Coexistence of splenic hemangioma and vascular malformation of the lower extremity in a child: a case report

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SUMMARY: Gawrych E, Walecka A, Kwas A, Materny J, Sawicki M. Coexistence of splenic hemangioma and vascular malformation of the lower extremity in a child: a case report. Turk J Pediatr 2012; 54: 436-439.

We report a rare finding of the coexistence of splenic hemangioma and progressive vascular malformation of the left lower extremity in a child. The lesion on the left calf was described as a vascular malformation in computed tomography and magnetic resonance. At the age of one year, the abdominal Doppler ultrasound was normal. The examination was repeated at the age of six years due to recurrent pain in the left hypochondrium and revealed giant multiple splenic hemangiomas. The girl underwent splenectomy at the age of 14 years. Histological findings demonstrated multiple cavernous hemangiomas. We present our case report regarding the diagnosis of spleen hemangioma and indications for surgical management in children.

Key words: vascular malformations, splenic hemangioma, diagnostics, child, splenectomy.

The Mulliken and Glowacki[1] classification system is an important tool in establishing the diagnosis of vascular malformations and hemangiomas and distinguishing between the different types. This system divides endothelial malformations into large groups, hemangiomas and vascular malformations, based on their natural history, cellular turnover and histology. Venous malformations are dysplasias of small and large venous channels associated with a variable amount of hamartomatous stroma. They are often found in the extremities and rarely regress[2]. Splenic hemangioma represents the most common benign primary neoplasm of the spleen. Its prevalence at autopsy ranges from 0.03% to 14%[3]. Splenic hemangioma may occur as part of generalized angiomatosis, as seen in Klippel-Trenaunay syndrome or Kasabach-Merritt syndrome[4]. It often has latent clinical symptoms, but spontaneous ruptures have been reported. Ultrasound often presents round, echogenic masses, with or without cystic areas[5]. In computed tomography (CT), it appears as single or multiple lesions that are usually homogeneous, hypodense, or multicystic. They may contain calcifications and generally demonstrate peripheral enhancement after intravenous contrast injection[6]. The final diagnosis is made after histological assessment of a resected spleen. Wilkox et al[7]. Recommended observation of patients with only small, asymptomatic splenic lesions, which met the radiologic criteria for hemangiomas. The treatment usually consists of splenectomy. However, in children, preservation of the spleen is taken into account, as an essential part of the immune system development. In children, splenectomy is associated with an increased incidence and severity of infections[8]. We think that the presented case is interesting since coexistence of a splenic hemangioma and vascular malformations of the extremities in children is a rare congenital disorder.

Case Report

A female child of healthy parents was born by cesarean section, in good condition, with a birth weight of 2200 g. The one-year-old child was observed to have a soft and painless tumor, blue in color, around the left lateral malleolus, gradually widening. It was surgically removed at the Department of Pediatric Surgery and Oncology, Pomeranian Medical University, in Szczecin, Poland.

In the histological examination, cavernosus hemangioma was diagnosed. An abdominal ultrasound was normal with no signs of

tumor. In subsequent years, other superficial and nodular enlarged vessels along the lateral surface of the left calf were found. Arteriography did not reveal any arterial lesions. Ascending venography (with and without ankle compression) showed multiple, dilated collaterals of the anterior tibial veins and segmental dilatation of the proximal femoral vein. Histological examination of surgically excised subcutaneous dilated veins and vascular tumor of the 4th toe of the left foot showed a vascular malformation. Due to progression of vascular lesions in the left lower extremity and left-sided inguinal lymphadenopathy, we performed CT and magnetic resonance imaging (MRI) angiography, which revealed multiple, dilated superficial vessels in the calf, popliteal fossa and one-third of the distal thigh. We also found a conglomerate of pathologic vessels localized just next to the bifurcation of the popliteal artery (Fig. 1). Bilateral inguinal lymphadenopathy was an additional finding. The right lower extremity did not reveal any abnormalities.

