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KASABACH-MERRITT SYNDROME – A COMPREHENSIVE REVIEW

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ABSTRACT: Haemangiomas are vascular lesions caused by abnormal blood vessel proliferation, especially in children. Kasabach-Merritt syndrome (KMS) is a rare and life-threatening thrombocytopenia and consumptive coagulopathy associated with aggressive vascular tumors in infancy and younger children. Kasabach-Merritt phenomena is a unique phenomenon in kaposiform hemangioendotheliomas (KHE) and tufted angiomas (TA), occurring in 70% of KHE cases and less often in TA instances. Risk factors for KMS include patient age under 6 months, lesion size greater than 5.0 cm, and the presence of a mixed lesion. Diagnosis of KMS is essential for preventing hemorrhagic complications and establishing a treatment plan. Physical examination, imaging, and biopsy are essential for identifying the underlying vascular tumors. Severe thrombocytopenia, a component of Kaposiform hemangiomas, is a potentially fatal side effect of vascular tumors in children. Treatment aims to manage thrombocytopenia and avoid lethal outcomes. According to SEOP (Spanish Society of Pediatric Oncology) guidelines for vascular tumors complicated by KMS, including vincristine, aspirin, and ticlopidine (VAT therapy). Corticosteroids are the most commonly used acute treatment for KMP, with vincristine being suggested as an effective option.

INTRODUCTION: Haemangiomas are vascular lesionscaused by aberrant blood vessel proliferation. Among children, these are the most prevalent neoplasms. Because of their unique characteristics and behavior, they are very significant in clinical practice. While some haemangiomas are enormous and cause severe deformity, others are tiny and barely visible [1]. Kasabach-Merritt syndrome (KMS) is a rare and life-threatening thrombocytopenia and consumptive coagulopathy associated with aggressive type of vascular tumor in infancy and younger children. It was initially discovered in 1940 by Katharine Krom Merritt and Haig Haigouni Kasabach [2, 3].

It is exclusively related to two kind of vascular tumors such as Kaposiform hemangioendothelioma (KHE) and less frequently in tufted angioma (TA), but not with the more common infantile hemangioma (IH) [4]. The syndrome results in a thrombocytopenic purpura, prolonged PT and activated partial thromboplastin time (APTT), the presence of D dimer and fibrin split products with or without microangiopathic hemolytic anemia, hypofibrinogenemia, consumptive coagulopathy [disseminated intravascular coagulation (DIC)]

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from platelet trapping and aggregation within a specific type of hemangioma (enlarging vascular tumor) [2, 3]. The hypercoagulable state in vascular malformations is caused by anomalies in the endothelium or vessel wall, which in turn leads to abnormal blood flow and subsequent turbulence or stasis, as well as greater tendency of clot [5]. The risk of KMS is greater with increased depth and infiltration of the vascular tumor and with retroperitoneal or intrathoracic involvement [6]. On contrast, in vascular tumors like KHE and related KMP, thrombocytopenia and hypofibrinogenemia are causes secondary with intralessinal trapping with platelet activation and fibrinogen consumption leading to cause high bleeding [5].

Hemangiomas in Kasabach-Merritt syndrome typically occur in the head and neck, trunk, perineum, and upper and lower extremities.About 50% of them have a fully formed lesion at birth. The mortality rate is increase up to 10-37%of cases [7]. All ethnicities and genders appear to be equally harmed. Etiologies of death vary, including invasion or compression of vital organs by the vascular tumor, massive hemorrhage, heart failure, infections, iatrogenic complications. In 1997, investigators from two distinct groups showed that the vascular lesions linked with KMS (or KMP) were not IHs, as previously believed [8]. Treatment was according to SEOP (Spanish Society of Pediatric Oncology) guidelines for vascular tumors complicated by KMP and included vincristine, aspirin and ticlopidine (VAT therapy) [9]. This review highlights the clinical manifestation, pathogenesis, diagnosis, risk factor, complication and treatment of KMP in underlying vascular tumors, specifically KHE, which if untreated may result in significant morbidity and mortality.

