

## A rare case of giant cavernous splenic haemangioma

*Rijedak slučaj divovskog kavernoznog hemangioma slezene*

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### Summary

**Introduction:** Tumors of spleen are generally rare disorders. The most common primary splenic neoplasm is haemangioma, although very rare. Giant, cavernous splenic haemangioma is extremely rare, less than 100 cases have been reported in the literature.

**Case report:** We present a case of giant haemangioma in a 65 years-old patient. Radiological diagnostic methods were not conclusive. Magnetic resonance imaging showed an expansive, heterogenous lesion with signs of necrosis and haemorrhage in the central part. Those characteristics raised suspicion in malignant neoplasm, especially haemangiosarcoma and metastatic neoplasm. The surgical team performed total splenectomy, with special attention to possible bleeding as a severe complication. Pathohistological finding was undoubted. There were certain signs of benign, cavernous splenic haemangioma with areas of coagulative necrosis.

**Conclusion:** We need to be aware of such rare entity. MRI is the most reliable diagnostic method, but the pathohistological analysis is definitive. Surgery is a treatment of choice.

**Key words:** splenic neoplasm, cavernous haemangioma, splenomegaly, giant haemangioma

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### Sažetak

**Uvod:** Tumori slezene općenito su rijetke neoplazme. Najčešći primarni tumor slezene je hemangiom, iako izuzetno rijedak. Divovski, kavernozni hemangiomi slezene iznimno su rijetki, s manje od 100 slučajeva opisanih u literaturi.

**Prikaz slučaja:** Prikazujemo slučaj izuzetno rijetkog divovskog hemangioma u 65-godišnje bolesnice. Radiološke dijagnostičke metode nisu bile uvjerljive. Magnetska rezonancija pokazala je ekspanzivnu, heterogenu leziju sa znakovima nekroze i krvarenja u središnjem dijelu. Ove karakteristike pojačale su sumnju na malignitet neoplazme, posebice sumnju na hemangiosarkom ili metastatsku neoplazmu.

**Zaključak:** Moramo biti svjesni ovog rijetkog entiteta. Magnetska rezonancija je najpouzdanija dijagnostička metoda, ali je patohistološka analiza konačna. Kirurški zahvat je tretman izbora

**Ključne riječi:** neoplazma slezene, kavernozni hemangiom, splenomegalija, divovski hemangiom

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### Introduction

Tumors of the spleen are extremely rare disorders. Generally, spleen tumors are categorised into tumors of primary and tumors of secondary (metastatic) origin. Primary spleen tumors are further categorised into: lymphoid neoplasms – neoplasms of white pulp origin and vascular neoplasms – neoplasms which

arised in red pulp of the spleen.<sup>1</sup>

Vascular neoplasms of the spleen which are infrequently encountered and represent the majority of the nonhematologic / nonlymphoid neoplasms are commonly presented as multifocal lesions. Primary malignant neoplasms involving the spleen

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are lymphoma and angiosarcoma. Primary benign neoplasms arising in the spleen are hemangioma, lymphangioma, littoral cell angioma, splenic cyst and solid lesions such as hamartoma and inflammatory pseudotumor.<sup>1,2</sup>

Metastatic diseases in the spleen are rarely encountered; when a patient has metastatic lesion in the spleen, it is probably not the only site with metastasis.<sup>2</sup> Spleen metastases occur in 2-9 percent of untreated cancer patients.<sup>1</sup>

The most common benign tumor of the spleen, although rare, is haemangioma. Giant, cavernous type of haemangioma are extremely rare. Less than 100 cases have been reported in literature.<sup>3</sup> The average age of patients is 63 years. Splenic haemangiomas are mostly asymptomatic and discovered incidentally.<sup>4</sup> The majority of the tumors are small, on average 2cm, but very rarely, like in our case, they can reach a large size with consequent splenomegaly and abdominal pain. Small tumors can be observed, but in the case of large tumors surgical treatment is indicated. The basic surgical treatment is splenectomy.<sup>5</sup>

We present a case of a rare giant splenic hemangioma, preoperatively suspected of angiosarcoma or metastatic splenic tumor.

