accepted: 24-02-2024, Published: 18-03-2024
Volume 6, Issue 1, 2024, Page No. 17-21

The kasabach-merritt phenomenon in a new born: Case report and review of literature

Gertrude Katameya Biselele[1], Manix Ilunga Banza[2*]
[1] Department of Public Health, Faculty of Medicine, University of Lubumbashi, Democratic Republic of Congo
[2] General Surgeon, Department of Surgery, Faculty of Medicine, University of Lubumbashi, University Clinics of Lubumbashi, Lubumbashi, Democratic Republic of Congo

Abstract
Kasabach – Merritt syndrome was firstly described in 1940. It's encompasses a triad of capillary hemangioma, thrombocytopenia and consumptive coagulopathy. Kasabach – Merritt syndrome develops on kaposiform hemangioendothelioma or tufted angioma, confirmed by anatomopathologic analysis which sampling is sometimes difficult to achieve because of bleeding. Consensual care is not established but corticotherapy treatment is unanimous, alone or associated to interferon α 2a or 2b, vincristine, the surgical removal of the mass is controversial.
We report a case of a 5-day old male newborn, with a vascular mass aspect to right forearm who developed on the second day of life an important gastro-intestinal hemorrhage, reason on transfer in teaching university hospital of Lubumbashi. Two complete blood transfusions has been administered to him and biological sampling showed normocytic anaemia, afibrinogenaemia, a severe thrombocytopenia, a disturb of blood test coagulation evoking the Kasabach – Merritt syndrom. After many discussions and consultations, a biopsy was achieved and a diagnosis of tufted angioma has been retained. In the context of the work up of spread, a chest radiography and abdominal ultrasonography were realized without anomalies. Despite the administration of corticotherapy associated to vitamin K, the newborn died the fifth day of life in an important bleeding chart due to consumption coagulopathy.
This study's aim is to present a rare case of congenital giant angioma associated to Kasabach – Merritt syndrom, to present difficulties release to diagnosis and therapeutics in a poor background in doing a literature review and associated.

Keywords: Angioma, newborn, kasabach, merritt syndrome

Introduction

Described for the first time In 1940 by Haig Haigouni kasabach and katharine Krom Merritt, Kasabach-Merritt was used to indicate the association of thrombopenia, afibrinogenemia, and a bulky angioma [1].
It was considered as a complication of classical childhood hemagiomas until 1997.
However, nor Enjolras's works, neither Sarkar's that demonstrated how Kasbach-Merritt's syndrome occured on some vascular tumors, kaposi-like hemangioma and endothelioma, and tufted hemangioma, have confirmed this [2, 3]. The diagnosis is suspected in front of a fast growing vascular tumor, which does not tend to regress spontaneously, unlike a childhood hemangioma [4]. Sarkar and al. proposed the term phenomenon rather than syndrom, because of its occurance on vascular tumors of which histology is different, and its response changeable to treatment [3].
Currently, the phenomenon of Kasabach-Merritt may be defined by the association of one purplish red vascular tumor, quickly far-reaching, of one thrombopenia, of a varying extent of disseminated intravascular coagulation, with sometimes anemia generally occuring to a newborn or an infant aged of less than 6 months [5] although Martinez and al. reported that certain cases in a context of anasarca in placenta and fetus, may be found In utero [6].
The vascular tumor may be congenial or acquired, and many locations have been described in litterature, although skin location on limbs was the most frequent; other locations may be retroperitoneal, mediastinal, pelvic, abdominal viscera, mesenteron [5].
The etiopathology is not clearly demonstrated, but the main hypothesis of this Kasabach-Merritt's phenomenon is a sequestration of platelets in vessels, due to an abnormal and proliferating endothelium [7-8].
This sequestration of platelets should bring about an activation of platelets and consumption of coagulating factors in arise of DIC (disseminated intravascular coagulation), which should encourage thromboses and intratumoral hemorrhage [8]. The clinical feature is varying and may include a brownish red or purplish blotch or small papules interspersed erythematous macula. There may be pale halo surounding the lesion, an inconspicuous hirsutism, a hyperhydrosis. The infiltration and pain to palpation are almost continuons [9]. Nodular and tumoral forms are scarce [5] but described as purplish red affecting limbs and the trunk [3, 10]; the described as badly limited, hardened, often hot, and painful in palpation [10].
Biological examens show a coagulopathy of consumption state with a profound thrombopenia, low platelets rates, a decrease of fibrinogen, a increase of D-dimere and fibrin degradation products (FDP), sometimes an associated iron deficiency anemia due to hemorrhagic syndrome or hemolytic in relation to microangiopathy [7-8]. Biopsy indication of the Kasabach-Merritt phenomenon mass is controverted because of the high risk of hemorrhage due to a surgical gesture and quick spreading infection access risk constituted in this field [5]. Two lesions are brought out: kaposiform hemangioendotheliome and tufted angioma. Dilated lymphatic vessel, hemosiderin deposits, microthrombi are observed in both types of lesions which can coexist on the same skin sample and make anatomopathological examination difficult [11, 12].