Kaposiform Hemangioendothelioma and Tufted Angioma:

Clinical Manifestation: The Kasabach-Merritt phenomena is unique to KHE and TA; it happens in 70% of KHE cases and less often in TA instances [10]. In 1993, Zukerberg and colleagues identified KHE as a unique entity from infantile hemangioma (IH) due to its locally invasive development, aggressive course, and "focal Kaposi- like appearance" [8]. The most typical appearance of kaposiform hemangioendothelioma is a hard, single, purpuric, growing cutaneous or soft tissue lesion, while 10% of patients with KHE lack skin involvement. Multiple tissue planes are frequently implicated, and the margins of the tumor are sometimes unclear. Over 70% of people with KHE develop the KMP, which is a potentially fatal consumptive coagulopathy. Despite its low incidence, KHE can result in morbidity and mortality in both adults and children [11]. Kaposiform hemangioendothelioma primarily affects the trunk, retroperitoneum, and extremities [5, 11]. Therefore, early diagnosis and effective treatment are essential for enhancing patients' long-term prognosis. Uniform guidelines for neonates with KHE are lacking [11]. Prenatal and adult-onset KHE have been described. Adults rarely develop kaposiform hemangioendothelioma, and KMS is not always present. Previously, KMP risk indicators included KHE lesions with a maximal cutaneous diameter of more than 8 cm or those with retroperitoneal or mediastinum infiltration. KHE lesions that spread into several anatomical areas may enhance the risk of KMP, according to Stacy E. Croteau *et al* [12]. Therefore, several treatment modalities have been developed. Prior to 2016, no prospective studies had demonstrated the efficacy and safety of these modalities [11].

Infancy or early childhood, tufted angiomas are rare benign vascular tumors that grow slowly. Tufted angiomas typically exhibit clinical characteristics with common hemangiomas, infantile hemangiopericytomas, and kaposiform hemangioendotheliomas (KHE). The characteristic "cannonball" look of tufted angiomas is caused by the proliferating endothelial cells that form tufts or lobules in the dermis [13].

Epidemiology: According to Croteau *et al.,* the prevalence of KHE has been assessed at 0.91 per 100,000 children per year, and the incidence of KHE is estimated at 0.07 per 100,000 based on observed instances at the Vascular Anomalies Center of Children's Hospital Boston for KHE from 1991 to 2009 [14]. Approximately 90% of these cases appear by the first year of age mortality rate is as high as 30% to 40% as a result of uncontrollable hemorrhage [14, 15]. Because KHE presents at a younger age and has a vascular cutaneous lesion, it can be mistaken for an infantile hemangioma [15].

Histopathology: Both TA and kaposiform hemangioendothelioma originate from the lymphatic and capillary endothelium. Kaposiform Hemangioendothelioma is an infiltrative tumor that typically affects the dermis, muscles, and subcutaneous fat. KHE can be identified histologically by uneven sheets of spindle-shaped endothelial cells and unique slit-like vascular channels. Immunohistochemical staining for lymphatic markers, podoplanin (D2-40), LYVE1, and Prox-1 is positive, while GLUT-1, a marker for infantile hemangioma, is negative. Additionally, immunohistochemical stains for the vascular markers CD31 and CD34 show good results [5]. Tufted angiomas are typified by irregularly sized capillary nodules or tufts that resemble a "cannonball." Immunohistochemistry shows positivity for endothelial and lymphatic vascular markers CD34, CD31, and D2-40.It is a slow-growing tumor; Kasabach-Merritt syndrome (KMS) and consumptive coagulopathy are uncommon consequences [13]. TA exhibits the same immunophenotype as KHE and produces positive Prox-1, D2-40, LYVE1, CD31, and CD34 staining results [5].

Pathophysiology: Physiological pathways have not yet been identified. According to some authors, the core pathophysiology of KMP involves platelet binding, activation, and consumption within the aberrant vascular structure, which results in fibrinogen consumption and degradation as well as the activation and consumption of additional coagulation components [16, 17].