### Case presentation

The Ethics Committee of Cantonal Hospital Zenica approved this Case Report and the patient gave us informed consent for the data we used in this article also. Due to urinary problems and feelings of discomfort and tension in the abdomen, a 65 years-old man came to the Primary Health Care Service where the general practitioner did some tests and prescribed an ultrasonography (US). The patient was afebrile and felt well. Except for the painless lump in the left upper abdomen, there were no other clinical signs. Physical examination revealed splenomegaly. The blood tests were normal. US revealed a large, well circumscribed, heteroechoic tumor lesion in the upper lobe, measuring 110mm. There were no ascites. The physician prescribed urgent Magnetic Resonance Imaging (MRI) (Figure 1). Magnetic Resonance Imaging (MRI-Siemens Magnetom Avanto 1.5 T, Erlangen, Germany) of the abdomen showed an enlarged spleen in its upper part and a small hepatic cystic lesion. There was a large, well-defined, heterogeneous, spherical expansive lesion with a maximum diameter of 102mm, involving the upper half of the spleen (Figure 2). The lesion showed hyperintensity on T1 and T2-weighted images, suggestive of necrosis and haemorrhage. The tumor

was close to the adrenal gland, kidney and stomach, displaced adjacent vessels and bowel loops. The liver was enlarged too. At the S2 layer and subcapsular, there were banded zones of T2W hyperintensity, measuring approximately 36×11mm. The lesion showed central hypointensified areas and a peripheral zone of DWI hyperintensity up to 10mm, which is a characteristic of sclerotic haemangioma. Heterogenous, expansive lesion of the spleen, according to MRI characteristics, indicated primary splenic neoplasm, whereby malignant features of angiosarcoma or metastatic neoplasm cannot be ruled out, especially because of the well-defined and non-infiltrative margin.

The medical team presented a classic and laparoscopic splenectomy in treatment to the patient, with all their benefits and risks in this case. He underwent laparotomy with total splenectomy. The spleen was enlarged, measuring approximately 110mm×100mm×50mm with a cystic lesion involving the upper pole.

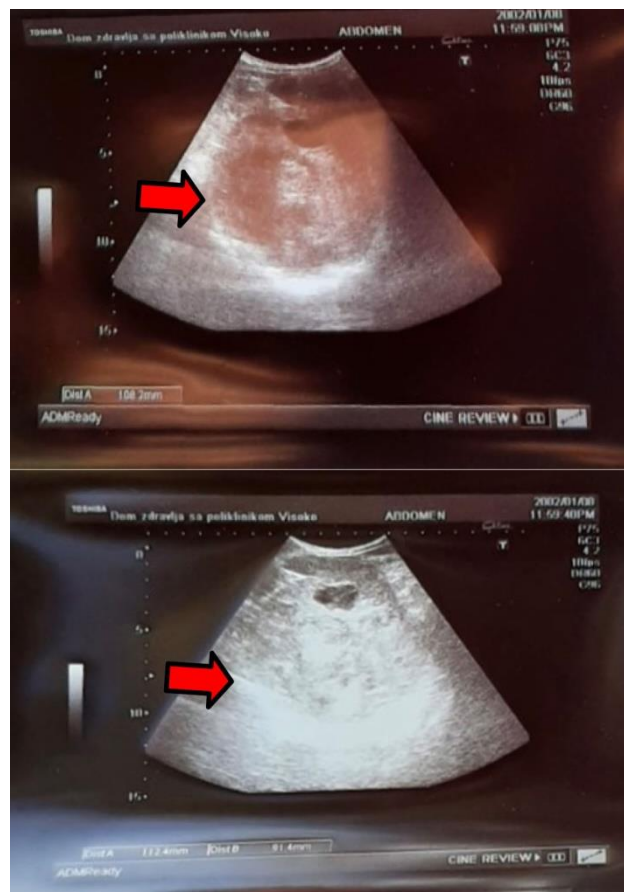


Figure 1 Ultrasound showing well circumscribed, heteroechoic tumor lesion (Medical Centre Visoko)  
 Slika 1. Ultrazvuk pokazuje jasno razgraničenu, heteroehoičnu tumorsku leziju (Dom zdravlja Visoko)

