

Calcified Subgaleal Hematoma With Secondary Cranial Deformity in a Patient With Kasabach-Merritt Phenomenon

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Abstract: We present here the case of a 25-month-old boy who, in conjunction with his preexisting Kasabach-Merritt phenomenon (KMP), developed a massive calcified subgaleal hematoma (SGH) with secondary cranial deformity. Kasabach-Merritt phenomenon is a rare condition that involves a secondary consumptive coagulopathy in the setting of congenital hemangiomatous lesions. The vascular lesions of KMP are often large, singular, and cutaneous, but may also be diffuse, and involve internal structures. Subgaleal hematomas are a rare condition, most frequently observed in neonates as a complication of instrument-assisted delivery. There have been few cases reported beyond the perinatal period, but those that are present within the literature have typically resulted from some instance of minor trauma to the scalp. Most cases resolve spontaneously and without complication. However, our patient's SGH developed into a large, organized, and calcified lesion, likely with some contribution from his hematologic deficit. In addition, this subgaleal lesion resulted in a cranial deformity, ultimately requiring surgical evacuation and reconstruction. We believe the case presented here represents the first report of an SGH in the setting of KMP.

Key Words: Calcified hematoma, chronic hematoma, cranial deformity, Kasabach-Merritt, subgaleal hematoma

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Subgaleal hematomas (SGHs) are a rare condition, most frequently observed in neonates as a complication of instrument-assisted delivery. There have been few cases reported beyond the perinatal period, but those that are present within the literature have typically resulted from some instance of minor trauma to the scalp.^{1–3} Subgaleal hematomas are typically self-limiting and usually resolve with conservative management and without

complications.^{3–7} There have also been scarce reports of SGH occurring in the setting of a congenital coagulopathy. In these instances, it is thought the SGH is a secondary occurrence, a complication of the preexisting hematologic deficit.^{8–15}

Kasabach-Merritt phenomenon (KMP) is a rare condition first described in 1940 as a consumptive coagulopathy in the setting of a congenital hemangiomatous lesion. The vascular lesions of KMP are often large, singular, and cutaneous, but may also be diffuse, and involve internal structures.^{16–19} We present here the case of a young male patient who, in conjunction with his preexisting KMP, developed a massive calcified SGH with secondary cranial deformity.

CLINICAL REPORT

The patient was a 25-month-old boy with a history of KMP coincident with hemangiomas of the spleen, who presented to our clinic on referral from his primary care physician after hitting his head after a fall 3 months previously. At the time of injury, he developed an egg-sized swelling on the left side of his forehead, which was initially treated with conservative management. During the following month, the swelling continued to increase in size. Subsequent computed tomographic scan findings suggested the mass to be an SGH. At that time, it was thought that the mass would resolve over time and so no attempt was made to drain the fluid. The last measured head circumference taken 2 weeks before presentation was 54 cm.

Before presentation to our clinic, the patient underwent partial embolization of the splenic lesions in an attempt to correct his coagulopathy. After that procedure, he recovered from his thrombocytopenia but continued to have low fibrinogen ranging from 100 to 120 mg/dL.

On presentation to our office, he had a very large fluid collection over his upper forehead that extended out into both temporal and parietal regions (Fig. 1). He had several large firm masses within the fluid collection, which were presumed to be a partial calcification of the mass. The remainder of his head and neck examination result was normal. A subsequent computed tomographic scan with contrast of the head ruled out potential complications and causes such as cerebrospinal fluid collection and also suggested significant calcification with an associated cranial deformity. Because of the extremely large size of the mass and the lack of improvement during the preceding several months, he was taken to the operating room for surgical drainage with debridement of the calcifications and cranial recontouring.

The hematoma was exposed using a coronal incision (Fig. 2). The anterior portion just under the forehead had completely calcified within the capsule, and there was very firm, skull-like bone present. After excision of the anterior portion of the capsule and drainage of the hematoma, there remained a very large ridge extending from the forehead into the parietal region and over the vertex of the skull that perfectly outlined the hematoma. There was also a significant skull

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FIGURE 1. A and B, Frontal and side profile photograph of the patient at the time of presentation demonstrating a large SGH.

depression secondary to the compressive effects of the hematoma (Fig. 3). The calcified tissue and ridges were debrided to achieve a more natural cranial contour. There was the additional concern that aggressive recontouring of the bone would result in bleeding secondary to the patient's coagulopathy so some contour deformity remained. Two large Jackson-Pratt drains were placed. There were no complications, and the patient tolerated the procedure well. He was kept for observation overnight and was discharged home the following day.

