Massive liver haemangioma causing Kasabach–Merritt syndrome in an adult

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ABSTRACT

Liver haemangiomas are common, but their size very rarely exceeds 40cm. Most people with liver haemangiomas are asymptomatic, and diagnosis is usually made incidentally during imaging for other complaints. When a liver haemangioma is symptomatic or produces complications, surgical intervention may be warranted. Kasabach–Merritt syndrome is an uncommon complication reported in certain rare vascular tumours in children, with only a few cases reported in adults. The syndrome describes a consumptive coagulopathy initiated within a vascular tumour, mainly tufted angiomas and kaposiform haemangioendotheliomas and, less commonly, giant haemangiomas. The process can extend beyond the tumour and become disseminated in certain cases due to trauma or surgery. The definitive treatment for giant liver haemangiomas can include arterial embolisation, surgical excision, hepatectomy or even liver transplantation. We report the case of a 32-year-old woman with a 42 × 32 × 27cm (18,870ml) liver haemangioma associated with Kasabach–Merritt syndrome. The diagnosis was challenging, even with proper imaging, owing to the rarity of the condition. It was achieved with an exploratory laparotomy with biopsy.

KEYWORDS

Liver tumour – Giant haemangioma – Cavernous haemangioma – Kasabach–Merritt syndrome – Consumptive coagulopathy

Accepted 7 May 2020

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Background

Liver haemangiomas are very common and usually cause no symptoms. There are very few reports in the literature of cavernous haemangiomas larger than 40cm, and here we present what we believe to be the largest one reported so far. Treatment of liver haemangiomas is usually surgical and is reserved for symptomatic or complicated cases. Kasabach–Merritt syndrome, mostly reported in children and rarely in adults, describes a consumptive coagulopathy initiated within the vascular tumour. The rarity of this syndrome in adults makes the diagnosis very challenging; hence, a high index of suspicion and a good knowledge of the phenomenon is critical for making the correct diagnosis.

Case history

A 32-year-old woman with a non-significant medical history presented to our institution for evaluation of an

Figure 1 The abdomen of the patient

intra-abdominal mass that had been enlarging for four years. Upon examination, the patient had a very large abdomen, completely occupied by the mass (Fig 1). Magnetic resonance imaging showed a huge intraperitoneal mass, lateralised to the right side, measuring 42 × 32 × 27cm in maximal diameters, significantly displacing the abdominal viscera (Fig 2). The radiologist suspected a peritoneal solitary fibrous tumour based on the size and location of the tumour. Laboratory results reported a platelet count of 114,000/ μ l, a haemoglobin of 5.6g/dl, haematocrit of 20%, total bilirubin of 3.13mg/dl (direct was 0.79mg/dl), and an international normalised ratio (INR) of 1.65. The findings were suggestive of haemolytic anaemia with a thrombocytopenic consumptive coagulopathy related to the tumour.

The decision was taken to do an exploratory laparotomy to make a definite diagnosis after correcting the patient's coagulopathy. Upon entering the abdominal cavity, an enormous well-vascularised mass was immediately visible (Fig 3). After careful exploration, it was found to be originating from the right liver. The gallbladder was identified and resected, however, further dissection along the cystic duct was not possible, as the mass completely invaded the hilum and the whole right lobe. Multiple biopsies were taken and sent to pathology. Owing to the atypical appearance of the tumour, its extensive invasion of the right liver and pedicle and the absence of a definite diagnosis, we decided to defer the resection until pathology results were available, allowing for possible adjuvant therapies before resection. The working diagnosis was a

Figure 2 Axial (a) and sagittal (b) T2-weighted magnetic resonance images showing the intraperitoneal origin of the mass, which had an intermediately-high T2 signal intensity. Unenhanced (c) and dynamic contrast-enhanced (d and e) magnetic resonance images, obtained with the volumetric interpolated breath-hold examination, showing the early peripheral discontinuous contrast enhancement and its centripetal progression.

giant liver haemangioma complicated by Kasabach-Merritt syndrome.

