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Paediatric splenic littoral cell angioma: a rare benign surgical finding

Naczyniak z komórek brzeżnych śledziony u dziecka – rzadkie łagodne rozpoznanie chirurgiczne

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Abstract

Littoral cell angioma is a rare primary vascular tumour of the spleen arising from littoral cells lining the venous sinuses of the red pulp. It is exceptionally uncommon in children and may be difficult to distinguish preoperatively from other benign or malignant splenic lesions. The article presents the case of an 8-year-old boy in whom a cystic splenic lesion was detected incidentally during abdominal ultrasound. Due to the uncertain nature of the lesion, the patient remained under clinical and imaging surveillance for more than two years. Eventually, laparoscopic hemisplenectomy was performed. Histopathological examination established the final diagnosis of littoral cell angioma. This case highlights the diagnostic difficulty of paediatric splenic lesions and the role of surgical treatment in achieving definitive diagnosis and management.

Keywords: child, spleen, littoral cell angioma, splenic neoplasms

Streszczenie

Naczyniak z komórek brzeżnych to rzadki pierwotny nowotwór naczyniowy śledziony, wywodzący się z komórek brzegowych wyścielających zatoki żylne czerwonej miazgi. W populacji pediatrycznej występuje niezwykle rzadko i może być trudny do odróżnienia przedoperacyjnie od innych łagodnych lub złośliwych zmian śledziony. W artykule przedstawiono przypadek 8-letniego chłopca, u którego torbielowatą zmianę w śledzionie wykryto przypadkowo podczas badania ultrasonograficznego jamy brzusznej. Ze względu na niejasny charakter zmiany pacjent pozostawał pod obserwacją kliniczną i obrazową przez ponad dwa lata. Ostatecznie wykonano laparoskopową hemisplenektomię. Badanie histopatologiczne pozwoliło ustalić ostateczne rozpoznanie naczyniaka z komórek brzeżnych. Przypadek ten podkreśla trudności diagnostyczne związane z pediatrycznymi zmianami śledziony i rolę leczenia chirurgicznego w uzyskaniu ostatecznego rozpoznania i postępowania terapeutycznego.

Słowa kluczowe: dziecko, śledziona, naczyniak komórek brzeżnych śledziony, guzy śledziony

INTRODUCTION

Primary tumours of the spleen are uncommon in children and are most often benign, with cysts, vascular malformations, and hamartomas representing the majority of cases[1]. Littoral cell angioma (LCA) is a particularly rare vascular tumour arising from littoral cells lining the venous sinuses of the splenic red pulp. It exhibits both endothelial and histiocytic differentiation and has been reported predominantly in adults, whereas paediatric cases remain exceptionally rare[2,3].

The clinical manifestations and imaging features of LCA are nonspecific and frequently resemble those of other benign splenic lesions, including epithelial cysts and haemangiomas[4,5]. Consequently, establishing a correct preoperative diagnosis is difficult, and in most cases the diagnosis is made only after splenectomy or partial splenic resection, based on histopathological evaluation[2,6,7].

Only a limited number of paediatric cases have been described to date, and the available data remain fragmented. Reported cases demonstrate considerable heterogeneity in clinical presentation, imaging findings, haematological abnormalities, and surgical management. While some children remain asymptomatic and are diagnosed incidentally, others present with abdominal pain, splenomegaly, anaemia, thrombocytopaenia, or pancytopaenia[3,8,9]. Due to the rarity of the disease and the absence of characteristic radiological features, differentiation from other benign and malignant splenic lesions remains difficult prior to surgery.

A case is presented of an 8-year-old child with a long-standing splenic lesion that was initially considered a cystic malformation and was ultimately diagnosed as littoral cell angioma following laparoscopic hemisplenectomy. This case illustrates the diagnostic challenges posed by benign splenic lesions in the paediatric population and highlights the crucial role of histopathological examination in establishing a definitive diagnosis.

