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CASE REPORT

Splenosis mimicking metastatic abdominal and pelvic tumours in a 12-year-old female patient

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ABSTRACT

Splenosis is a heterotopic autotransplantation of splenic tissue after rupture of the splenic capsule caused by injury or during elective splenectomy in the management of haematological diseases. Splenosis might cause nonspecific abdominal symptoms but is usually asymptomatic, and it is diagnosed accidentally as an abdominal tumour. It is rarely diagnosed in the paediatric population. We describe a 12-year-old female patient with hereditary spherocytosis admitted to the hospital 3 years after elective splenectomy and cholecystectomy. In medical imaging abdominal and pelvic tumours together with enlarged lymph nodes were found. During laparoscopic surgery many nodules were found with morphology similar to splenic tissue in the omentum, on the surface of small intestine, colon and peritoneum. Results of histopathological examination confirmed splenosis. Splenosis should be considered in differential diagnosis of abdominal tumours in paediatric patients with a history of elective splenectomy or spleen injury.

KEY WORDS:

splenectomy, childhood, splenosis, abdominal tumours, pelvic tumours.

INTRODUCTION

Splenosis is a heterotopic autotransplantation of the splenic tissue. It is usually preceded by a spleen injury or splenectomy. Splenosis is usually asymptomatic, but it can cause non-specific abdominal symptoms or lead to recurrence of the haematological disease for which the splenectomy was performed. It is often diagnosed accidentally during tests performed for other indications unrelated to splenosis. Due to its non-specific course and symptoms, it may raise suspicion of a neoplastic disease [1–4]. The reported prevalence of splenosis following trauma in adults is 26–67% [5–7] and 59% in children [8]. The data on the prevalence of splenosis following elective splenectomy is limited. In children, splenosis is extremely rare - isolated cases have been described [1, 2, 9–15] with a single report of splenosis after splenectomy without rupture of the splenic capsule during the surgery [14, 15]. Below we present a case of splenosis mimicking metastatic abdominal and pelvic tumours in a 12-year-old girl, who underwent splenectomy for congenital spherocytosis. During the procedure the spleen capsule was not macroscopically damaged.

CASE REPORT

A 12-year-old female patient with hereditary spherocytosis (HS), diagnosed during the first month of life,

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after laparoscopic removal of the gallbladder and spleen in April 2018 (there was no macroscopic rupture of the organ capsule during the procedure), was admitted to a district hospital in July 2021 because of a headache with photophobia, nausea, and asthenia. Since 2019, the girl had been under the supervision of a gastroenterology clinic due to symptoms suggestive of irritable bowel syndrome. There was a positive family history of HS. The father of the patient was also affected by HS and had undergone a splenectomy in the past. On admission to the hospital, the child's general condition was assessed as fair. On the physical examination, pain on palpation and palpable lumps in the area of the lower abdomen were of concern.

The ultrasound examination revealed numerous hypoechoic structures, mainly oval, in the true pelvis, above the bladder, and in close proximity to the iliac vessels. The largest one, located adjacent to the left bladder wall, was approx. 24 × 34 mm. Initially the image could correspond to the pathologically enlarged lymph nodes.

Therefore, computed tomography of the abdominal cavity and pelvis was performed (Figure 1). Numerous lesions of size up to 45 × 28 × 32 mm (the largest to the left of the bladder, with contrast enhancement similar to the spleen density) as well as intraperitoneal and retroperitoneal lymph nodes were visualized. Splenosis was suspected. Headaches and photophobia subsided, but gastrointestinal complaints persisted.

After 5 days, the patient was transferred to the Department of Paediatric Haematology and Oncology at Clinical Hospital No.1 in Zabrze for further diagnostics. Basic laboratory tests were within the normal range, and selective markers of neoplastic diseases were negative. The chest X-ray examination showed no abnormalities. Based on the computed tomography image analysis of the abdominal cavity and pelvis, the suspicion of splenosis was supported. Because the neoplastic nature of the lesions could not be ruled out, a laparoscopic surgery was performed. Splenic implants were visualized intraoperatively in greater omentum, mainly in true/lesser pelvis, middle abdomen, also in wall of the cecum and ascending colon, the parietal peritoneum of the right flank and the mesentery of the small intestine.

