Pylephlebitis with Portal Cavernoma, a Rare Complication of Appendicular Peritonitis in Pediatric Settings: About an Observation

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Abstract
Pylephlebitis is a septic thrombosis of the portal vein, most often secondary to an infectious site starting intra-abdominal. Common in adults, its occurrence in children is rare. The disease presents a low mortality, however the evolution in some cases can lead to acute complications such as acute intestinal necrosis, or chronic such as the evolution to hepatic cirrhosis.

We report a case of Pylephlebitis associated with the development of a subacute portal cavernoma, which occurred on the sixteenth day following the operative follow-up of peritonitis of appendicular origin in a 12-year-old boy. Radiological investigations made it possible to make the diagnosis and to assess the complications. The treatment required surgical revision with drainage of a residual collection from the right iliac fossa associated with medical treatment based on broad-spectrum antibiotic therapy and anticoagulation with good clinical and radiological progress.

Keywords: Pylephlebitis; Portal Cavernoma; Imaging

Introduction
Portal septic infection is a thrombosis of infectious origin of the portal vein, one of its intrahepatic branches and/or the spleno-mesenteric system. It is most often secondary to an infection starting intra-abdominal located in the territory drained by the portal system.

Technical advances in modern imaging now allow the diagnosis to be made at the onset of the disease in order to avoid the occurrence of serious consequences such as hepatic cirrhosis.

Clinical Observation
This is a 12-year-old boy who presented with pain in the right hypochondrium on the 16th day following an appendectomy postoperative following an appendicular abscess.

He comes from a family of 05 children and the interrogation did not reveal any particular medical or surgical history. The clinical examination found in a patient with good hemodynamic state pain in the right hypochondrium radiating to the umbilicus. The body temperature was estimated at 38.5 degrees.

Biologically, except for a hyperleukocytosis at 17,000 ml/mm$^3$ and a C-reactive protein at 116 mg/L, the remainder of the laboratory examination was unremarkable.

He underwent a first-line abdomino-pelvic ultrasound which revealed a non-dysmorphic liver, of homogeneous echostructure, a portal trunk increased in caliber, containing echogenic material and not taking the color coding testifying to its thrombosis. The hepatic artery was also enlarged in size but permeable (Figure 1). The abdomino-pelvic scannographic complement after injection of contrast product carried out the same day confirmed the thrombosis of the portal system by objectifying a defect of enhancement of the splenomesaric convergence extended to the portal trunk and its right and left dividing branches as well as evidence of a hepatic and perivesicular hilar collateral venous circulation reflecting a portal cavernoma (Figure 2). The scanner also objectified a collection at the level of the right iliac fossa with enhanced wall after iodine contrast with a densification of the neighboring fat and hepatosplenic pre-suppurative foci (Figure 3).

The patient underwent surgical drainage of the residual collection from the right iliac fossa combined with antibiotic therapy. Thrombosis of the portal system benefited from treatment with a low molecular weight anticoagulant secondarily relayed by an anti-vitamin K. The evolution was marked by clinical and radiological improvement.
Figure 2: Abdominal CT scan after contrast in axial A, coronal B and C view showing: A and B: Splenomesaraic convergence enhancement defect extended to the portal trunk and its right and left dividing branches (red star). C and D: Hepatic hilar collateral venous circulation (red arrow) and perivesicular (white arrow) related to a portal cavernoma.

Figure 3: Abdomino-pelvic tube after contrast injection in axial section (A, B and C) showing: A: Several hepatic lesions (red arrowhead) hypodense of millimeter size related to hepatic pre-suppurative lesions. B: lower polar splenic hypodense lesion also related to a pre-suppurative lesion (yellow arrow). C: Hypodense collection with enhanced wall after injection in connection with an abscess of the right iliac fossa (red arrows).
Intra-abdominal septic foci are the most common local cause of portal vein thrombosis [1,2]. It is a rare condition in the pediatric population and its true incidence remains difficult to establish [3]. It is often associated with a favorable pro-thrombotic terrain [4,5]. In the majority of cases, pylephlebitis is the consequence of the gradual spread of suppurative thrombophlebitis, secondary to an intra-abdominal infectious site.

The most common etiologies of this pathology are acute ascending disorders of an abdominal organ such as pancreatic necrosis, appendicitis, acute cholecystitis, acute or perforated diverticulitis [6-8]. In our case, the pylephlebitis was secondary to an abscess of appendicular origin.

Clinical signs are nonspecific, fever is almost constant, abdominal pain is frequent, with or without nausea and diarrhea [9,10]. In our case, the symptomatology was dominated by febrile right hypochondrium pain in the early postoperative after appendectomy. The diagnosis can be made in the acute phase with abdominal pain, but occlusion of the portal vein by a thrombus often goes unnoticed [3].

Faced with the non-specificity of the clinical presentation, imaging plays a fundamental role in the diagnostic approach. Ultrasound is the first means of imaging in hepato-biliary pathologies. Coupled with color doppler, it allows a good study of the gate system and its tributaries. This examination thus has very good sensitivity and very good specificity in the diagnosis of portal and mesenteric thrombosis with figures ranging from 89 to 99% reported in the literature [2,6].

The ultrasound appearance is the presence of echogenic material within an augmented portal vein and the absence of a color Doppler signal within the vein or its intra and extra hepatic branches [11,12].

The abdomino-pelvic scanner thanks to its good resolution allowing a multiplanar study offers a better study of the abdomino-pelvic cavity. It makes it possible to make a complete work-up, to establish the diagnosis, to exclude other differential diagnoses and to look for complications such as the occurrence of a portal cavernoma or signs of portal hypertension.

Early-stage CT scan confirms portal vein thrombosis as an endoluminal defect, assesses full or partial extension to the rest of the portal system. It also shows arterIALIZATION of the hepatic parenchyma downstream of the thrombus and the development following the obstruction of the portal flow of porto-portal anastomoses and the development in a few days, up to 5 weeks, of a serpiginous venous network in connection with peribiliary, perivesicular, epigastric and peri-duodenal varices [13,14]. In our case, Doppler ultrasound and abdomino-pelvic CT scan after injection of contrast product made it possible to make the diagnosis with certainty and to assess the complications.

Biologically, it is accompanied by an overt inflammatory syndrome with leukocytosis on the complete blood count with an increase in C-reactive protein. Biology can also rule out other causes that can lead to occlusion of the portal trunk, such as coagulation factors.

The main objective of the treatment of acute portal thrombosis except the main cause is to prevent the extension of the thrombosis or even to obtain a permeabilization of the latter in order to avoid the development of portal hypertension.

The recommendations at this stage are in the direction of the systematic use of anticoagulants except in the case of major contraindications. Anticoagulant treatment should be continued for a minimum of 6 months. In the absence of permeabilization beyond this, the thrombosis can be considered to be present in a chronic state.
The prognosis of septic pylephlebitis largely depends on the rapidity of diagnosis and treatment, but also on the sensitivity to antibiotics of the germ in question, the underlying condition and also the patient’s background [15,16].

In our case, the patient continued the antibiotic treatment for 10 days, anticoagulation for 06 months and regular ultrasound checks to look for complications, in particular signs of portal hypertension.

Conclusion

Acute pylephlebitis is a difficult diagnosis to make clinically, hence the interest of imaging in a suggestive clinical setting associating an intra-abdominal infectious site. In addition to its rarity in the pediatric population, the particularity of our observation is the occurrence of an acute portal cavernoma. Such a development has been rarely reported in the pediatric population.

The initiation of an early anticoagulant treatment prevents progression to portal hypertension or even hepatic cirrhosis in extreme cases.

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