Kaposiform hemangioendothelioma and tufted angioma could thus belong to the same histological spectrum [12, 13].
An ultrasound, an arteriovenous doppler ultrasound, and a CT-scan are sometimes performed ; a standard x-ray can show regional osteolysis called Gorham's sign [13]; however, there is no study to establish the specific caracteristics of the Kasabach-Merritt phenomenon tumor [5].
The treatment of the Kasabach-Merritt phenomenon is not codified. Most of publications deal with isolated cases or small series of cases which make the effectiveness of treatment difficult to be established. In the initial phase of the Kasabach-Merritt phenomenon, systemic corticosteroids are often used at the rate of 2 to 5 mg/kg/day. The initial dose must be maintained for 6 weeks at least, in the event of a response [14]; sometimes association of dipyridamol and interferon alpha 2b may be used. In case of failure, adjuvant treatment consisting of embolization of the feeding artery or chemotherapy [15]. However, the therapeutic approach remains multidisciplinary requiring a Pediatric oncologist, a Hematologist, a Dermatologist, a Pediatric surgeon, a Plastic surgeon in the event of a large mass and a Radiologist. The treatment persues two main objectives : the controle of coagulopathy and the eradication of the vascular tumor [7-8]. The emergent treatment aims to correct hematological disorders, especially when an invasive procedure is planned or when hemorrhage occurs [8]. It may include the administration of flesh frozen plasma, cryoprecipitates, and transfusion of packed blood cells. Surgical excision of the Kasabach-Merritt phenomenon vascular tumor must be discused according to its size, adjacent anatomic structures's degree of invasion and the aesthetic of damage. It's often considered when the skin tumor is single, superficial, located in an unvital area; completely resectable [15-17]. The mortality rate varies, depending on publications despite the treatment ; as Sarkar and al. reported 24 % in a study [3], and Enjolras and al. 13% [2]. The etiologies of death vary namely vital structures invasion or compression by the vascular tumor, massive hemorrhage, heart failure, infectious and iatrogenic complications [7, 2]. The objective of this work as for this, is to present a rare case of congenial vascular tumor of the forearm associated with the Kasabach-Merritt phenomenon who died on the 5th day of life, to bring out diagnostic and therapeutic difficulties by reviewing different litteratures.