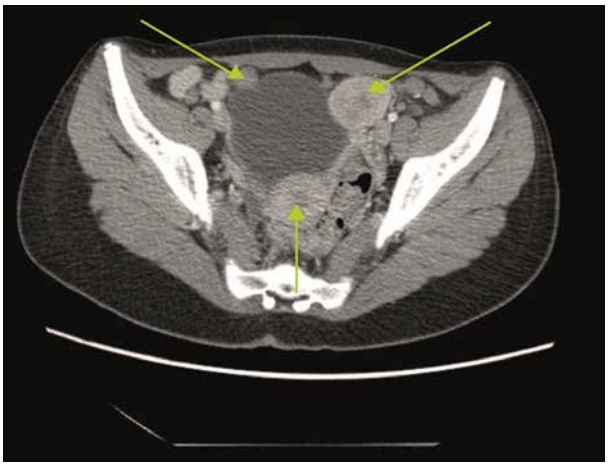

FIGURE 1. Computed tomography scan at presentation: numerous lesions in pelvis, the largest to the left of the bladder, with a contrast enhancement similar to the spleen density

The lesions in the greater omentum and in the true pelvis were larger, even up to 3 cm (Figure 2A). The lesions in the parietal peritoneum were very small and patchy, while those localized in the mesentery reached up to 2 cm (Figure 2B). The mesenteric lymph node (7 × 3 mm) with macroscopically nodular morphology was also visible. The biopsy of the lesions and adjacent lymph node was taken for histopathological examination. The postoperative period was complicated by abdominal pain and diarrhoea.

The histopathological examination of nodular lesions revealed splenic tissue with a predominance of red pulp, significantly dilated sinuses filled with erythrocytes, sparsely represented white pulp, single lymphatic nodules, and small lymphocyte clusters around the spiral arteries (Figure 3). The lymph node showed reactive changes without specific features. The result of the histopathological examination finally confirmed the diagnosis of splenosis and excluded neoplastic disease.

DISCUSSION

Splenosis is an autotransplantation of the splenic tissue in a heterotopic location, usually preceded by a spleen

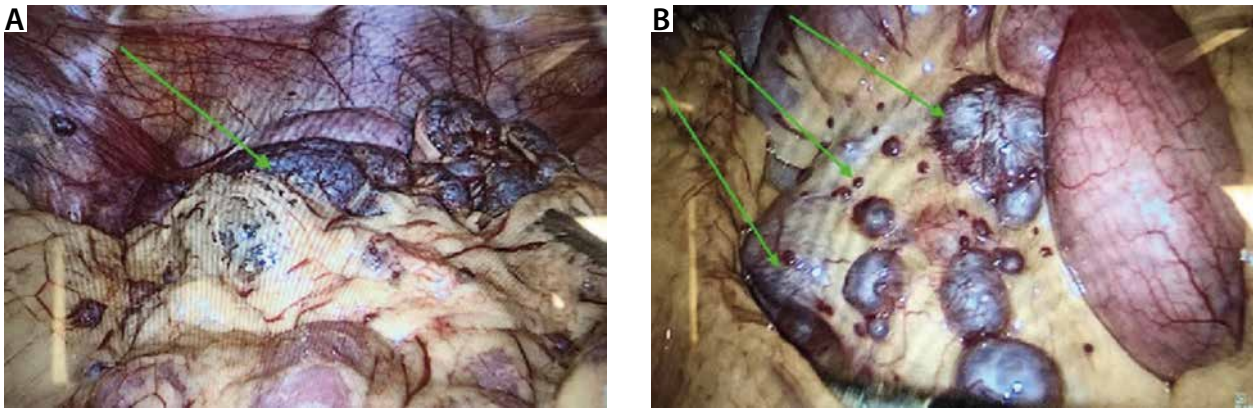

FIGURE 2. A) The largest tumour (arrow) located in greater omentum. **B)** The largest tumour (arrow) located in greater omentum