The drains were removed on postoperative day 13. The patient looked well on examination but had a residual bony ridge above his brows. On a subsequent follow-up 1 year after the procedure, the forehead ridging had resolved and the patient had excellent cranial contour (Fig. 4). After his cranial reconstruction, the patient underwent a splenectomy for treatment of his KMP and has been free of hematologic problems since that procedure.

KASABACH-MERRITT PHENOMENON

First described in 1940 as a case of thrombocytopenic purpura associated with a giant capillary hemangioma in an infant, the cardinal features of KMP include the association of an enlarging vascular lesion, profound thrombocytopenia, a consumptive coagulopathy (with associated hypofibrinogenemia), and some degree of microangiopathic hemolytic anemia.^{18,19} Kasabach-Merritt phenomenon is a rare disorder that can affect infants from the time of birth, or it may appear later in infancy in association with expansion of the vascular malformation. Since the original description of this syndrome, it has been determined that the lesions are not true hemangiomas but are actually distinctive vascular tumors that include tufted hemangiomas and kaposiform hemangioendotheliomas.²⁰ The lesions are typically cutaneous and solitary, but may be diffuse,



FIGURE 3. Intraoperative photograph of patient following surgical resection of the SGH demonstrating the calcified component of the lesion and secondary cranial deformity and ridging.

involving internal structures such as the liver or spleen. However, neither the site nor the size of the vascular lesions can reliably predict the development of KMP or its outcomes.^{16–19} Consumptive coagulopathy being a defining clinical feature of KMP, the pathophysiology is thought to involve platelet trapping by abnormally proliferating endothelium within the hemangioma. This results in platelet activation and the resulting consumption of other clotting factors (eg, fibrinogen) by intralesional thrombosis.^{21,22} Complications associated with KMP include shock, intracranial bleeding and other internal hemorrhage, and cardiac failure from high-volume arteriovenous shunting.²² Although the natural course of the syndrome is resolution within childhood, KMP has been associated with a mortality rate of up to 40% in some reports, with the major cause of death being hemorrhage secondary to the consumptive coagulopathy.^{19,22}

The approach to management of these patients is 2-pronged, with initial efforts toward controlling the acute consumptive coagulopathy, followed by eventual eradication of the offending vascular lesion.^{23,24} Minimally invasive techniques such as embolization of feeder arteries have been used with varying degrees of success; however, the accepted definitive treatment of the hemangiomatous lesions associated with this disorder is surgical intervention and excision. Although there is risk involved in the surgical excision of these lesions in the setting of an uncontrolled coagulopathy, it is always followed by immediate normalization of hematologic parameters

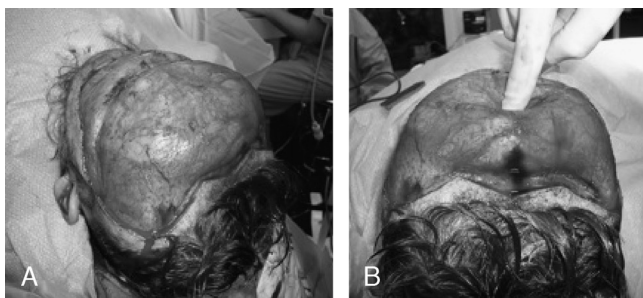


FIGURE 2. A and B, Intraoperative photographs of the patient demonstrating the forehead portion of the hematoma with a thick, calcified capsule.



FIGURE 4. Patient at follow-up visit 1 year after surgical intervention demonstrating resolution of the subgaleal lesion and cranial deformity.

and is curative for either single cutaneous lesions or multiple lesions of the liver and spleen.^{25,26}

SUBGALEAL HEMATOMA

Subgaleal hemorrhage and subsequent hematoma is a relatively uncommon condition, seen mainly in neonates secondary to instrument-assisted delivery. This condition is almost unheard of beyond childhood, a function of the increased vascularity of the subgaleal space in the younger age group. Because the scalp has 5 distinct layers—skin, subcutaneous connective tissue, the galea aponeurosis, loose areolar tissue, and pericranium—there are several planes within which fluid may collect with varying causes and consequences. Between the galea and the pericranium, within a loose areolar layer, run emissary veins that connect the dural venous sinuses to the superficial scalp veins.³ It is this group of vessels that, when damaged, are responsible for SGH.²⁷ In children, the galea is loosely attached to the pericranium, and the area also has a larger supply of blood vessels that makes it more susceptible to bleeding.¹ This space is not limited by cranial sutures, but instead is defined by attachment of the galea anteriorly to the supraorbital ridges, laterally to the zygomatic arches and auricular muscles, and posteriorly to the tissues of the posterior triangle and nuchal ridge.²⁸ Because of the expansive size of the potential space within this layer, and the ease with which it may be dissected under hydrostatic pressure, there is the potential to accumulate a large amount of fluid with potentially serious consequences.