Patient and Observation

A local medical center transferred us a 2 days male elderly newborn, for congenial mass on the forearm and emission of blood from anus. The history reveals that he was with a mass on the forearm which obstructed the delivery. In the history, the mother was 35 years old, with an obstetric identity Parity4 Gestation3 Abortion0 Death0, living in a mining area, and did not follow prenatal assessment nor had any ultrasound been carried out during pregnancy. She points out that none of her 3 previous children had any malformation. The pregnancy was full term (38 weeks), and the vaginal delivery made expulsion difficult due to the mass In the forearm. The newborn born at term from a single-fetal pregnancy, with an APGAR of 8/9/10. He is the 4th of 4 alive children in a healthy apparence, without any visible congenial malformation found. At birth, even if the newborn' s appearence was normal, doctors noted the presence of a vascular-looking mass on the right forearm. Then he developped a lower digestive hemorrhage manifested by rectal bleeding and anemia, 12 hours later. The hemoglobin level was 8.9g%, 25% of the hematocrit, the blood group O rhesus positive, on the 1st day of life. So, the newborn has been transfused 20 mg/ kg of whole blood. Despite the transfusion, the newborn was still anemic on the 2nd day of life with 9,4 g% of hemoglobin and 26% of hematocrit, then a 2nd 20 mo/kg whole blood transfusion was administrated again. Considering non-improvement of the signs of anemia and the number of transfusions administrated to the newborn, the diagnosis of hemorrhagic disease to the newborn, and a tumor with macroscopically vascular appearence in this newborn, the insufficiency from the technical platform for the treatment of vascular-looking mass, he was transferred to us on the 3rd day of life to the Teaching Hospital of Lubumbashi for better management. On admission we assessed him, and in the physical examination we noted a full-term newborn with pallor of eyelids conjunctivas, anicteric bulbar conjunctivas, a normotense fontanelle, pallor of the oral mucous membranes, moist tongue.
The thorax was tachycard with 183 beats/minutes, respiratory rate 46 cycles/minutes, temperature 36,4 degrees celcius.
The abdomen was not bloated, supple, and depressible, normotensive, no hepatomegaly nor palpated splenomegaly.
The right upper limb, especially the forearm, wrist, and hand on the dorsal side shows an ovoid mass extending from two fingers widths above the radial styloid process to the base of the right upper limb.
Metacarpophalangeal articulation of the fingers, based on a sessile implantation, with a long axis 15 cm transverse, warm to palpation, and grayish in color, encapsulated by a translucent membrane, dotted with a multitude visible vessels [Figure 1] and with a small bleeding visible on the upper surface of the mass [Figure 2].

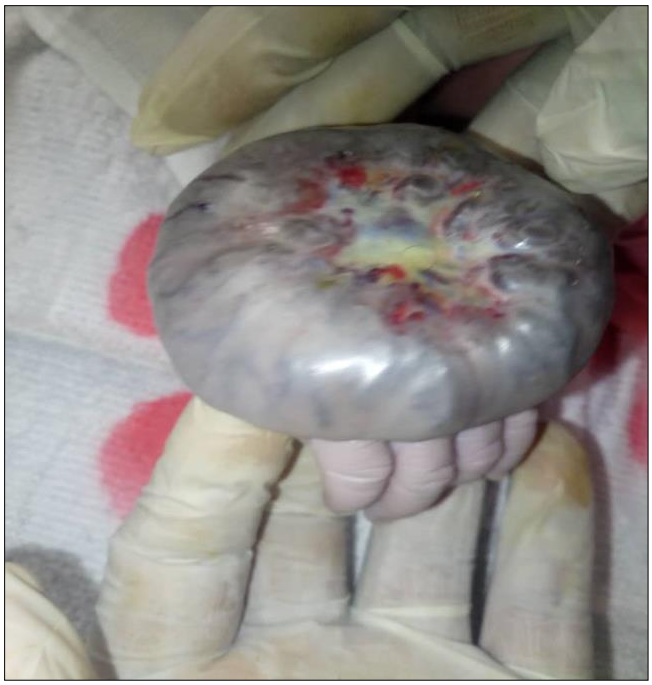

Fig 1: Front vew of the vascular mass in thé foream

Fig 2: Profil view

There is no abnormality clinically detected on the left limb and lower ones.
In view of these elements, we concluded in a congenial vascular tumor associated to hemorrhagic disease of the newborn.
Paraclinical inquiries were carried out and were about **Biology:** Hemoglobin 8, 6 g%, Hematocrit 24%, Blood group and rhesus O+, Mean corpuscular volume (MCV) 92 microgram /mm3, Mean corpuscular hemoglobin concentration (MCHC) 30%, Platelets 40 000/mm3, D-dimer 800/mm3, Fibrinogen 1.8g/l ;

Coagulation test: Prothrombin time (PT) 59%, Activated partial thromboplastin time (APPT): 58 seconds, Bleeding Time (BT) 4 min 30 sec;. Abdominal ultrasound was carried out to look for non-clinically visible abdominal malformations, and no malformation was revealed.