The patient's thrombocytopenia was aggravated for the first two days postoperatively, but she recovered to her original values with supportive management alone. One week later, the pathology confirmed the diagnosis of cavernous haemangioma. We decided to offer arterial embolisation to shrink the mass before attempting to do a right hepatectomy, however, the patient refused surgery. She was discharged on aspirin and bisoprolol, and was requested to follow-up regularly in the clinic.

Discussion

Cavernous haemangiomas are the most common benign lesions of the liver, with a prevalence ranging between 0.4% and 20% in the general population.¹ They are usually asymptomatic and discovered incidentally on imaging studies done for other reasons. Haemangiomas are usually classified according to their size into small (0–3cm), medium (3–10cm) and giant (over 10cm), with extremely rare reports of sizes reaching more than 40cm in diameter, as in our patient. The size of our patient's tumour at maximal diameters was 42 × 32 × 27cm, with an estimated volume of 18,870ml, making it, to the best of our knowledge, the largest cavernous haemangioma reported in the English literature so far.

Determining the organ of origin constitutes a challenge with such giant masses. Imaging with computed tomography (CT) or MRI is usually highly sensitive for diagnosing liver haemangiomas, but it is not very specific (55% for

CT and 86% for MRI).¹ On MRI, the mass is typically hypointense on T1-weighted series, hyperintense on T2, and has discontinuous nodular peripheral enhancement that progresses centripetally. Haemangiomas can be mistaken for malignant tumours of the liver, as both masses can be hyperintense on T2-weighted sequences; however, increasing echo time and acquiring diffusion-weighted images can help make the diagnosis.¹

Surgical treatment of giant liver haemangiomas includes enucleation, hepatic resection, or even liver transplantation. Arterial embolisation has been reported to successfully reduce the tumour size and allow for easier resection.² Surgical resection is usually reserved for haemangiomas causing compressive symptoms, such as abdominal discomfort or pain, or when it is associated with complications such as haemorrhage, rupture, thrombosis, infarction that can constitute a surgical emergency. Kasabach-Merritt syndrome is a more rare complication of haemangioma that may warrant surgical resection.³

Kasabach-Merritt syndrome, first described in an infant by Kasabach and Merritt in 1940, is a syndrome that associates giant haemangiomas with consumptive coagulopathy.⁴ Associated laboratory abnormalities include thrombocytopenia, hypofibrinogenaemia, increased fibrinolysis, prolonged INR and elevated D-dimer levels. The syndrome is usually described in children in association with tufted angiomas and kaposiform haemangioendotheliomas. It is seldom seen in adults and is rarely associated with cavernous haemangiomas, as in our patient. The pathophysiology is poorly understood, but it is thought that the abnormal endothelium within the tumour leads to platelet trapping and activation with subsequent consumption and activation

Figure 3 Intraoperative images of the liver haemangioma

of the coagulation cascade, leading to the laboratory findings of consumptive coagulopathy.⁴ The process seems to be localised in the tumour rather than generalised, as suggested by previous studies, but it may be exacerbated by surgery or biopsies, extending into a more disseminated process, manifesting as further thrombocytopenia, anaemia or developing venous thrombosis.⁵

In the few reported adult cases, the presenting symptoms include abdominal pain, increased mass size, disseminated intravascular coagulopathy, bleeding, fractures and high output cardiac failure. Owing to the rarity of this syndrome, no clear guidelines for management are available. In addition to the surgical options discussed above, chemotherapy, steroids, anticoagulants, radiation therapy, aspirin, propranolol, and others have all been reported as possible treatment options for Kasabach–Merritt syndrome.⁴

Liver haemangiomas can reach astounding sizes, as in our patient, and may have dramatic unexpected complications like Kasabach–Merritt syndrome. It is vital for surgeons, radiologists and internists alike to know of such possibilities to correctly diagnose and treat them when encountered.

Acknowledgements

We would like to thank the members of the Faculty of Medical Sciences at the Lebanese University for their continuous support and guidance.

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