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Acute pancreatitis revealing a rare form of pancreatic anomaly: Complete agenesis of the dorsal pancreas: About one case

Iken Taha*; Chetoui Ayoub; Remmal Adib; Yazghi Martah Lamyae; Nuiakh Jihan; Omari Tadlaoui Sara; Soulami Fatine; Mohammed Amine El Mouden; Ajana Rabii; Anass Ahallat; Younes Aggouri; Said Ait Laalim; Alaoui Rachidi Siham

***Corresponding Author: Iken Taha**

Radiology Department, University Hospital Tanger-Tetouan Alhoceima, Tangier, Morocco.

Email: ikentaha@gmail.com

Abstract

Agenesis of the dorsal pancreas is a rare entity that is most often revealed by its complications, notably acute or chronic pancreatitis, hyperglycaemia, and more rarely polysplenia.

We report a case of a complete agenesis of the dorsal pancreas revealed by an acute pancreatitis.

The diagnosis was made on the basis of the abdominal CT scan which showed the absence of the isthmic, corporal and caudal portion of the pancreas, only the head and the arciform process were present.

Through this case we discuss one of the main complications of corporal-caudal agenesis of the pancreas and the difficulties in the etiological diagnosis of acute pancreatitis.

Keywords

Agenesis of pancreas; Pancreatitis; Dorsal pancreas; Malformation of the pancreasis.

Abbreviations

IRM: Magnetic Resonance Imaging; ULN: Upper Limit Normal; DPA: dorsal pancreatic agenesis.

Introduction

The pancreas develops from the primitive gut through two outlines, dorsal (the most important) and ventral.

It is an exocrine but also an endocrine gland. The exocrine part is the most voluminous. It is a tubulo-acinar gland. The endocrine part is represented by islets of Langerhans disseminated in the parenchyma, predominantly in the tail.

It is a gland with an oblique direction upwards and to the left, flattened from front to back, concave towards the back, rolling up on the rachis, between the twelfth thoracic vertebra and the third lumbar vertebra [1].

Four parts are described: the head, the isthmus, the body and the tail. The head is the most complex portion. Its lower part appears pyramidal, externally based and passes behind the mesenteric vessels. The upper part is roughly trapezoidal to the left of the duodenum and to the right of the mesenteric vessels.

The pancreatic parenchyma comes in a variety of shapes, sizes, and curves. It's important to remember that the pancreas is a flat organ that abuts the posterior abdominal wall and is 4 to 8 cm tall.

The left part of the pancreaticosplenic omentum is positioned in the pancreaticosplenic omentum if the head and body are well secured by the peritoneum and its attachments. The tail of the pancreas can be more or less movable and occupy varied regions in the left hypochondrium depending on the development of this omentum and its relationship with the splenogastric omentum. In fact, any shape or form is feasible. They shouldn't cause any diagnostic difficulties.

Complete pancreatic agenesis, dorsal and ventral, is incompatible with life. A few cases have been described in association with multiple other malformations. Partial ventral, total dorsal or partial agenesis (hypoplasia) are also rare. The latter are sometimes associated with polysplenia syndrome. They may be responsible for diabetes mellitus and abdominal pain. The diagnosis is made by CT scan or MRI which show the absence of a part of the pancreas or a short dorsal pancreas.

Case Presentation

The patient was B. A, 41 years old, married, G2P2, with a history of cholecystectomy 5 years ago.

The patient consulted for transfixing epigastric pain, radiating to the back with an antalgic position in gunshot. The pain was prolonged and resistant to the usual analgesics.

The general clinical examination on admission found a conscious, pale, apyretic patient with a polypnea of 32 cycles/minute and a tachycardia of 85 beats/minute. Blood pressure was 126/84 mmHg.

Abdominal examination revealed meteorism and abdominal tenderness localized to the epigastrium and the right hypochondrium. The gynecological, cardiovascular and respiratory examinations were unremarkable.

On the paraclinical level, Lipasemia was 12 times ULN. The blood count showed a hyperleukocytosis of 10600 white blood cells per cubic millimeter. The liver workup showed ASAT at 126 IU/I, ALAT at 64 IU/I and alkaline phosphatase at 135 IU/I.

The abdominal CT scan showed the absence of the isthmic, corporal and caudal portion of the pancreas, only the head and the arciform process were present and have a normal aspect (Figures 1,2,3).

For the etiological research, the interrogation ruled out a drug or alcohol intake, the patient was cholecystectomized. The lipid and calcium levels were normal (Calcemia 105mg/l, Cholesterolemia 1.79 g/l, triglycerides 1.02 g/l, HDL 0.66 g/l, LDL 0.93 g/l).

