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Rare Case of a Newborn with Kasabach-Merritt Syndrome

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Abstract

Kasabach-Merritt Syndrome consists of a set of signs and symptoms associated with capillary hemangioma with thrombocytopenia, which causes bleeding accompanied by petechiae, ecchymosis, and spontaneous hematomas. This present study aims to report a rare case of cervical lymphangioma (cystic hygroma) that later evolved into Kasabach-Merritt Syndrome, demonstrating the importance of systemic evaluation of veno-lymphatic malformations. Although rare, prompt diagnosis and therapy are necessary due to the high severity and potential fatality of the condition."

Keywords

Kasabach-Merritt Syndrome, Cervicofacial Lymphangioma, Hemangioma

1. Introduction

Lymphangiomas, also called "cystic hygromas," "lymphangiomatoses," and "circumscribed lymphangiomas," are benign tumors of the lymphatic vessels. They result from the sequestration of portions of primitive embryogenic lymphatic origin that do not efficiently anastomose with larger lymphatic channels and are characterized by lymphatic tissue malformations with highly variable distribution, size, and characteristics. Lymphangiomas present clinically as nodular, painless swellings that are usually located superficially. Lymphangiomas occur in the head and neck in approximately 50% - 70% of reported cases. Approximately 50% of lesions are observed at birth, while 90% develop by 2 years of age. Notably, approximately 4% of all vascular tumors and 25% of benign tumors affecting children are lymphangiomas [1].

Kasabach-Merritt syndrome consists of a set of signs and symptoms associated

with capillary hemangioma with thrombocytopenia (platelets totaling approximately 10,000/mm) and a long-lasting tumor that generates bleeding associated with petechiae, ecchymoses, and spontaneous hematomas. In fact, extensive tuberous hemangiomas evolve rapidly during the first year of life. Thrombocytopenia, associated with the trapping of platelets within the vascular malformation, consists of an extensive proliferative hemangioma, which leads to consumption coagulopathy. Although there are generally no other associated malformations, the systemic evaluation of veno-lymphatic malformations is important [1] [2].

Most patients with Kasabach-Merritt syndrome present with tufted hemangiomas or kaposiform hemangioendotheliomas, which need to be treated because of thrombocytopenia associated with bleeding, possible disseminated intravascular coagulation (DIC), severe anemia, and eventually death. It can occur during the growth process of hemangioma and is later associated with contusion of the lesion. In Kasabach-Merritt syndrome, there is continuous consumption of platelets and clotting factors, as well as triggering of fibrinolysis [1]-[3].

Splenic hemangioma is characterized by a benign tumor that is more frequent in the spleen, although it is sometimes difficult to diagnose and can be part of the Kasabach-Merritt syndrome. Compared with hemangiomas, splenic hemangioendotheliomas have seldom been reported and are characterized by well-circumscribed lesions that are properly separated from the splenic parenchyma, with moderate cellular atypia, little mitotic activity, and an intermediate histological stage between hemangioma and angiosarcoma [4] [5].

In this article, we report a rare case of a cervical lymphangioma (*i.e.*, cystic hygroma) that later evolved into Kasabach-Merritt syndrome.

2. Case Description

Patient F.S.N, female, to term, was born in the maternity ward of a universitary tertiary hospital with an intrauterine diagnosis of extensive cervical lymphangioma. She received injections of OK-432 in 4 sessions shortly after birth, which induced significant regression of the lesions after treatment, as shown in **Figure 1**.

The patient was followed-up at the hospital's otorhinolaryngology and hematology outpatient clinic for cervical lymphangioma. At 1 year and 9 months of age, she needed emergency services when her mother noticed a change in color in the cervical region and hemangiomatous characteristics, in addition to the appearance of lesions on the face, ears, and lower limbs (**Figure 2**).

The patient's mother also reported increased fatigue and changes in the controlled pattern. Laboratory tests, chest X-rays, and echocardiograms (**Figure 3(b)**) revealed the need for drainage (190 ml) of pericardial effusion with Marfan puncture and hospitalization in the intensive care unit to control acute respiratory hemorrhage resulting from airborne transmission and coagulation. Following disseminated intravascular coagulation requiring orotracheal intubation, intensive support, and hematological control, the patient was discharged after 17 days.