CASE REPORT

An 8-year-old boy was admitted to the Department of Paediatric Surgery at the Medical University of Warsaw Hospital for elective surgery of a cystic lesion located in the lower splenic pole. The lesion was incidentally discovered in January 2023 during an abdominal ultrasonographic examination performed due to suspected umbilical hernia. It was described as a hypoechogenic, predominantly solid mass measuring 46 × 31 × 41 mm in the left adrenal region. At that time, the patient had experienced two infectious episodes between November and December 2022, during which he reported knee pain and abdominal pain, without additional symptoms. Laboratory tests revealed the following values: white blood cell count 2.77 × 10^9/L, haemoglobin 13.1 g/dL, and platelet count 179 × 10^9/L. To verify the ultrasonographic findings, a computed tomography (CT) scan was performed, revealing a multilocular cystic lesion

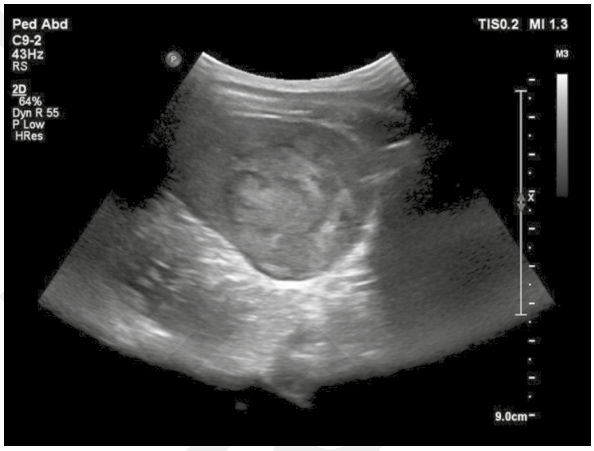

Fig. 1. Grayscale abdominal ultrasonography showing a hypoechoic, predominantly solid lesion within the spleen

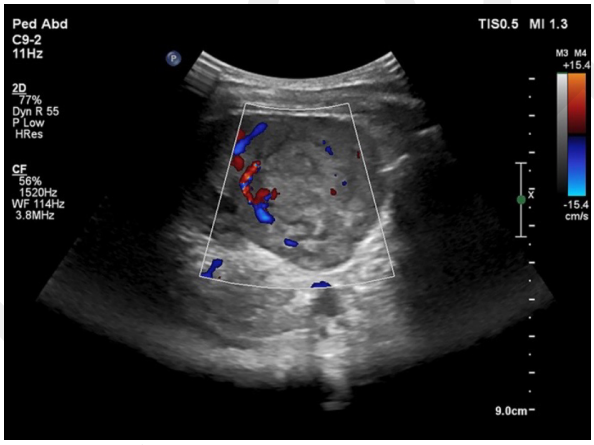

Fig. 2. Doppler ultrasonography demonstrating vascular flow within the splenic lesion

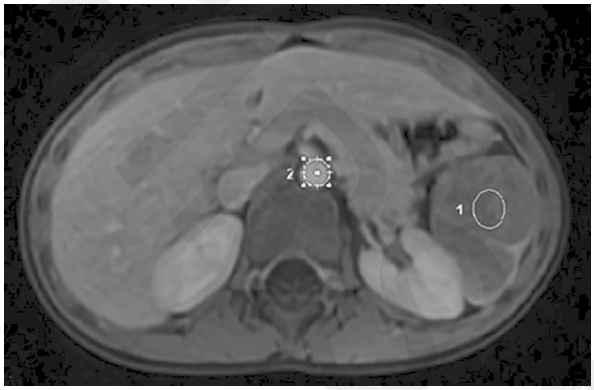

Fig. 3. Magnetic resonance imaging of the spleen in the sagittal plane (April 2024), showing the splenic lesion

in the lower pole of the spleen measuring approximately 40 × 30 mm. Based on this examination, malignancy was excluded, and the patient was referred for outpatient follow-up. In April 2023, follow-up abdominal ultrasonography showed that although the size of the lesion had not changed, its character had transformed from cystic to solid with moderate vascular flow (Figs. 1–2). This finding

