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Septic thrombophlebitis of the portal vein – a case report

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Septic thrombophlebitis of the portal venous system, also known as pylephlebitis, is a rare clinical condition with nonspecific clinical presentation, in which imaging has a crucial role in its prompt diagnosis. We report a case of a 13-year-old male who presented with fever, jaundice and hepatomegaly. Ultrasound and computed tomography revealed portal vein thrombosis, hepatomegaly, splenomegaly, and mild ascites. No primary focus of infection, underlying thrombophilia or malignancy was identified, and broad-spectrum antibiotic therapy and anticoagulation were initiated, resulting in clinical improvement and partial recanalization of the portal vein. Clinical and imaging follow-up should be maintained to exclude the development of portal hypertension.

Keywords: LIVER; PORTAL VEIN; THROMBOSIS; MULTIDETECTOR COMPUTED TOMOGRAPHY

INTRODUCTION

Septic thrombophlebitis of the portal venous system, also known as pylephlebitis, is typically secondary to infection of the structures drained by or contiguous to it (1). It is a rare condition with nonspecific clinical presentation, in which imaging has a crucial role in its diagnosis and follow-up (2).

CASE REPORT

We report a case of a 13-year-old male with no relevant background, who presented with a prolonged history of fever, mild jaundice and hepatomegaly. Laboratory workup revealed anaemia, increased C-reactive protein, unconjugated hyperbilirubinemia and elevated liver transaminases. In this setting, abdominal ultrasound (US) was requested, which revealed hepatomegaly and portal vein (PV) and superior mesenteric vein (SMV) thrombosis, further characterized by contrast-enhanced computed tomography (CECT). CECT in the portal-venous phase better depicted occlusive thrombosis of the SMV, non-occlusive thrombosis of the main trunk of the PV and occlusive thrombosis of all its segmental branches except for segment VIII. The walls of the thrombosed veins showed mural enhancement (Figure 1). Of notice, liver parenchyma of the thrombosed segments showed hyperenhancement. Although no intra-abdominal primary focus of infection was found, there were multiple mildly enlarged retroperitoneal lymph nodes with reactive morphological features. Other relevant imaging findings included a small amount of ascitic fluid in the pelvis and splenomegaly.

The patient was admitted for inpatient care with management encompassing broad-spectrum antibiotic therapy and anticoagulation, with clinical improvement. Blood cultures were negative and further workup excluded other conditions, namely intra- and extra-abdominal infections, underlying thrombophilia or malignancy. In this setting, a presumptive diagnosis of idiopathic pylephlebitis was made. Anticoagulation was instituted for six months after discharge, during which partial recanalization of the right segmental branches of the PV was possible, while the left main branch remained occluded.

DISCUSSION

Septic thrombophlebitis of the portal venous system usually begins in the small mesenteric veins, a process that then extends proximally to the larger portal veins (1). De-