The patient's condition evolved with the appearance of hemangiomatous

lesions on the lower limbs, upper eyelids, cervical region, and ears (Figure 2), and the presence of hemangioma in the supraglottic portion of the larynx was also observed via nasofibroscopy. After the appearance of multiple hemangiomatous lesions and complications such as DIC, Kasabach-Merritt syndrome was diagnosed. Systemic corticosteroids were administered, but the child's clinical condition worsened. Magnetic resonance imaging (Figure 3) of the entire abdomen showed the presence of splenic hemangiomas and lymphangiomas with hemodynamic repercussions, and splenectomy was indicated. The spleen of the patient is shown in Figure 4.

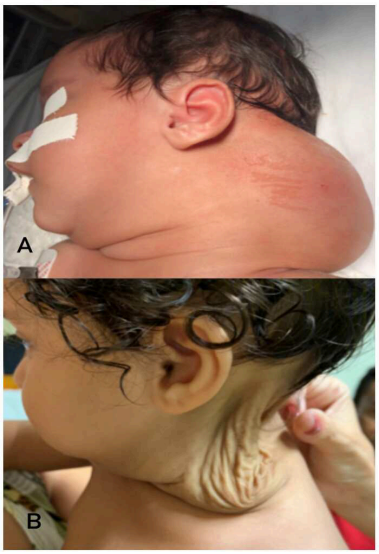

Figure 1. (A) Cervical cystic hygroma before OK 432 (B) After 4 OK-432 sessions.

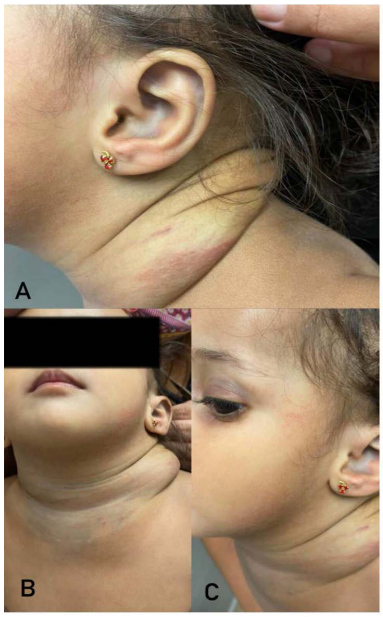

Figure 2. Hemangiomatous lesions in (A and B) the cervical region and (C) the periorbital region.

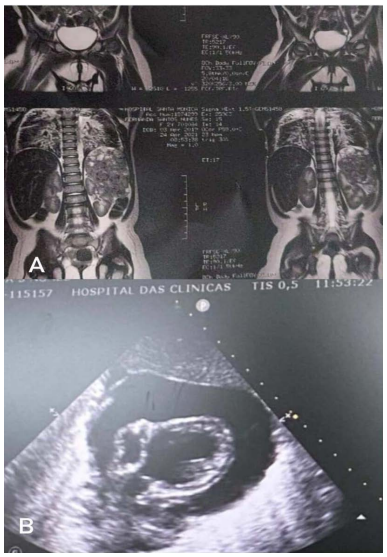

Figure 3. (A) Coronal sections of the MRI of the patient's abdomen (B) Echocardiogram showing pericardial effusion.

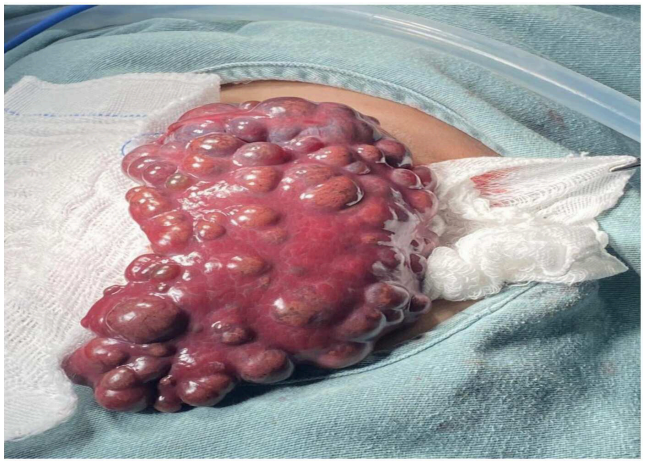

Figure 4. Patient's spleen during splenectomy.

After 29 days in the intensive care unit, the patient was discharged with outpatient assistance. Computed tomography (CT) scans of the neck performed 1 year after the last hospitalization and splenectomy showed the presence of a predominantly cystic heterogeneous formation in the posterior cervical region measuring 6.2 × 4.5 × 2.8 cm (volume = 40 cm^3), while the most recent laboratory tests showed mild thrombocytopenia and prolonged prothrombin activity.

