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Kasabach-Merritt Syndrome: about a case

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Abstract

Kasabach-Merritt Phenomenon (KMP) is a coagulopathy characterized by profound thrombocytopenia due to platelet entrapment within a pre-existing vascular tumor, tufted angioma or kaposiform hemangioendothelioma. It usually occurs in newborns and infants under 6 months of age. Congenital forms are not uncommon, and some diagnoses have even been made in utero. Imaging is used to confirm the diagnosis and look for deep-seated localizations. This syndrome can be life-threatening, with the risk of haemorrhage, coagulopathy, thrombosis, compression and heart failure. Therapeutic management must be early and multidisciplinary, adapting benefits to therapeutic risks in order to improve prognosis. We report a new case of Kasabach Merritt syndrome, and review the literature to highlight the diagnostic, therapeutic and evolutionary difficulties.

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Introduction

Kasabach-Merritt syndrome is a mainly pediatric entity classically associating a benign vascular tumor, mainly cutaneous and sometimes visceral, thrombocytopenia and a consumption coagulopathy. This syndrome is mainly described in association with two vascular tumors: hemangioendothelioma kaposiforme (HEK) and tufted angioma (TA). It usually occurs in newborns or infants under 6 months of age. Congenital forms are not uncommon, and some diagnoses have even been made in utero. This syndrome is always potentially serious, due to the haemorrhage associated with coagulopathy, and numerous medical and surgical treatments have been proposed. We report on a new case of Kasabach Merritt syndrome, and review the literature to highlight the diagnostic and therapeutic difficulties involved.

Observation

The 4-month-old infant was an only child of non-consanguineous parents, with a vaginal delivery at the end of a well-monitored pregnancy. A maternal cousin had been treated for cervical angioma. Symptoms began at 15 days of age, with the appearance of macules and then purplish papules on the inner surface of the right thigh, which rapidly progressed to an enormous purple-red, hot, tender skin swelling with a circumscribed indurated base extending from the thigh down to the right leg, which was neither ulcerated nor infected (figures 1,2). The right foot was edematous, and the right thigh was 37 cm in circumference. The infant's hemodynamic status was reassuring, with ecchymosis at the injection site.

Biological workup showed microcytic hypochromic anemia (HB: 6.5 GMV 74, MCHR 22), WBC 12,000 with severe thrombocytopenia (5,000/mm3) and hemostasis workup showed prothrombin 85%, high d-dimer levels (41882 ng/ml normal <500 ng/ml), fibrinogen 1.90 g/l normal. His serum electrolytes and renal function tests were within normal limits. Liver function tests revealed normal liver enzymes. Doppler ultrasound was consistent with a vascular, hypoechoic and hypervascularized tumor. Abdominal ultrasound, brain CT and chest X-ray revealed no other associated abnormalities. MRI of the lower limb revealed an intensely raised, early, infiltrative soft-tissue area over the anterior fascia of the right thigh, possibly related to a thigh hemangioma. Skin biopsy in favor of a tufted angioma.

A diagnosis of tufted angioma complicated by MerRitt kasabach thrombocytopenia was made on the basis of the clinical triad of thrombocytopenia, hemorrhagic tendency and presence of a vascular tumor. The infant was transfused with packed red blood cells (platelet IC), then started on oral prednisolone 1 mg/kg/d + adjuvant therapy for 6 weeks, tapering off over several months, and acetylsalicylic acid 10mg/kg/d, Ticlopidine 10mg/k/d and Sirolumus 1mg 1cp/d for 3 months, combined with intermittent compression bandaging. The evolution was marked, one month after the start of treatment, by a rapid improvement in the platelet count, with a change in the coloration of the hemangioma and regression of its volume (4 cm wide). Translated with DeepL.com (free