A chest X-ray was performed and revealed no abnormality.
A biopsy sample was taken, usine a biopsy puncture needle on the 4th day of life to confirm the diagnosis, and histological analyses were thus carried out;
The diagnosis of Tufted Angioma was given on histopathological analysis, unfortunately results reached us on the 10th day, the newborn had already died.
Persistent anemia and above all active digestive hemorrhage associated with hematuria during the 4th and 5 th days of life led the newborn in whom undertaken medical treatment consisted of corticosteroid therapy (2 mg/kg of prednisolone intravenously), associated to vitamin K, to death.
The delay in anatomopathological diagnosis is also a cause of uncorrect management in view of litterature.

Discussion

Kasabach-Merritt syndrome also called Kasabach-Merritt phenomenon, is caracterized by a triad maid by hemangioma, thrombocytopenia, and a coagulopathy of consumption [1]. In adults, it is a rare and its incidence is not known. It generally occurs in certain vascular tumors such as tufted angioma and kaposiform hemangioendothelioma [2, 3], which are scarce in the pediatric literature, with respectively 40 % and 10% as incidences [5]. In some cases, these tumors may be described in patients who have not developed Kasabach-Merritt syndrome, however [9, 10, 18], and the incidence of the occurrence of Kasabach-Merritt syndrome on the Kaposiform hemangioendothelioma and tufted angioma is respectively tively 70% and 10% [19]. It is a rare pathology, but more common in children. In wang's series, all 17 patients were newborns aged from 17 hours to 28 days [19]; which is our patient's case, a 3-day-old newborn who developped the Kasabach-Merritt syndrome on the 2nd day of life on a intauterine-developped mass. In a series of 33 cases by Lyons and al. however, the patients were aged from 2 weeks to 20 years with an average age of 3 years and 9 months [18], which is broadly higher than our observation. As our patient is male, this does not allow us to predict a predominance of sex and almost all of the studies described do not confirm any predominance of one sex over the other since they relate either to single cases [9], as in our case or to small samples [18, 19]; Wong says that there is therefore no gender predilection [9]. Kaposiform hemangioendothelioma is a rare, locally aggressive vascular tumor that has been discussed as a possible intermediate malignancy [8]. It is congenial or appears in infants, rarely in adulthood [3, 18, 10]. Tufted angioma is a rare begin vascular tumor, sometimes congenial and often acquired in childhood [5]. In our observation where the tumor which was a tufted angioma, and appeared from the birth, In relation to the location, in his series, Wong SN found that tufted angiomas predominated on limbs [9]; which is the case the case for our observation where the mass was located In the upper limb. However, several locations are described in the literature, notably the neck [1], the left scapula, but also viscera such as the liver [15, 16]. As for laterality, studies do not justifies a predominance of one side over the other, there is no predilection site [2]. The clinical feature of tufted angioma varies, consisting of a plaque, red-brown or purplish, or an erythematous macule dotted with small papules ; nodular or tumoral forms are rare [2], which is the case of our observation, or the clinical form is a well-demarcated ovoid mass, dotted with multiple vessels. Biopsy indication for the mass is controversial due to the hemorrhagic risk linked to the surgical procedure and the risk that is constitutes a gateway to a rapidly spreading infection in this area [7, 14, 8]. The diagnostic confirmation of this mass, however is anatomopathological. Therefore, in our case, biopsy was carried out by qualified personal ; a consultant in this case. Tufted angioma and Kaposiform hemangioendothelioma lesions can coexist on the same skin sample and make pathological examination difficult [11]. This latter also indicates a possible correlation between the histological appearence of the Kasabach-Merritt phenomenon and the biopsy date. The appearence of tufted angioma is often observed in early phase of the Kasabach-Merritt phenomenon, while the appearence of Kaposiform hemangioendothelioma is predominant In the active phase of this condition [2, 11]. The biopsy performed in our case on the third day of life in full evolution of the disease does not allow us to affirm or refute this correlation.
This tumor grows quickly varying from a few months to around ten years, then stabilizes [9]. The death of our child on the fourth day of life unfortunately, does not allow us to support this assertion. The mortality rate varies depending on publications : 24% in Sarkar's study and 13% Enjolras's series [2, 3]. The lethal risk is significant for visceral forms and larger cutanous forms; which is the case in our observation where the mass was very large [2].
Etiologies of death vary, including invasion or compression of vital organs by the vascular tumor, massive hemorrhage, heart failure, infections, iatrogenic complications [2, 7]. Our