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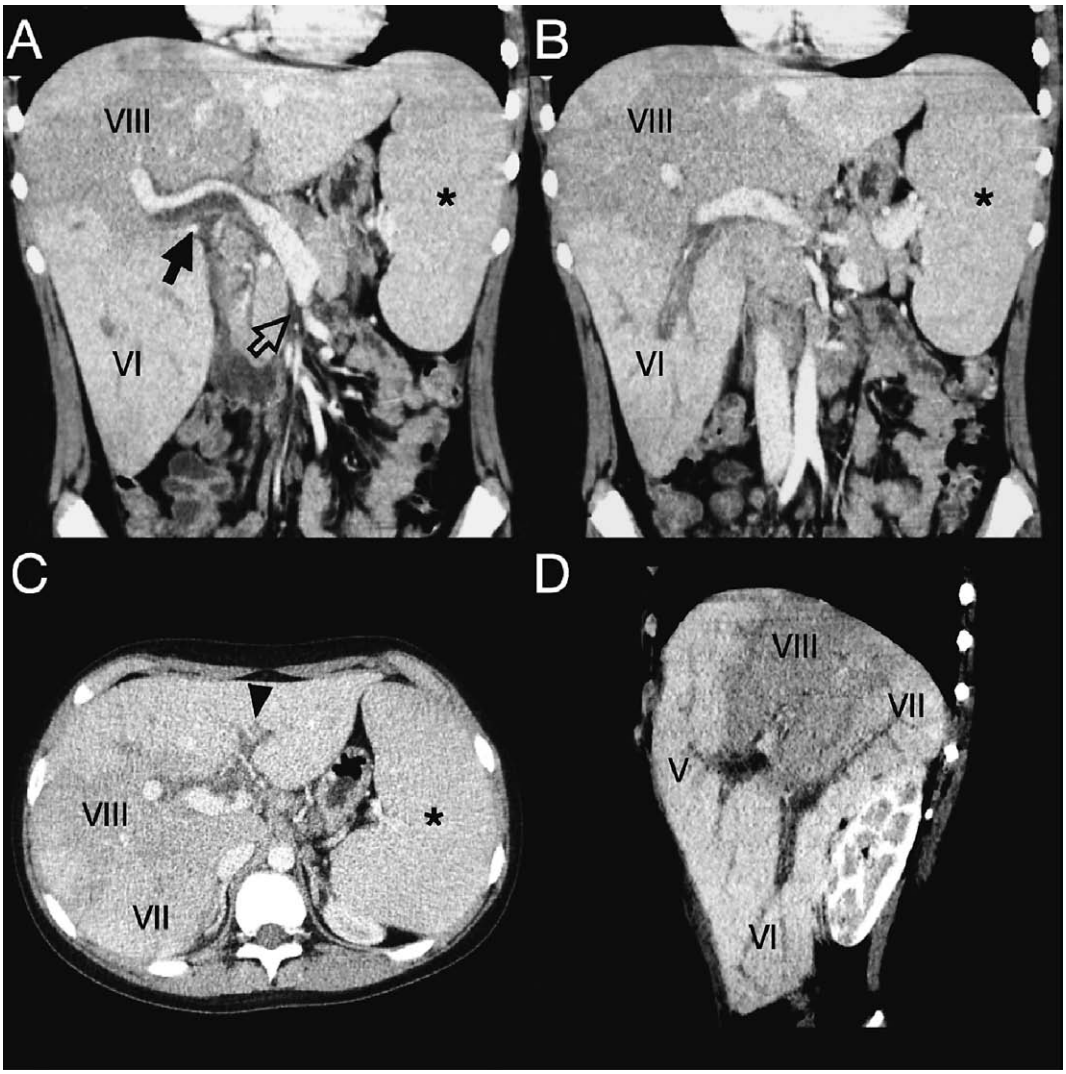

FIGURE 1. Contrast-enhanced computed tomography in the portal-venous (PV) phase demonstrating enlarged liver, with non-occlusive thrombus in the PV trunk (arrow) and occlusive thrombosis of the left PV (arrowhead) and of the branches of segments V, VI and VII. The parenchyma of the thrombosed segments shows hyperenhancement compared to normal enhancement of segment VIII, due to arterial compensatory flow. Thrombosis of the superior mesenteric vein is also partially seen (arrowhead), as well as splenomegaly (asterisk).

layed diagnosis can lead to complications such as hepatic abscesses and bowel ischemia (3).

It has an estimated incidence of 2.7 cases *per* 100 000 persons-year and a mortality rate of up to 25% (2, 4), presenting in any age group. The most common aetiology in paediatric age is appendicitis, whereas diverticulitis, pancreatitis and cholangitis are more common in adults. Nevertheless, some cases of pylephlebitis remain idiopathic, as occurred in the case presented (2). Other risk factors include hypercoagulable states, malignancy, recent abdominal surgery or patient immobility (4, 5).

Diagnosis is often delayed due to nonspecific clinical manifestations, such as abdominal pain, fever and nausea, with hepatomegaly and jaundice presenting in more advanced cases. Furthermore, symptoms referable to the primary focus infection may be minimal (2). Laboratory abnormalities are also nonspecific, including leucocytosis, elevated liver transaminase levels, anaemia and hyperbilirubinemia (6). Blood cultures are advised, although bacteraemia is not always detected, with reported positivity rates between 23% and 80% (1, 2). When positive, organisms are usually from bowel flora, most commonly *Bacteroides fragilis* and *Escherichia coli* (7).

Imaging has thus a fundamental role, with portal-venous phase CT being the modality of choice in the acute setting, detecting PV thrombosis, its complications and possible primary intra-abdominal focus of infection (2). Typical imaging findings include an enlarged vein with partial or complete filling defect by a hypoattenuating blood clot surrounded by a rim-enhancing venous wall. The affected parenchyma

usually demonstrates hyperenhancement due to arterial compensatory flow secondary to hepatic flow autoregulation. Additional complications such as hepatic abscesses and bowel ischemia should be ruled out (8). Doppler US and magnetic resonance imaging are most typically used in the follow-up of these patients.