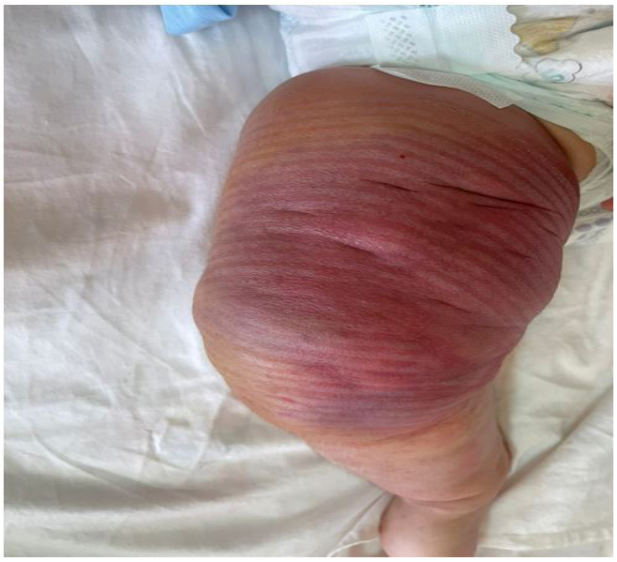

Figure 1: Ventral view of Kasabach-Merritt syndrome.

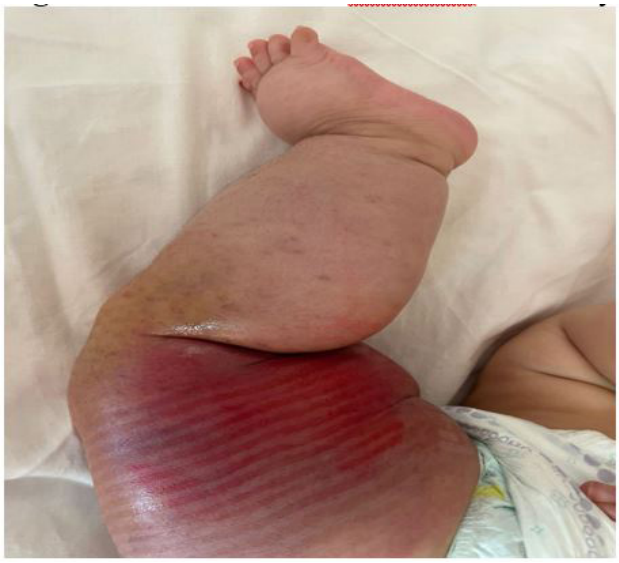

Figure 2: Dorsal view of Kasabach-Merritt syndrome.

version)

Discussion

Kasabach-Merritt syndrome (SKM) is a syndrome that associates a particular vascular lesion, often conveniently classified as an immature hemangioma, with a hematological syndrome dominated by major thrombocytopenia [1]. It is a rare syndrome, thought to complicate 1% of angiomas. It most often affects boys. The age of onset is early: 25% of Kasabach-Merritt syndromes are present in the neonatal period and 80% of cases before 3 months of age [2].

Pathophysiology: The pathophysiological mechanism of SKM remains poorly understood. The initial event is the trapping of platelets by the tumor due to the interaction between the abnormal tumor endothelium and platelets; platelet aggregation and activation then activate coagulation and thus the consumption of coagulation factors. The triggering factor(s) are still unknown. It would appear that the larger the tumour and the deeper the infiltration, the greater the risk of SKM [3, 4]. According to the literature, at least half of all HEK cases are complicated by SKM. Thus, in an infant or newborn, the presence of a vascular tumor of the HEK or AT type constitutes a major risk of developing SKM [4].

Clinical forms of Kasabach-Merritt syndrome:

SKM localizations are cutaneous and more rarely visceral.

CUTANEOUS LOCATION:

There is no anatomical site of predilection [2]. The clinical appearance of the vascular tumor is highly variable: ecchymotic and purpuric, inflammatory or hemorrhagic. Its surface is smooth and shiny, sometimes very fragile, bordering on rupture. On palpation, the tumour is indurated and tender [6]. Petechiae and ecchymosis are present around and at a distance [3, 5].

Visceral localization: In the literature, the vascular tumour of SKM has been observed particularly in the cervico-thoracic region (deep cervical region, mediastinum) or abdomino-pelvic region (retroperitoneum, etc.), which may cause abdominal distension [3,5]. The visceral form of SKM is essentially associated with HEK.

Biology: Thrombocytopenia is constant, often profound and associated with a decrease in fibrinogen levels, a significant increase in D-Dimers and fibrin degradation products, sometimes with iron-deficiency anemia related to the hemorrhagic or hemolytic syndrome of microangiopathy [7], [8].

Differential diagnosis: The differential diagnosis includes, among others, infantile hemangioma, tufter angioma and lymphangiomatosis kaposiforme. The latter occurs mainly in the mediastinum and lungs, in older patients, but is also associated with SKM [9].