There exist enough cases in the perinatal population such that SGH has been reasonably well characterized in that group. There is a reported incidence of between 1.5 and 4 per 10,000 births. Most cases are the result of vacuum-assisted or forceps-assisted delivery, although SGH has also resulted from fetal scalp vein catheterization and blood sampling or can occur spontaneously.^{9,15,29–32} Diagnosis is made clinically by the presence of a boggy, diffuse swelling of the scalp that crosses suture lines.³¹ The condition is typically self-limiting, and the natural course is usually resolution within 1 to 5 weeks with minimal intervention. And although conservative management is usually sufficient, complicated cases can sometimes require extensive supportive care and surgical evacuation. Serious complications can certainly arise, and there are a number of cases describing the development of hypotension, anemia, hyperbilirubinemia, consumptive coagulopathy, hypotonia, encephalopathy, seizures, disseminated intravascular coagulation, renal failure, and respiratory failure.^{9,29,31,33,34} There have also been reports of long-term sequelae including impaired auditory function, epilepsy, and cerebral palsy secondary to extracranial cerebral compression and ischemic injury to the brain from massive SGH.^{33,35} The mortality rate in this group has been reported to be as high as 22.8%.³²

Beyond the perinatal period, SGH is seen with increasing rarity, and only a handful of cases have been reported past early childhood. Subgaleal hematoma in this population is typically the result of some tangential or radial trauma to the scalp and is usually located in the frontal or parietal regions.^{1–3} Although the cause can be associated with more serious injuries such as skull fractures, SGH has more often been reported as insults as innocuous as hair braiding and pulling and, in some instances, with no determinable mechanism.^{3,7,10,13,36,37} Onset is often insidious, with the hematoma developing slowly for several days to weeks.³ Again, these cases are usually self-limiting and may resolve spontaneously in 1 to 5 weeks with minimal complications.^{3–7} Aside from anemia, the most frequently described complication involves violation of the attachments of the galea to the facial bones and subperiosteal extension into the orbit. This can result in facial edema, proptosis, elevated intraocular pressure, decreased vision, corneal ulceration, persistent blindness and, in some instances, otorrhea, and airway

compromise.^{2,4,10,11,36,38,39} In rare cases, often involving additional complicating factors, patients have developed subacute or chronic SGH with evident organization and calcification and, in one instance, neoplastic transformation to an aggressive osteoblastoma.^{40–43} When intervention has been required, it has typically involved percutaneous drainage, drain placement, and compression dressings.^{13,42,44} Usually, there is an attempt to avoid surgical intervention because this has been associated with increased risk of infection and occasionally resulted in reaccumulation of blood and the need for transfusion.^{5,6,45} However, several reported cases did require surgical evacuation for definitive treatment.^{2,4,10,11,37}

Although rare, there are a number of SGH cases that were complicated by the concomitant presence of a congenital coagulopathy. In the neonatal population, there have been several individual reports of SGH associated with deficiencies of factor VIII, factor IX, and vitamin K, one of which was fatal.^{8,9,14,15} In older children, there have been reports of SGH in association with deficiencies of factor VII and factor XIII, von Willebrand disease, and a congenital hypofibrinogenemia. In at least 1 instance, the SGH was the initial presentation of the bleeding disorder.^{10–13} In addition, 1 case series described the presence of coagulopathies in up to 29% of patients with SGH, although the specific disorders were not elaborated.³² In all of these cases, it was presumed that the SGH was a secondary complication of the coagulopathy.

DISCUSSION

Although there have been a number of cases reported of patients developing SGHs secondary to preexisting coagulopathies, we believe the case presented here represents the first report of an SGH in the setting of KMP.^{8–15} Whereas most cases resolve spontaneously and without complication, our patient's SGH developed into a large, organized, and calcified lesion, likely with some contribution from his lingering hematologic deficit. Although it is difficult to say with certainty that our patient's condition was a direct result of his congenital KMP, the chronic course in this instance implies extraordinary circumstance and at least strongly suggests the possibility. In addition, this subgaleal lesion resulted in a cranial deformity, ultimately requiring surgical evacuation and reconstruction. Although surgical intervention is certainly an established therapeutic modality in refractory cases of SGH, hematoma calcification indicating a need for cranial remodeling is exceedingly rare.^{2,4,10,37,43} We found evidence of only one other case in which a calcified SGH would have reasonably required a similar procedure but which was not performed in that instance.⁴³ Nevertheless, treatment in our patient followed the standard course for each of his rare individual maladies. As such, although it represents a unique coincident episode of 2 rare disorders, this case further strengthens the currently accepted therapeutic methods for each condition on the foundation of a successful outcome.

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