newborn died of hemorrhage following a consumption coagulopathy detected through thrombocytopenia; Maguiness states that 12% to 50% of death in Kasabach - Merritt syndrome are due to significant hemorrhage, Disseminated Intravascular Coagulation (DIC), local invasion of vital organs, heart failure, multiple organ failure syndrome, sepsis [7]. The standard treatment of the Kasabach-Merritt syndrome is not established [5], because publications relates either to an observation or small series of cases. Therefore, the efficiency of the treatment is difficult to establish by consensus. The therapeutic approach is multidisciplinary (Oncohematologist, dermatologist, radiologist, Pediatric surgeon, Pediatricians) [7, 8]. The objectives of the management of Kasabach-Merritt syndrome are: the controle of the coagulopathy, and the tumor eradication.

The emergent treatment aims to correct hematologic troubles, especially when an invasive procedure or In the event of hemorrhage [8]. It may include the administration of fresh frozen plasma, cryoprecipitates, transfusion of packed red blood cells [5], thus justifying our unfortunate transfusion of fresh blood due to lack concentrated red blood cells in our institution, but which nevertheless makes it possible to correct the anemia of a part, and bring the coagulation factors on the other part. Authors are not animous on the non-transfusion of platelets which would stimulate the process and increase risk of bleeding, with rapidly consumption of the infused platelets, however [1]. Systemic corticosteroid therapy is an integral part of the medical treatment [19, 5], alone or combined to vincristine, propanolol, dypiridanol, aspirin-ticlopidine, which justifies our corticosteroid treatment with prednisolone intravenously 2mg/kg/ day [16, 17]. Vincristin helps exhibit antimitotic activity, and antiangiogenesis. It is given intravenously at a dosage of 1 to 2 mg/m2/ week, and in the event of failure of monochemotherapy or life risk, we can combine it to polychemotherapy [5]. The Kasabach-Merritt syndrome's 1st sign of therapy is the rise in platelets rate. Then follows the involution of the vascular tumor in weeks **[14]**, although some authors affirm that the evaluation of the criteria of answers currently do not exist [17]. Surgical excision is discussed, however, depending on the tumor's size, its invasion in adjacent anatomic structures, should be considered In a non-vital area, when it is completely resectable [5]. Our patient had a resectable tumor in an accessible area, but his condition did not allow major surgery with significant morbidity and mortality, while Velin reported a total cure without sequelae of Kasabach-Merritt syndrome after totally excised the angioma [16]. Embolization of the vascular tumor can be contemplated if feeding vessels are visualized on arteriography [15], however it exposes us to a risk of migration of emboli, ischemia of a vital organ, exacerbation of hematological troubles, formation of collateral vessels and relapses [5]. Our technical plateform does not allow us this type of surgery however. Despite all these treatment attempts it remains difficult [16], and mortality exceeds 50% [20].

In conclusion, Kasabach-Merritt syndrome causes a triad combining afibrigenemia, thrombocytopenia, and a large angioma. It remains a rare condition, but one to be known in the clinic. The treatment remains controversial according to authors, although.

Conflicts of Interest

Authors declare no conflict of interest

Contribution of Authors

All authors took part in the development of this current work.

All declare having read the final version before submission.