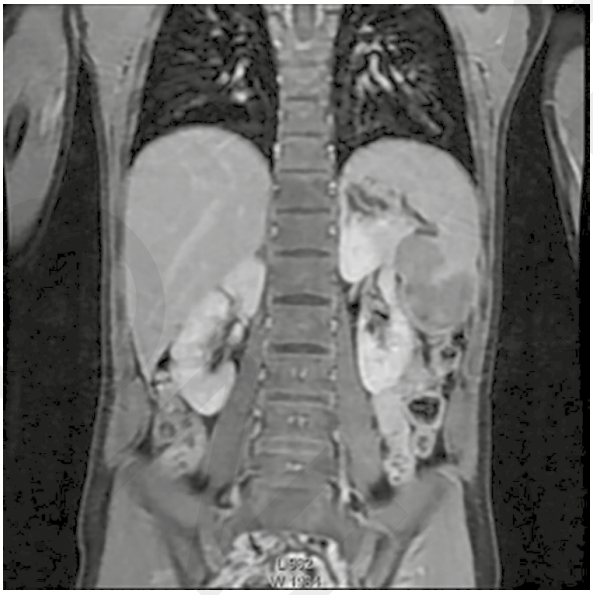

Fig. 4. Magnetic resonance imaging of the spleen in the coronal plane (April 2024), showing the splenic lesion

was confirmed by CT in September 2023. Laboratory tests at that time were within normal limits. Further abdominal ultrasonographic examinations performed in December 2023 and November 2024 demonstrated progressive growth of the lesion, measuring 57 × 38 × 52 mm in December 2023 and reaching approximately 6 cm at its greatest dimension in November 2024. Magnetic resonance imaging (MRI) performed in April 2024 demonstrated a splenic lesion measuring 31 × 51 × 38 mm in the sagittal and coronal planes (Figs. 3–4). Follow-up MRI in January 2025 confirmed further enlargement of the lesion to 53 × 58 × 43 mm, without changes in morphology (Fig. 5). Notably, the radiology report described atypical vascularisation of the lesion, raising concern regarding its biological behaviour and directly supporting qualification for surgery. The patient was subsequently reviewed at the clinic, and elective surgery was scheduled. In July 2025, the patient was admitted to the hospital for laparoscopic hemisplenectomy. On admission, he was in good general condition, with no significant abnormalities on physical examination. Laparoscopic hemisplenectomy was performed, during which the mass located in the lower pole of the spleen was completely removed with no damage to the cysts and submitted for histopathological examination. No signs of intraoperative bleeding were observed. The surgical wounds were closed with single sutures, and a 16F drain was placed in the left upper abdomen. On the third postoperative day, the drain was removed, and following an uneventful postoperative course, the patient was discharged home after six days of hospitalisation. Histopathological examination revealed multiple irregular cysts within the splenic tissue with a predominance of vascular spaces. Immunohistochemical analysis showed positive staining for CD68 and CD31, supporting the diagnosis of LCA. One month after surgery, follow-up outpatient ultrasonography revealed no residual tumour or evidence of recurrence.

DISCUSSION

Littoral cell angioma is a rare vascular tumour of the spleen, with the vast majority of reported cases occurring in adults. Occurrence in children is exceptional and usually limited to isolated case reports or small case series[3,8,9]. Because of its rarity, LCA is seldom included in the initial differential diagnosis of paediatric splenic lesions.

Review of the available paediatric literature demonstrates that LCA may occur across a broad age range, from early infancy to adolescence. Most lesions are discovered incidentally during imaging performed for unrelated indications, although symptomatic presentations associated with abdominal pain, splenomegaly, anaemia, thrombocytopaenia, or pancytopaenia have also been reported[3,8–10]. As in the present case, prolonged radiological follow-up before surgical treatment is frequently described, reflecting the difficulty in establishing a reliable preoperative diagnosis.