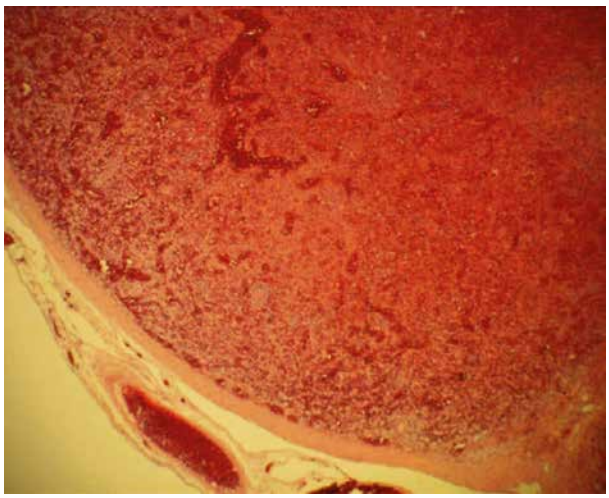

FIGURE 3. The histopathological examination of nodular lesion showing splenic tissue with a predominance of red pulp, significantly dilated sinuses filled with erythrocytes

injury and posttraumatic splenectomy or an elective splenectomy due to haematological diseases. Only a few cases of splenosis in children aged 4–18 years have been described in the literature [1, 2, 9–15]. Most of them report splenectomy after previous spleen injuries, while only single ones report this condition after elective splenectomy in patients with haematological disorders, such as HS [9, 15], Wiskott-Aldrich Syndrome [14], and primary immune thrombocytopaenia [13]. In the described cases, there was usually a release of the spleen parenchyma into the abdominal cavity because of a rupture of the capsule [1, 9] or a sac in which the spleen was fragmented prior to removal [13].

However, splenosis has rarely been found in children after splenectomy during which there was no damage to the splenic capsule. This suggests that even a small number of spleen cells can cause splenosis [14, 15]. It is possible to overlook a slight rupture of the organ capsule. In adults, splenosis also occurs most often after spleen injuries and traumatic splenectomy [11, 13, 15].

There are limited data on the prevalence of splenosis in adults after selective splenectomy for haematological diseases. Return of splenic function after post-traumatic splenectomy, which may indicate splenosis, is reported in 26–67% of adults [5, 7] and 59% of children [8]. It should be noted that these studies were performed on small groups of patients, and they differ in methodology. For this reason, the actual prevalence of splenosis is not precisely defined. In the analysed literature on splenosis in children, the time from spleen injury or splenectomy due to the haematological diseases to the diagnosis of splenosis is from about 1 year [10, 13] to 6 years [2, 14]. However, in patients who have undergone splenectomy in childhood due to trauma, the time to diagnosis varies from 5 months [4, 10, 16] to as much as 55 years [3], 10 years on average [4, 12, 16]. The asymptomatic course and the fact that most of these people are unlikely to undergo any other medical imaging or surgery makes it even more difficult to estimate the prevalence of splenosis [2, 11, 14, 16] and may explain why it is diagnosed less frequently in children than in adults [12].

Rupture of the splenic capsule caused by trauma or during splenectomy leads to the formation of splenic implants. This can occur as a result of direct contact of the released spleen parenchyma with well-vascularized tissue (as is the case within the abdominal cavity) or through the spread of spleen cells through the blood vessels to distant organs [2, 11, 13].