At the age of six years, the child presented recurrent dragging pain in the left hypochondrium. The pain was non-radiating and had no precipitating or relieving factors. A blood count showed normal hemoglobin level and normal platelet count. An abdominal Doppler ultrasound showed multiple hyperechoic lesions, with sizes varying between 2.5 cm and 5 cm, suggesting multiple hemangiomas. In the next year of life, an increase in the size of the splenic lesions was observed, reaching 5 cm to 12 cm in diameter. Therefore, multiphasic abdominal CT was performed, which showed multiple, hypodense foci in the spleen. The lesions demonstrated peripheral enhancement after intravenous contrast injection (Fig. 2A, 2B). We decided to observe changes in the spleen with follow-up ultrasonography and to perform splenectomy at a later age in our patient.

The girl underwent an elective splenectomy at the age of 14 years. The spleen measured 14 cm x 10 cm x 3 cm. On the cut surface, we found multiple solid angiomatous lesions, randomly distributed throughout the entire spleen (Fig. 3). Most of the lesions were blue-red, spongy nodules and measured up to 2 cm in diameter. A diagnosis of multiple cavernous hemangiomas of the spleen was confirmed in the histological examination. The patient withstood the procedure well and recovered uneventfully. One year after the procedure, the girl was comfortable and did not experience any infection.

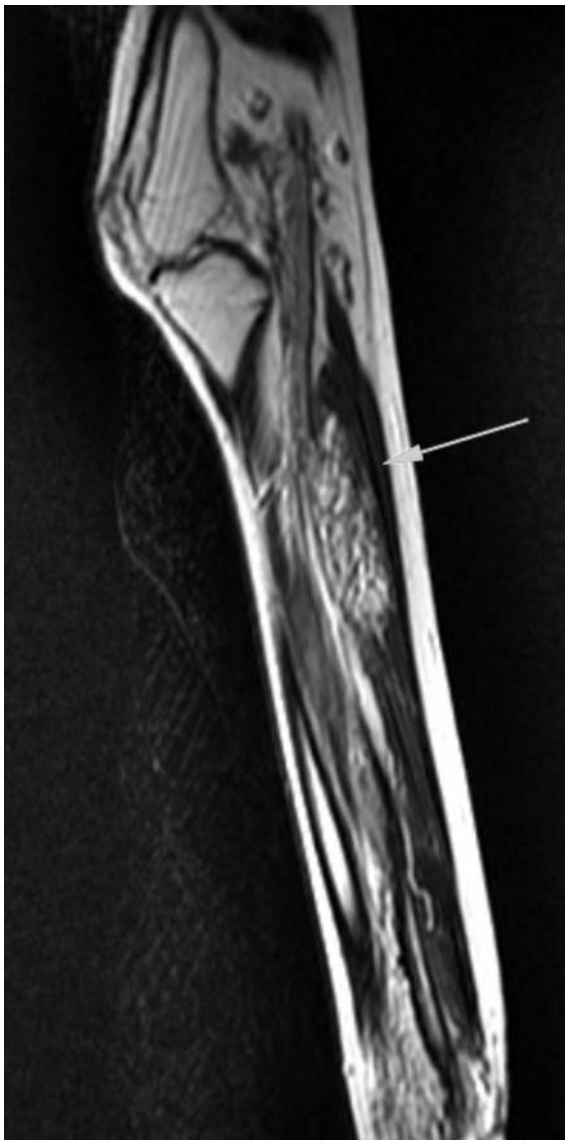

Figure 1. MRI of the calf (T2-weighted image) revealed a complex of pathologic vessels (arrows), which corresponded to arteriovenous malformation (AVM).

Discussion

Vascular malformation is present at birth and enlarges proportionately with the growth of the child. They do not involute spontaneously and may become more apparent as the child

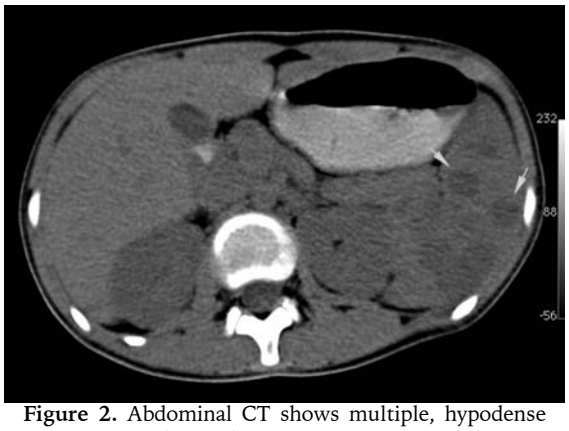

Figure 2. Abdominal CT shows multiple, hypodense splenic lesions (arrows) in the native phase **(A)**. In the venous phase **(B)**, lack of any contrast enhancement was noted, which is characteristic for cavernous hemangioma.