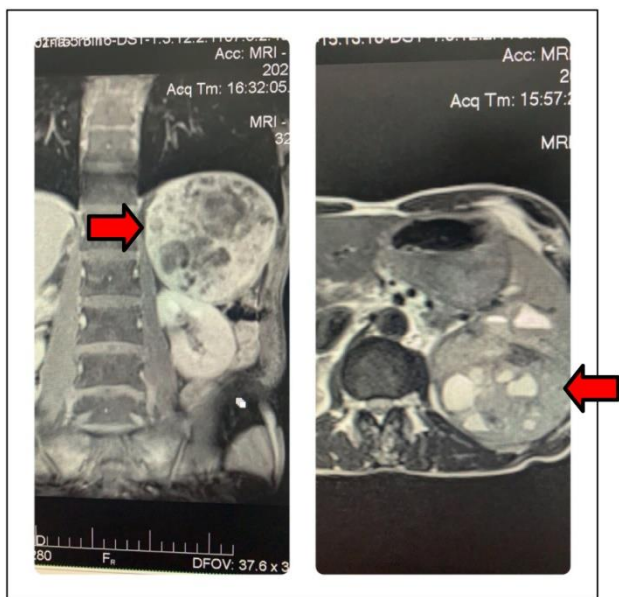


Figure 2 MRI longitudinal and sagittal section showing the massive splenic lesion (Cantonal Hospital Zenica)

*Slika 2. MRI uzdužni i poprečni presjek koji prikazuje masivnu leziju slezene (Kantonalna bolnica Zenica)*

The fluid in the splenic cyst was brownish in colour. Macroscopic examination of the specimen revealed a well delineated tumor of the upper half of the spleen (Figure 3). The cut surface of the tumor was multicystic in appearance, and to a lesser extent solid, pink in color. The tumor was hemorrhagic almost entirely. The cystic part was filled with abundant hemorrhagic content, and the central part of the tumor was softened and yellow. By microscopic analysis (Olympus, model CX31 RBSF), the spleen was occupied by a tumor that was well demarcated and non-encapsulated. The tumor was made up of multiplied vessels of different calibers with blood. Most of them were distinctly dilated, cavernous lumen, lined with a single layer of endothelial cells, with normal cytomorphological appearance, without pronounced atypia and without mitoses. Extensive necrosis of the infarction type was present in the central area. Fibrin thrombi were present in some blood vessels. Immunohistochemical analysis (VENTANA BenchMark ULTRA) showed that the tumor endothelial cells were positive for vascular markers CD31 and CD34; while the marker of proliferative cell activity was low, up to 2% (Figure 4). The definite histopathological diagnosis was giant cavernous haemangioma. The post-operative period was uneventful.

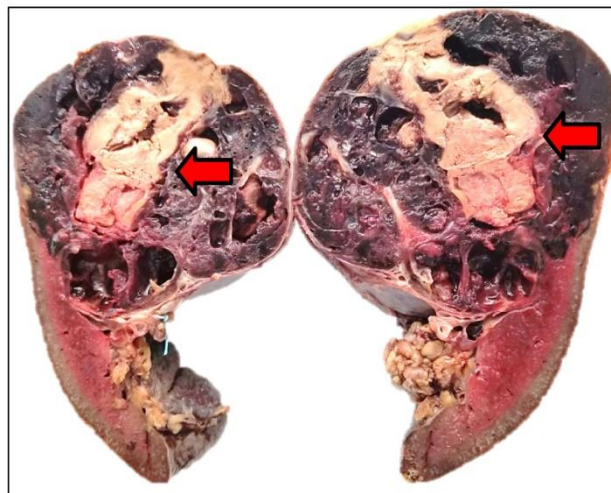


Figure 3. Macroscopic image showing an expansive haemorrhagic lesion of the upper splenic lobe with central necrotic area (red arrow) (Cantonal Hospital Zenica)

*Slika 3. Makroskopski prikaz ekspanzivne hemoragijske lezije gornjeg pola slezene sa središnjim nekrotičnim područjem (crvena strelica) (Kantonalna bolnica Zenica)*