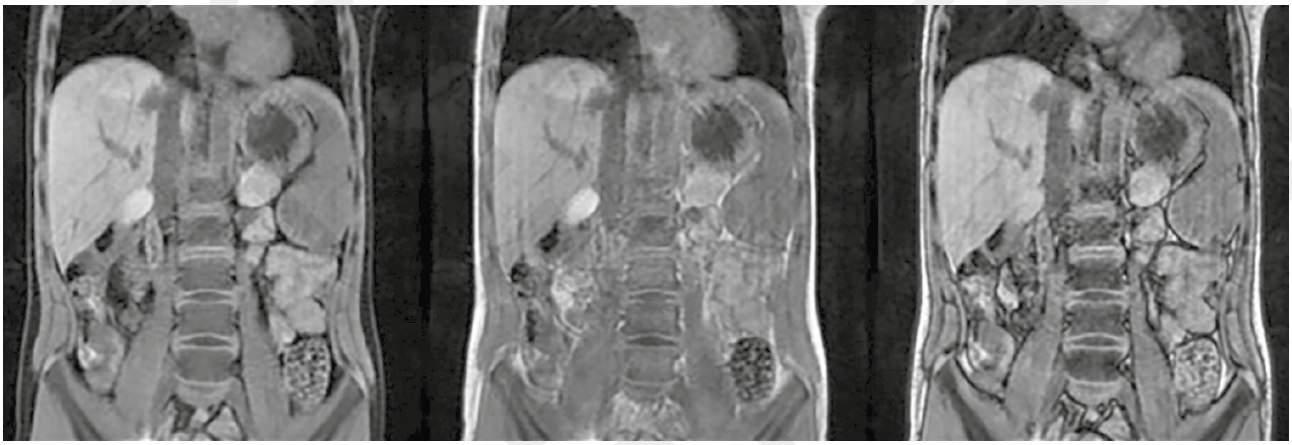

Fig. 5. Follow-up magnetic resonance imaging (January 2025), demonstrating further enlargement of the splenic lesion prior to surgical treatment

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Radiological findings in LCA are heterogeneous and lack characteristic features. Depending on the internal structure of the lesion, imaging may demonstrate cystic, solid, or mixed morphology, while Doppler ultrasonography often reveals minimal or absent vascular flow[4,5]. In the present patient, the lesion initially resembled a splenic cyst and later evolved into a predominantly solid mass with limited perfusion, illustrating how the appearance of benign splenic tumours may change over time and complicate interpretation. Previous paediatric reports have also shown considerable variability in imaging presentation. Some lesions were initially interpreted as splenic cysts, whereas others raised suspicion of haemangiomas, hamartomas, lymphangiomas, or even malignant splenic tumours[5,8,10]. In several published cases, CT and MRI findings remained inconclusive despite repeated examinations. These observations support the view that imaging alone is insufficient for accurate preoperative identification of LCA.

Even advanced imaging modalities, including CT and MRI, often fail to reliably distinguish LCA from other benign splenic lesions[4,5,11]. As demonstrated in the present case, definitive diagnosis was ultimately established only after surgical resection and histopathological evaluation, which remains the diagnostic gold standard for LCA[2,7].

The management of splenic lesions in children remains controversial, particularly when lesions appear benign and are detected incidentally. Conservative management with serial radiological surveillance is frequently advocated. However, progressive enlargement, atypical vascularisation, evolving imaging characteristics, or persistent uncertainty regarding the biological nature of the lesion may justify surgical intervention[1,12]. In such situations, surgery serves both diagnostic and therapeutic purposes.

Management strategies described in the literature vary depending on lesion size, symptomatology, imaging appearance, and institutional experience. Historically, total splenectomy was considered the standard treatment for splenic vascular tumours because of concerns regarding malignancy and intraoperative bleeding risk. More recently, increasing emphasis has been placed on spleen-preserving procedures, especially in paediatric surgery, due to the immunological importance of splenic tissue and the risk of overwhelming post-splenectomy infection[13–15]. Several paediatric centres currently favour laparoscopic partial splenectomy for focal benign splenic lesions whenever technically feasible[13–15]. In the present case, progressive lesion enlargement and atypical vascularisation supported the decision to proceed with laparoscopic hemisplenectomy.

Histopathological examination remains essential for final diagnosis. Typical immunohistochemical findings reported in paediatric cases include positivity for CD31 and CD68, with variable expression of other endothelial and histiocytic markers[2,7,10]. Similar findings were observed in the present patient. Although LCA is generally regarded as a benign lesion, associations with immunologic disorders and visceral malignancies have been reported in adults, which may additionally contribute to concern during preoperative evaluation[7,10].