Typically, splenic implants are located within the abdominal cavity. Mostly on the surface of the small intestine, colon, greater omentum, mesentery, in the lodge after splenectomy or radical nephrectomy, under the diaphragm, on the parietal peritoneum, especially at the site of port for laparoscopy, and even in the mesentery of the appendix [17]. Splenosis may also occur in the pelvis or subcutaneously, especially at the site of surgical access [3, 13, 18]. There have been reports of thoracic splenosis, both with and without diaphragm rupture [1, 15]. The spread by blood vessels explains the cases of splenosis in the liver parenchyma [18–21], pancreas [4], mammary gland, and even the brain [11, 12]. In the described case, the location of the lesions is typical and includes the greater omentum, intestinal wall and its mesentery, parietal peritoneum, and pelvis.

In most cases, splenosis is asymptomatic and can be detected during surgery or medical imaging unrelated to its occurrence [1–4]. Splenic implants localized in the abdominal cavity can cause intestinal obstruction and related signs and symptoms such as abdominal pain, nausea, vomiting, constipation, and fever [2, 9–12, 16]. Appendicitis or hydronephrosis due to pressure on the appendix or ureters also can occur [17]. During routine physical examination a tumour in the abdomen can be palpated – it can be asymptomatic [14, 19, 20] or it can cause abdominal pain [18, 21, 22]. Location in the thorax may cause haemoptysis and pleural pain [2, 13, 15]. Another manifestation may be the recurrence of the haematological abnormalities due to which the splenectomy was performed. In such cases, splenosis should be differentiated with missed accessory spleen [2, 13, 15]. Depending on the location of the lesions and the history of the disease, splenosis should be differentiated from tumours of the liver, pancreas, or chest, but also from lymphomas, endometriosis [9, 18], metastases of a diagnosed neoplastic disease [4] or metastases of unknown origin [3], or tumour recurrence after resection [1]. On admission, our patient reported non-specific symptoms, such as headache, asthenia, nausea, and photophobia. Most of the complaints were not caused by splenosis or increased haemolysis in the course of spherocytosis, which suggests that splenosis was accidentally diagnosed.

In addition, after splenectomy, there was a suspicion of irritable bowel syndrome. The symptoms indicating

this disease might be caused by splenosis. Interestingly, the abdominal and pelvic ultrasound performed in March 2021 did not reveal pathological structures. The clinical course we report is similar to other cases in which tumours in the abdominal cavity [3, 16, 19, 20] or enlarged lymph nodes were detected during hospitalization or routine examinations [22].

For diagnosis, tests such as computed tomography and magnetic resonance imaging are used. These procedures enable the localization and assessment of the extent of the lesion. However, on their basis, its nature cannot be unequivocally determined [1, 14]. Scintigraphy with sensitive technetium-99 m-labelled heat-denatured red blood cells (Tc-99 m-DRBC) additionally enables non-invasive differentiation from tumours. Tc-99 m-DRBC is a gold standard in the diagnosis of splenosis. Unfortunately, this method is difficult to access in routine diagnostics. In doubtful cases, a biopsy of the lesion or laparoscopic sampling for microscopic examination is necessary [20–22]. In this patient, due to the ambiguous results of imaging tests suggesting neoplastic disease, the final diagnosis was made after laparoscopic surgery, and it was based on the histopathological examination of the lesions. Treatment should only be performed in symptomatic individuals, and it varies slightly depending on the clinical picture. The treatment consists mainly of surgical removal of the lesions that cause the obstruction together with the necrotic intestine [9, 12, 16]. Similarly, when haematological disorders recur, foci of splenosis are removed, e.g. by omentectomy or resection of a part of the intestine with the lesion [2, 13, 15]. Surgical removal of the lesions should not be undertaken in patients with asymptomatic clinical course, who were diagnosed accidentally [19, 21, 22].

Considering that the suspicion of irritable bowel syndrome may be doubtful, and that gastrointestinal complaints are organic, treatment in the future may be difficult. The number and spread of splenosis foci exclude the possibility of effective surgical treatment.

CONCLUSIONS

Splenosis should be considered in the differential diagnosis of disseminated abdominal lesions in paediatric patients with medical history of spleen injury or splenectomy due to haematological disease.

DISCLOSURE

The authors declare no conflict of interest.

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