IMAGERY: Ultrasound examination is useful for establishing the diagnosis of an often hyper-echogenic mass. Color Doppler confirms the diagnosis with 84% sensitivity and 98% specificity. CT and MRI, on the other hand, help to better define these lesions, particularly in the case of deep-seated localizations [10]. On MRI, these tumors appear poorly limited, T1 isosignal, T2 hypersignal, diffusely enhanced after gadolinium injection and infiltrating subcutaneous tissue [7]. Other diagnostic tools include digital subtraction angiography, bone scintigraphy and standard radiology for regional osteolysis [7]. Ultrasound-guided biopsy can be used to differentiate SKM from other malignant neoplasia, but its use is controversial due to the risk of haemorrhage associated with the surgical procedure, and the risk it represents as a gateway to a rapidly spreading infection in this terrain. TA and HEK lesions may coexist on the same skin sample, making pathological examination difficult [6, 11]. Enjolras et al. suggest a possible correlation between the histological appearance of SKM and the date of biopsy. The appearance of TA is more frequently observed in the early phase of SKM or in the residual stage. In active SKM, HEK is predominant [6, 12]. Labeling for GLUT-1 is negative in SKM [4].

Treatment: The prognosis can be life-threatening in the short term. The aim of treatment is to arrest tumor growth and promote rapid involution, while preserving the function of affected organs [13], [14]. Emergency treatment is aimed at correcting haematological abnormalities, in particular transfusions of packed red blood cells and fibrinogen supplementation (fresh frozen plasma, cryoprecipitate) may be necessary. e Platelet transfusions should be avoided, as they may aggravate coagulation abnormalities; they should only be considered in cases of active, life-threatening haemorrhage [15]. Thus, hemorrhagic skin manifestations such as purpura or ecchymosis do not justify platelet transfusion.

Numerous treatments have been tried. High-dose systemic

corticosteroid therapy (approx. 2-3 mg/kg, sometimes bolused at doses 10 times higher) has long been considered the first-line treatment, although its efficacy has never been demonstrated; in fact, this treatment has most often been disappointing, at least as monotherapy; it is still most often used as first-line therapy, and above all in combination with other treatments. Some cases of radiotherapy and interferon efficacy have been published in the literature, but these treatments are used less and less because of their serious and frequent side-effects: well-known radiotherapy sequelae, neurological complications of interferon in infants such as spastic diplegia. Because of its antiaggregant properties, pentoxyphillin was rarely used successfully and has now been abandoned. So, for some years now, it seems to be generally accepted, based on individual experience, that the main therapeutic resources are:

1. Restraint with bandages, topographically permitting, which mechanically reduces tumor volume

2. Anti-platelet agents: a combination of aspirin and ticlopidine at a dosage of 10 mg/kg/d each [7].

3. Anti-fibrinolytics: aminocaproic acid and tranexamic acid have been used successfully [7].

4. Low-molecular-weight heparin combined with coagulation factors can control consumption coagulopathy [4].

Surgical excision is discussed on the basis of tumour size, invasion of adjacent anatomical structures, and should be considered in a non-vital area when it is completely resectable [1]. Moreover, anesthetic management represents a real challenge, given the severe anemia, thrombocytopenia and coagulopathy requiring preconditioning [16].

• Cryotherapy: its main application is in the treatment of small, superficial, well-defined lesions in the proliferative phase.

• Laser: several sessions are usually necessary to obtain a halt in growth or even the start of involution.

• Vincristine Apart from its anti-mitotic activity, it inhibits angiogenesis. It binds electively to platelets, which may increase its concentration in the PKM mass.

• Sirolimus, an inhibitor of the mTOR pathway, is currently the recommended treatment for HEK complicated by KM. Indeed, several studies have demonstrated the good tolerability and efficacy of this molecule in this type of vascular tumor, in comparison with older treatments such as general corticotherapy or vincristine.

• Close monitoring after treatment discontinuation is necessary in order to rapidly identify recurrence and discuss reintroduction of sirolimus [17].

In our patient, given the ease of administration, the efficacy reported in the literature and its availability in our hospital structure, treatment with Sirolumus was preferred, with a good therapeutic response to date.

Conclusion

Kasabach-Merritt syndrome is a serious condition. It can be responsible for systemic consumption coagulopathy with a fatal outcome in 20-30% of cases. Treatment decisions must be multidisciplinary. It constantly weighs up the short-term vital prognosis against the adverse effects of the treatments used.

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