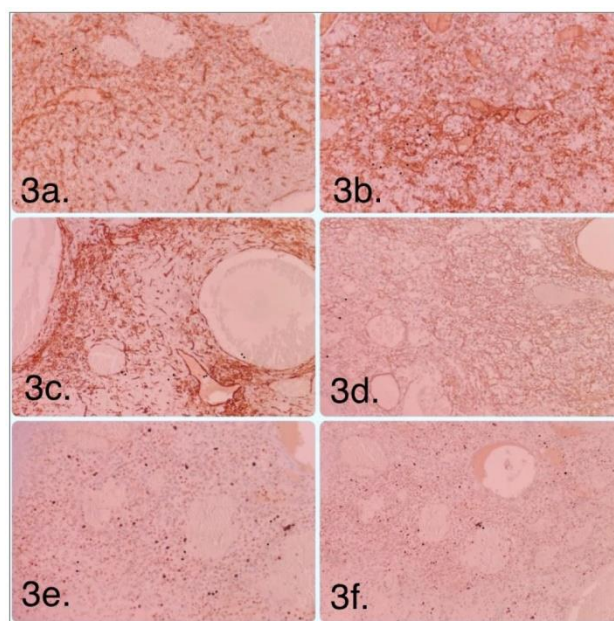


Figure 4 Immunohistochemical analysis with characteristic stainings of the haemangioma; 4a and 4b CD31  $\times 20$ ; 4c CD34  $\times 20$ ; 4d SMA  $\times 20$ ; 4e ki67  $\times 20$ ; 4. ki67  $\times 10$ .

*Slika 4. Imunohistokemijska analiza s bojenjem karakterističnim za hemangiome; 4a. i 4b. CD31  $\times 20$ ; 4c. CD34  $\times 20$ ; 4d. SMA  $\times 20$ ; 4e. Ki67  $\times 20$ ; 4f. Ki67  $\times 10$ .*

## Discussion

Spleen tumors are rare in clinical practice. Although rare, the most common benign neoplasms of the spleen are hemangiomas and they are the most common incidental finding. Hemangiomas are usually up to 2 cm in size, while those of gigantic dimensions are a rarity. Giant splenic hemangiomas are most commonly with a mild clinical picture, due to slow growth primarily. The first presentation or detection of a tumor varies, with an average age between 51 and 63 years<sup>5</sup>, but giant splenic hemangioma was reported in literature in a 10-years-old boy.<sup>3</sup> Preference for gender or race has not been reported. Asymptomatic abdominal mass occurs in 30-45% of cases.<sup>6</sup>

Splenomegaly, abdominal pain, dyspnea, diarrhea or constipation are significant features in several described cases. Tumors can affect an entire organ, can be single or multiple as a part of generalized angiomatosis when they occur in multiple organs, primarily in the liver (as in our case) and skeleton.<sup>7</sup> In our case, MRI-described liver lesion has also the characteristics of haemangioma. Haematological characteristics of splenic haemangioma, mainly in the case of multiple and large haemangiomas, and may present as unexplained coagulopathy caused by platelet trapping, anemia, and thrombocytopenia, as part of the Kasabach Merritt syndrome.<sup>8</sup> In children, abdominal symptoms, such as pain and physical examination findings, are present when cysts are larger than 8 cm.<sup>3</sup>

In the case of giant neoplasms, thrombosis and infarction necrosis often occur and due to abnormalities in the vascular supply of the tumor. In these cases, spontaneous rupture with hemorrhage can also occur as the main complications of this disease. Rupture has been described in about 25% of such cases.<sup>9</sup>

Treatment options range from regular follow-up to complete splenectomy, depending on whether the tumor is symptomatic or asymptomatic, as well as the size of the tumor. When the tumor is small and asymptomatic, only regular monitoring is required. Partial splenectomy can be done when the tumor is symptomatic and located on one of the poles. Total splenectomy is performed when the tumor is large. Extremely rarely, laparoscopic splenectomy may be planned. Another procedure that can help is the embolization of a specific branch of the lienal artery.<sup>10</sup>

Although rare neoplasms, splenic hemangiomas are of great clinical importance which generally lies

in their awareness and differentiation from other conditions, especially metastatic neoplasms and malignant counterpart.

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