This case further demonstrates that apparently benign splenic lesions in children may conceal rare pathological entities. Careful follow-up, individualised therapeutic planning, and consideration of spleen-preserving surgical techniques remain essential when the biological behaviour of a splenic lesion cannot be clearly determined.

CONCLUSIONS

Littoral cell angioma is an exceptionally rare splenic tumour in children and poses a significant diagnostic challenge due to its nonspecific clinical and radiological features[3,8]. This case demonstrates that splenic lesions with a benign appearance may remain diagnostically uncertain despite extensive imaging and are frequently identified only after surgical resection. Laparoscopic partial splenectomy is a safe and effective approach that enables definitive diagnosis while preserving splenic tissue[13–15]. Awareness of littoral cell angioma should be maintained when evaluating paediatric splenic lesions, particularly those showing atypical imaging features or interval growth.

Conflict of interests

The authors do not report any financial or personal connections with other persons or organisations which might negatively affect the content of this publication and/or claim authorship rights to this publication.

Author contribution

Original concept of study; analysis and interpretation of data; writing of manuscript: JG, GDK. Collection, recording and/or compilation of data; critical review of manuscript; final approval of manuscript: JG, GDK, MW.

References

1. Silver DS, Pointer DT, Slakey DP: Solid tumors of the spleen: evaluation and management. J Am Coll Surg 2017; 224: 1104–1111.
2. Falk S, Stutte HJ, Frizzera G: Littoral cell angioma: a novel splenic vascular lesion demonstrating histiocytic differentiation. Am J Surg Pathol 1991; 15: 1023–1033.
3. Bedir R, Sehitoğlu I, Calapoğlu AS et al.: A rare case of splenic littoral cell angioma in a child. J Lab Physicians 2014; 6: 117–120.
4. Johnson C, Goyal M, Kim B et al.: Littoral cell angioma of the spleen: imaging features. Clin Imaging 2007; 31: 27–31.
5. Ertan G, Tekes A, Mitchell S et al.: Pediatric littoral cell angioma of the spleen: multimodality imaging including diffusion-weighted imaging. Pediatr Radiol 2009; 39: 1105–1109.
6. Anbardar MH, Kumar PV, Forootan HR: Littoral cell angioma of the spleen: cytological findings and review of the literature. J Cytol 2017; 34: 121–124.
7. Peckova K, Michal M, Hadravsky L et al.: Littoral cell angioma of the spleen: a study of 25 cases with confirmation of frequent association with visceral malignancies. Histopathology 2016; 69: 762–774.
8. Mou Y, Yang L, Wang J et al.: Case report and literature review: asymptomatic littoral cell angioma in a 3-year-old girl. Front Pediatr 2024; 12: 1383015.
9. Gao T, Wang X, Wei Q et al.: Littoral cell angioma of a huge spleen with peripheral blood pancytopenia in a 14-year-old boy: a case report and review of the literature. Front Pediatr 2024; 12: 1473930.
10. Bisceglia M, Sickel JZ, Giangaspero F et al.: Littoral cell angioma of the spleen: an additional report of four cases with emphasis on the association with visceral organ cancers. Tumori 1998; 84: 595–599.
11. Kaza RK, Azar S, Al-Hawary MM et al.: Primary and secondary neoplasms of the spleen. Cancer Imaging 2010; 10: 173–182.
12. Pugalenthi A, Bradley C, Gonen M et al.: Splenectomy to treat splenic lesions: an analysis of 148 cases at a cancer center. J Surg Oncol 2013; 108: 521–525.
13. Mac New HG, Fowler CL: Partial splenectomy for littoral cell angioma. J Pediatr Surg 2008; 43: 2288–2290.
14. Qingjiang C, Wang L, Chen X et al.: Laparoscopic management of benign splenic tumors in children. World J Pediatr Surg 2022; 5: e000419.
15. Wang X, Wang M, Zhang H et al.: Laparoscopic partial splenectomy is safe and effective in patients with focal benign splenic lesion. Surg Endosc 2014; 28: 3273–3278.

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