Management includes broad-spectrum antibiotic therapy for a minimum of 4-6 weeks, narrowed according to blood culture results. While controversial, anticoagulation therapy seems to lower thrombosis recurrence and mortality rate, with a recommended duration of at least 3-6 months or longer if an underlying thrombophilic condition is identified (9). In the case reported, partial recanalization of the PV was demonstrated, nevertheless, clinical and imaging follow-up should be maintained to exclude the development of portal hypertension (10).

REFERENCES

1. Plemmons RM, Dooley DP, Longfield RN. Septic thrombophlebitis of the portal vein (pylephlebitis): diagnosis and management in the modern era. Clin Infect Dis Off Publ Infect Dis Soc Am. 1995;21:1114-20. https://doi.org/10.1093/clinids/21.5.1114.
2. Choudhry AJ, Baghdadi YMK, Amr MA, Alzghari MJ, Jenkins DH, Zielinski MD. Pylephlebitis: a review of 95 cases. J Gastrointest Surg. 2016;20:656-61. https://doi.org/10.1007/s11605-015-2875-3.
3. Vanamo K, Kiekara O. Pylephlebitis after appendicitis in a child. J Pediatr Surg. 2001;36:1574-6. https://doi.org/10.1053/jpsu.2001.27052.
4. Abraham MN, Mathiason MA, Kallies KJ, Cogbill TH, Shapiro SB. Portomesenteric venous thrombosis: a community hospital experience with 103 consecutive patients. Am J Surg. 2011;202:759-63; discussion 763-4. https://doi.org/10.1016/j.amjsurg.2011.06.039.
5. Rea JD, Jundt JP, Jamison RL. Pylephlebitis: keep it in your differential diagnosis. Am J Surg. 2010;200:e69-71. https://doi.org/10.1016/j.amjsurg.2010.02.021.
6. Kanellopoulou T, Alexopoulou A, Theodossiades G, Koskinas J, Archimandritis AJ. Pylephlebitis: an overview of non-cirrhotic cases and factors related to outcome. Scand J Infect Dis. 2010;42:804-11. https://doi.org/10.3109/00365548.2010.508464.
7. Carnish E, Garrett A, Slome M. A case of septic portal vein thrombosis (pylephlebitis). Med Forum. 2015;15. https://doi.org/10.29046/TMF.015.1.020.
8. Duran R, Denys AL, Letovanec I, Meuli RA, Schmidt S. Multidetector CT features of mesenteric vein thrombosis. RadioGraphics. 2012;32:1503-22. https://doi.org/10.1148/rg.325115100.
9. Yoon SH, Lee M-J, Jung SY, Ho IG, Kim MK. Mesenteric venous thrombosis as a complication of appendicitis in an adolescent: a case report and literature review. Medicine (Baltimore) 2019;98:e18002. https://doi.org/10.1097/MD.0000000000018002.
10. García Figueiras R, Liñares Paz M, Baleato González S, Villalba Martín C. Case 158: pylephlebitis. Radiology. 2010;255:1003. https://doi.org/10.1148/radiol.10081543.

SAŽETAK

Septični tromboflebitis portalne vene – prikaz slučaja

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Septični tromboflebitis sustava portalne vene, također poznat kao pileflebitis, rijetko je kliničko stanje s nespecifičnom kliničkom prezentacijom, gdje slikovne pretrage imaju bitnu ulogu u brzom dijagnosticiranju ovoga stanja. Opisujemo slučaj 13-godišnjega dječaka s vrućicom, žuticom i hepatomegalijom. Ultrazvuk i kompjutorska tomografija otkrili su trombozu portalne vene, hepatomegaliju, splenomegaliju i blaži ascites. Nije utvrđeno nikakvo primarno žarište infekcije, osnovna trombofilija ili malignitet. Uvedena je terapija antiobioticima širokog spektra i antikoagulacija, što je dovelo do kliničkog poboljšanja i djelomične rekanalizacije portalne vene. Ovakav slučaj zahtijeva stalno praćenje kliničkim metodama i slikovnim pretragama kako bi se isključio razvoj portalne hipertenzije.

Ključne riječi: JETRA; PORTALNA VENA; TROMBOZA; MULTIDETEKTORSKA KOMPJUTORIZIRANA TOMOGRAFIJA