While blood parameters in KMP show alterations similar to those found in disseminated intravascular coagulation (DIC), it is important to stress that this is a limited process that occurs within the vascular lesion and should not be known as DIC. The process of platelet activation and aggregation is self-replicating, as dense bodies of activated platelets emit adenosine diphosphate (ADP), which works locally to activate other platelets [15]. Prothrombin time (PT) and activated partial thromboplastin time (APTT) are usually prolonged, fibrinogen is decreased, and FDPs and D-dimers are raised. It is possible for there to be localized intralesional bleeding, which would enlarge the lesion and worsen the underlying disease. KMP associated with schistocytic hemolytic anemia. Red blood cells undergo mechanical shearing when they come into contact with fibrin clots and rheological forces in the hemangioendothelioma's convoluted, constricted, and partially thrombosed vasculature. Vascular endothelial growth factor, which is secreted by KHE endothelial cells and platelets, further stimulates angiogenesis. Risk factors for Kasabach-Merritt syndrome (KMS) include patient age less than 6 months, lesion size greater than 5.0 cm, and the presence of a mixed lesion [17].

It is important note that there are two other rare condition involving vascular lesions that may also present with DIC. Blue rubber bleb nevus syndrome and Klippel- trenaunay- weber syndrome have also been associated with chronic or intermittent DIC. In these two conditions, venous malformation within various tissues and organs may rarely cause a similar chronic consumptive coagulopathy; however, these conditions are clinically and histologically distinct from KMS, where a single KHE or TA is the source of the hematologic abnormality. Aside from the various coagulopathy induced by KMS, the presence of a large vascular with many vessels causes significant cardiovascular stress. Increased blood flow may cause high-output cardiac infants may present with symptoms of particularly when the lesions are occult [18].

Diagnosis: Laboratory testing and the detection of an underlying vascular tumour are necessary for the diagnosis of KMP. Complete blood count, coagulation panel, and PTT should all be part of the laboratory examination. Thrombocytopenia may be severe. Coagulation studies will reveal a prolonged PT and PTT, hypofibrinogenemia, and increased D-dimer [19, 2, 5]. Early diagnosis is essential to prevent hemorrhagic complications and to establish the treatment plan [16].

A physical examination could be enough to determine the underlying, cutaneous lesion for identification. If a visceral tumor is suspected, further imaging such as Multiphase computerized tomography (CT) and magnetic resonance imaging (MRI) may be useful in establishing both the presence of the tumor and its local extent. MRI may indicate considerable gadolinium enhancement with dermal and subcutaneous thickening [19, 20].

A biopsy of the suspected underlying lesion should be explored to determine the identity of the underlying lession, however it is usually not viable, if KMP is already present due to the risk of hemorrhage [2]. While ultrasound can precisely identify and assess the size, shape, and interior features of lesions, elastography and contrast-enhanced ultrasound may also assess the hardness and softness of the tissue as well as particular internal blood flow patterns; 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 detecting kaposiform hemangioendothelioma and plexiform hemangioma using KMP [21].

On CT scans, KHE manifests as striated soft tissue shadows that improve with contrast delivery. Furthermore, CT is more useful for identifying bone erosion, remodeling, and osteolytic degradation. The CT imaging revealed morphological abnormalities in the involved muscles, including internal punctate or flocculent augmentation [22].

Rib morphological anomalies without significant bone loss. KHE's reported MRI characteristics typically include low signal intensities that are speckled or striated, as well as heterogeneous and high signal intensities on T2WI [23]. Making the distinction between KHE and other vascular tumorsparticularly infantile hemangiomasis beneficial. On MRI, infantile hemangioma presents as a regular and well-defined lesion with a uniformly high signal intensity on T2WI. With minimal damage to the surrounding bone [22]. In addition to infiltrative morphologic characteristics, MRI can assist detect bleeding inside the lesions, hemosiderosis, and grid-like lymphedema [24]. Both CT and MRI are effective techniques for identifying vascular lesions, but CT is faster and allows for a more complete assessment of vessels as well as the identification of common musculoskeletal abnormalities, which may be useful in the early diagnosis and treatment of KHE.The imaging findings showed significantly heterogeneous and ill-defined lesions, the majority of which infiltrated the surrounding tissues. Infantile masses with those features should be noticed from KHE in clinical practice [22].