grows. In the presented case, the first lesion on the left shank was observed in child's first year, but clinical progression was evident with the growth of the child. The majority of spleen hemangiomas are asymptomatic for a long time (75-80%), being diagnosed as an incidental finding or during investigative procedures for other disorders[7]. Normal results of the abdominal ultrasound of our patient in the first year of life decreased our vigilance, and we therefore did not include periodic abdominal examination. Abdominal pain first occurred in our patient at the age of six. The abdominal ultrasound revealed a splenic hemangioma at that time. Such lesions usually present asymptomatic slow growth and become symptomatic during the third to fifth decade of life[9].

Sonography is known as a useful examination tool of soft tissue masses, including those suspected to be hemangiomas or vascular malformations. In sonograms, hemangiomas are visualized as well-defined intrasplenic or pedunculated echogenic solid or complex cystic masses. Echogenic calcification with acoustic shadowing may be present[10]. The Doppler examination is crucial to distinguish low- and high-flow vascular malformations[11]. In the presented case, an abdominal Doppler ultrasound showed multiple large hyperechoic lesions from 5 cm to 12 cm in diameter in the spleen, suggesting multiple hemangiomas. As the next ultrasound showed an increase in lesions and more solid structure, the CT was necessary.

According to Chatzoulis et al.[12], CT and MRI fail to achieve appreciable sensitivity and specificity to define the origin of the giant splenic hemangioma. Velkova and Nedeva[13] report a combined sensitivity of 61.3% for ultrasound and CT scan in recognition of liver and spleen hemangiomas, and they emphasized the importance of digital angiography in the proper management of hemangiomas. Ferrozi et al[14]. described CT late- or delayed–phase contrast enhancement of cavernous hemangiomas as discrete mottled areas of heterogeneous attenuation rather than centripetal enhancement as seen in the liver. This finding suggests that the cystic spaces, often central in location, do not contain blood-filled vascular channels. Some authors reported cases of splenic hemangiomas evaluated in MRI. This diagnostic method is a noninvasive and highly sensitive technique that has proven valuable in the diagnostic process. It is the most valuable modality in the classification of vascular malformations and depicts the anatomic relation between the vascular lesion and adjacent organs[10,15]. Final diagnosis is based on histological assessment of a resected solid spleen tumor. Histologically, they are composed of endothelia-lined blood-filled spaces of different size as capillary or cavernous lesion[16]. In the presented case, the normal tissue of the spleen represented a minor part of the organ, as it was dominated by multiple spongy masses and fibrosis.

The treatment of splenic hemangiomas usually involves splenectomy. It is now generally accepted that splenectomy exposes children to

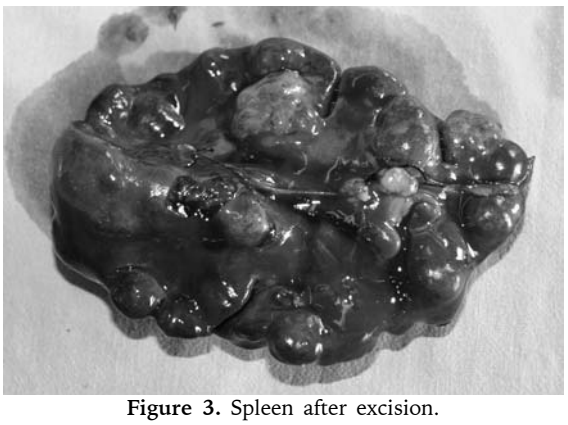

Figure 3. Spleen after excision.

the threat of overwhelming sepsis. The clinical appearance of postsplenectomy infections may vary from mild cases to fatal sepsis. The risk of septicemia is approximately 2%, and it is twice as a great for children younger than four years of age[8]. Jugenburg et al[17]. reported that patients after splenectomy performed up to age five presented a much higher risk of infection, with a rate of infection of 13.8% and a mortality rate of 1.5% compared to the group of older patients. In the presented case, splenectomy was performed at the age of 14 years as the risk of infection was lower.

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