The patient continued to take propranolol 3 mg/kg/day to control the hemangiomatous lesions described above and prednisolone 2 mg/kg/day for six months to treat the syndrome. Over time, significant regression of the hemangiomatous lesions in the cervical region, lower limbs, and occipital region was observed. The patient no longer presented with petechiae, ecchymoses, or spontaneous hematomas. Nasofibroscopy revealed a violaceous hemangiomatous lesion in the supraglottic portion of the larynx, with a significant volumetric reduction compared to

previous examinations.

Currently, the patient diagnosed with Kasabach-Merritt syndrome is receiving joint follow up every other month at the HC-UFG's otorhinolaryngology and hematology outpatient clinic. The patient continued to take propranolol at a dose of 10 mg every 8 h without prednisolone for 8 months, and the dosage of propranolol was reduced in the past 6 months, with no signs of progression of hemangiomatous lesions.

3. Discussion

In the reported case, a child born with cystic hygroma was treated with OK-432 and after 1.5 years presented with hemangiomatous lesions that evolved into respiratory failure within 48 h, which prompted the diagnosis of Kasabach-Merritt syndrome. The evolution differs from what has been reported in the literature to date because although veno-lymphatic diseases may have systemic involvement, such involvement is not described in cystic hygroma, which makes the case an exception in the literature [1].

Kasabach-Merritt syndrome is a consumption coagulopathy associated with the presence of large vascular lesions. Although its pathophysiology remains unclear, the rapid consumption of platelets and fibrinogen in the capillaries of hemangiomas is thought to constitute the root of DIC, which is the chief cause of death in patients with the syndrome. This complication was reported by the patient in a previous case [6].

The diagnosis of Kasabach-Merritt syndrome needs to be based on clinical manifestations, with the presence of hemangiomatosis in association with profound thrombocytopenia and consumption coagulopathy, as in the case described here. Some patients present with visceral forms involving organs such as the liver, spleen, lungs, brain, and intestines. The child in this case had a splenic hemangioma diagnosed based on imaging tests [7].

The diagnosis of Kasabach-Merritt syndrome can be confirmed by imaging and histological examination of hemangiomatous lesions. Ultrasound, CT, magnetic resonance imaging, and digital angiography are important diagnostic aids [8].

Kasabach-Merritt syndrome is not a frequent complication of hemangiomatosis, and few articles describe its treatment, meaning that recommendations are based on case reports or small case series [9]. The treatment of Kasabach-Merritt syndrome consists of supportive care, including the transfusions of platelets, cryoprecipitate, and fresh frozen plasma, as in the reported case [8]. The use of corticosteroids is indicated for patients with hemangiomas when thrombocytopenia is present, and in these cases, is the first option for pharmacological therapy. The drugs used were prednisone or dexamethasone for at least 2 weeks and until the symptoms resolved (2).

It is also necessary to introduce anticoagulant or antiplatelet medications to reduce the risk of bleeding, usually in association with corticosteroids, which are used to prevent platelet aggregation within tumor lesions. Tranexamic acid, used

in the case presented here, is an antihemorrhagic drug administered until hemangiomas resolve.

In view of the satisfactory response to supportive clinical therapy associated with anticoagulant and anti-hemorrhagic drugs, surgical treatment of lesions is typically unnecessary. However, surgical treatment with complete resection of the hemangioma is indicated for the resolution of thrombocytopenia and DIC that are unresponsive to conservative treatments. Despite receiving conservative clinical treatment with corticotherapy, the child in this case developed severe hemodynamic and hematological complications that required surgical therapy (*i.e.*, splenectomy) and brought about clinical improvement.

As described in other case reports, the clinical treatment of hemangiomas and other signs resulting from Kasabach-Merritt syndrome is not always satisfactory, which can make aggressive, even invasive, measures to resect hemangiomatous tumors. In our case, once the cause of the syndrome was recognized and treated, the patient did not present new clinical decompensations during follow-up by the hematology and otorhinolaryngology teams. After controlling for the syndrome and managing the persistence of multiple hemangiomatous lesions, propranolol was continued at a dose of 1 mg/kg/day, providing good control of the remaining lesions. There was no worsening or appearance of new lesions, and the patient did not experience recent hospitalizations or hematological decompensation.

4. Conclusion

Monitoring veno-lymphatic lesions is important, because even when the initial diagnosis is lymphangioma, there may be a change in the clinical evolution, and hemangiomatous involvement may appear that may be a systemic disease.

Conflicts of Interest

The authors declare that they have no affiliations with or involvement in any organization or entity with any financial interest in the subject matter or materials discussed in this manuscript.

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