Risk Factor: Risk factors for KMP in KHE include the lesion's location (more common in the head and neck region, mediastinum, or retroperitoneum), deep infiltration in the muscle or fascia, a tumor diameter larger than 5–8 cm, and the child's young age [16]. Risk factors are linked to the size, depth, and location of the lesion, as well as the age of onset [25]. Some cases, children with other congenital or genetic syndromes may have a higher risk, although these associations are not well defined. There is currently no strong evidence linking Kasabach-Merritt syndrome to specific ethnic groups or genetic backgrounds, but ongoing research may shed light on potential genetic risk factors.

Complication: K Yang, *et al*, reported the major complications other than thrombocytopenia were also recorded. Severe anemia and acquired hypofibrinogenaemia were the two most common haematological outcomes. Other common outcomes were bone-joint invasion or destruction, reduced range of motion, and chronic discomfort. Acute heart failure, active organ bleeding, pericardial effusion, and pleural effusion were relatively infrequent occurrences, occurring only in KMP patients [27].

Treatment: Among the potentially fatal side effects of vascular tumors in children is severe thrombocytopenia, which is a component of KMS [26]. KHE has notably high morbidity and mortality rates (up to 30%) due to severe associated complications and a lack of effective interventions [27]. Diagnosis and treatment of juvenile superficial vascular anomalies can be challenging due to their diversity and range of clinical symptoms. The goal of treating KMS associated with Kaposiform hemangiomas or tufted angiomas is to manage thrombocytopenia and avoid lethal outcomes [28, 29]. Expert recommendations exist, however there are no consensus guidelines for KMP management. Complete surgical excision and embolization of feeder vasculature remains the preferred method, however this is rarely achievable due to the concomitant cytopenia and coagulopathy [30]. Numerous multimodality regimens have been documented; however, they have a lot of adverse effects and varying degrees of efficacy. Includes beta-blockers, immunosuppressants, antibiotics, corticosteroid therapy, interferon therapy,

anticancer medications, antiplatelet therapy, and systemic analgesics. Supportive therapies include fresh frozen plasma (FFP) and platelet transfusions. To prevent additional cardiac compromise, patients with high-output cardiac failure require treatment [28]. However, this kind of malformation typically requires extensive and mutilating surgery. As a result, medical methods are being employed more frequently.

Corticosteroids are the most commonly utilized acute treatment for KMP [24]. Systemic corticosteroid therapy is an integral part of the medical treatment, alone or combined to vincristine, propanolol, dypiridamole, aspirin-ticlopidine, which justifies our corticosteroid treatment with prednisolone intravenously 2mg/kg/ day [31]. Over one-third of KMP patients shown sensitivity to methylprednisolone. Platelets progressively increasing within a week of taking corticosteroids is referred to as sensitivity [26]. The initial methylprednisolone dosage was 2–3 mg/kg/day. Less than a week was used at this dosage. Patients with extremely low platelet counts and life-threatening circumstances may benefit from a greater dose (5–6 mg/kg/day) [32]. The combination of systemic corticosteroids and vincristine is the mainstay of KMP treatment in many centres, and the treatment is listed as the first-line therapy in current guidelines. KHE's impact and risk are a tangible issue considering its young age, high complication rate, and hard therapy [27].

Sirolimus has recently been suggested as an effective treatment option for vascular tumor (VT) with KMP [30], as it results in an increase in the platelet count and a decrease in the tumor size [31]. Particularly sirolimus (rapamycin), as a targeted therapy for complex and refractory vascular malformations refractory to conventional treatment has been introduced with promising results [29]. Rapamycin inhibits the expression of numerous cytokines, including VEGF, and blocks the mammalian target of rapamycin mTOR/PI3K/AKT signal pathways, which generates anti-vascular growth and promotes death [32]. Zhanget al, demonstrate sirolimus as described above was rapid in terms of platelet count and coagulation function, especially when it was combined with steroid and embolization therapy. A more recent report by Harbers *et al*, suggested that low-dose sirolimus successfully cured therapy-resistant patients with congenital vascular malformation, although adverse events such as menstrual cycle disturbances were observed in young adult patients. In addition, Sirolimus does have side effects, including immune suppression, hyperlipidemia, and oral ulcers [33]. It is now the most widely used systemic medication for KHE, with a high response rate and long-term efficacy. For superficial KHE, the use of topical sirolimus is preferable, minimizing the potential infection risk of associated with oral administration [35].