References

1. Kasabach HH, Merritt KK. Capillary Hemangioma with extensive purpura. Am J Dis Child,1940:59:1063-70.
2. Enjolras O, Wassef M, Mazoyer E, Frieden IJ, Rieu PN, Drouet L, *et al*. Infants with Kasabach-Merritt syndrome do not have "true" hemangiomas. J Pediatr,1997:130:631-40.
3. Sarkar M, Mulliken JB, Kozakewich HPW, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with kaposiform hemangioendothelioma and not withcommon infantile hemangioma. Plast Reconstr Surg,1997:100:1377-86.
4. Vin Christian K, McCalmont TH, Frieden IJ. Kaposiform hemangioendothelioma. An aggressive, locally invasive vascular tumor than can mimic hemangioma of infancy. Arch Dermatol,1997:133:1573-8.
5. P Le Nouail, V Viseux, O Enjolras, Phénomène De Kasabach Merritt. Ann Dermatol Venereol,2007:134:580-6.
6. Martinez AE, Robinson MJ, Alexis JB. Kaposiform hemangioendothelioma associated with nonimmune fetal hydrops. Arch Pathol Lab Med,2004:128:678-81.
7. Maguiness S, Guenther L. Kasabach-Merritt Syndrome. J Cutan MedSurg,2002:6:335-9.
8. Hall GW. Kasabach-Merritt syndrome: pathogenesis and management. Br J Haematol,2001:112:851-62.
9. Wong SN, Tay YK. Tufted angioma: a report of five cases. Pediatr Dermatol,2002:19:388-93.
10. Gruman A, Liang MG, Mulliken JB, Fishman SJ, Burrows PE, Kozakewich HP, *et al*. Kaposiform hemangioendothelioma without Kasabach-Merritt phenomenon. J Am Acad Dermatol,2005:52:616-22.
11. Enjolras O, Wassef M, Dosquet Ch, Drouet L, Fortier G, Josset P, *et al*. Syndrome de Kasabach-Merritt sur angiome en touffes congénital. Ann Dermatol Venereol,1998:125:257-60.
12. Mulliken JB, Anupindi S, Ezekowitz RA, Mihm MC Jr. Case records of the Massachusetts General Hospital. Case 13-2004. A newborn girl with a large cutaneous lesion, thrombocytopenia, and anemia. N Eng J Med,2004:350:1764-75.
13. Kerhoas Nicolas K, Le Bidaut M, Dosquet C, Enjolras O, Stalder JF. Syndrome de Kasabach-Merritt d'un membre associé à une ostéolyse ou signe de Gorham. Ann Dermatol Venereol,1997:124:852-4.
14. Teillac Hamel D, Andry P, Bodemer C, Hubert P, Sebag G, Brunelle F, *et al*. Syndrome de Kasabach-Merritt de l'enfant. Ann Pediatr,1992:39:435-41.
15. Steven N Hochwald, Leslie H Blumgart. Giant hepatic hemangioma with Kasabach-Merrit Sydrome: Is the appropriate treatment enucleation or Liver transplantation. HPB surgery,2000:11(6):413-419.
16. Velin P, Dupont D, Golkar A, Valla JS. Neonatal Kasabach-Merritt syndrome healed by complete

surgical excision of the angioma. Arch Pediatr,1998:5:295-7.
17. Drolet BA, Scott LA, Esterly NB, Gosain AK. Early surgical intervention in a patient with Kasabach-Merritt phenomenon. J Pediatr,2001:138:756-8.
18. Lyons LL, North PE, Mac Moune Lai F, Stoler MH, Folpe AL, Weiss SW. Kaposiform hemangioendothelioma: a study of 33 cases emphasizing its pathologic, immunophenotypic, and biologic uniqueness from juvenile hemangioma. Am J Surg pathol,2004:28:559-68.
19. Wang P, Zhou W, Tao L, Zhao N, Chen XW. Clinicals analysis of Kasabach-Merritt syndrome in 17 neonates. BMC Pediatr,2014:14:146.
20. Croteau SE, Liang MG, Kozakewich HP, Alomari AI, Fishman SJ, Mulliken JB, *et al*. Kaposiform hemangioendotrhelioma: atypical features and risks of kasabach-Merrit phenomen in 107 referrals. J pediatr,2013:162:142-7.