Vincristine (VCR) binds to tubulin and acts by induction of apoptosis and inhibition of mitosis of tumor cells and endothelial cells. VCR is an inexpensive, easily available therapeutic approach with few unwanted effects, making it a good choice for resource-constrained clinics [30]. Vincristine 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 mono- chemotherapy or life risk, we can combine it to polychemotherapy [31]. It is always administered to patients with KMP who have not responded to corticosteroids.

Propranolol is a nonselective β-adrenergic antagonist with different efficacies in the treatment of KMP [34, 35], but its mechanism in treating KHE remains unclear [35]. Oral beta-blockers (propranolol), initially introduced in 2008, are now considered the first-line therapy for HHs.Vasoconstriction, endothelial cell apoptosis, and decreased angiogenesis have been proposed to explain the effects of propranolol on hemangiomas [36]. Wei *et al*, reported successful outcomes with oral administration of propranolol at a dose of 2 mg/kg/day. Over a treatment duration of 6–50 months, demonstrating good long-term safety with no serious adverse reactions [35].

Surgical excision and embolization have been reported a helpful in the management of KMS [18]. For the majority of KHE patients, early surgical intervention may be the most effective treatment. For low-risk patients, surgical treatment provides the advantage of resecting lesions immediately and directly, lowering the risk of thrombocytopenia and eliminating the need for long-term toxic

medications [32]. They have many case reports of success of transcutaneous embolization treatment in the literature. The arteries supplying the proliferation are embolized under radiologic guidance in this procedure. Embolization remains an effective procedure and may be used in combination with surgery and medical therapy [18]. Embolization has also been shown to be effective in KMP patients. Potential side effects include the danger of skin necrosis due to infarction of surrounding tissue and the possibility of life-threatening hemorrhage with the use of intraprocedural heparin with catheter placement [5].

Platelet transfusion alone was ineffective for KMP cases because platelet counts decreased rapidly to previous levels within 48 hours [32]. Platelet transfusion is not indicated except in the case of active bleeding since the transfused platelets can get trapped in the tumor and contribute to more aberrant coagulation and worsening of KMS [34].

Other therapeutic techniques that have been employed include compression and radiation. Localized compression therapy to the vascular tumor has been reported although it is not widely used. Radiation was the original treatment used by Kasabach and Merritt; its use remains controversial. Outcome of radiation therapy showed an increase in platelet count. The total radiation doses were 8-10 Gy. After no hematological abnormalities nor evidence of hemangioma on followup, limb-shortening were noted [18]. A response rate of 75% has been described with the use of radiation in conjunction with steroids; however, radiation may have long-term complications, including growth arrest, developmental delay, and secondary malignancies, and therefore should be used with caution in younger patients [5].

CONCLUSION: Kasabach-Merritt Syndrome (KMS) is a rare but life-threatening condition characterized by severe thrombocytopenia and consumptive coagulopathy associated with aggressive vascular tumors like Kaposiform Hemangioendothelioma (KHE) and Tufted Angioma (TA). Despite advancements in understanding its pathophysiology, KMS remains a significant clinical challenge, especially in pediatric patients. Early and accurate diagnosis is essential to prevent complications and to initiate timely treatment. While corticosteroids, vincristine, and emerging therapies like sirolimus have shown promise, treatment strategies continue to be refined. Future research into the molecular and genetic aspects of KMS could pave the way for novel targeted therapies, improving outcomes and quality of life for affected individuals. Collaborative and multidisciplinary approaches are crucial to optimizing the management of this complex syndrome.

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