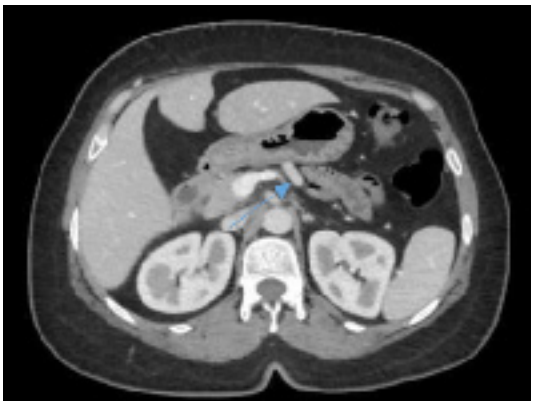

Figure 1: Axial C+ abdominal stage in portal phase showing the presence of the head of the arciform process only (arrow).

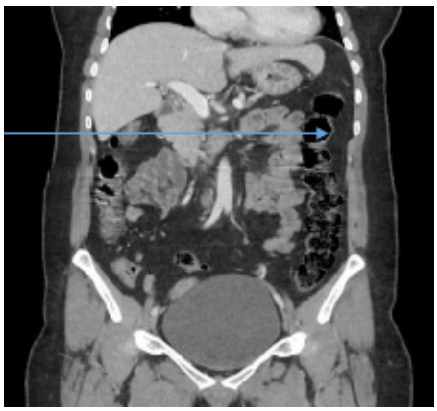

Figure 2: Coronal C+ abdominal stage in portal phase showing the presence of the head of the arciform process only.

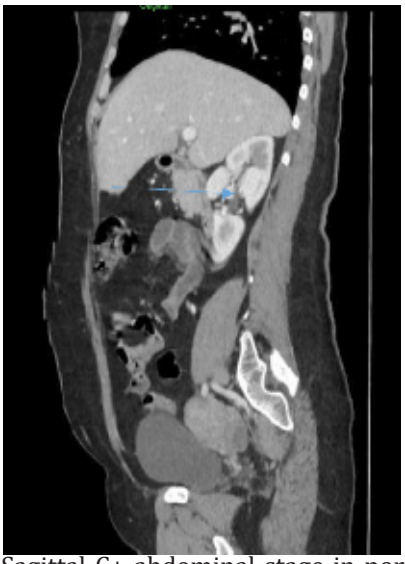

Figure 3: Sagittal C+ abdominal stage in portal phase showing the presence of the head of the arciform process only.

Discussion

Development of the pancreas begins in the fourth week of gestation. Two distinct buds arise from the duodenum. The ventral bud is born initially, followed by the dorsal bud. The ventral bud forms the majority of the head and unciform process. The dorsal bud gives rise to the upper part of the head, isthmus, body and tail. The ventral and dorsal parts fuse around the eighth week of gestation. The dorsal pancreatic duct becomes the main pancreatic duct, draining into the duodenum via the ampulla of Vater. The proximal part of the dorsal duct remains permeable and becomes the accessory pancreatic duct, drained by the minor papilla. Normally, a communicating duct develops between the 2 ductal systems. When this duct does not develop, it is called «pancreatic divisum». If there is agenesis of the dorsal pancreatic bud, a rare anomaly, the resulting absence of the body, tail and uncinate process, as well as the accessory duct of Santorini

and the minor papilla is observed. Because the islet cells reside in the tail of the pancreas, the development of diabetes has been attributed to this. Exocrine pancreatic dysfunction is rare.

The cause of acute onset abdominal pain is still undetermined. It has been hypothesized that the variant pancreatic duct system may cause insufficient drainage, resulting in acute, intermittent or chronic pancreatitis [2].

Dorsal pancreatic agenesis (DPA) is a very rare malformation that consists of the absence of the neck, body and tail of the pancreas derived from the dorsal endodermal bud. This anomaly was first described in 1911 by Schnedll and approximately 100 cases have been published to date. DPA is characterized by the absence of the pancreatic body and tail; if only the pancreatic tail is missing, it is called partial agenesis of the dorsal pancreas. If the pancreatic body is also absent and only the pancreatic head is present, it is called complete agenesis of the dorsal pancreas [2].

Some people have no symptoms, while others may develop hyperglycemia, diabetes mellitus, biliary tract obstruction, abdominal pain, pancreatitis or other conditions. It has been shown that hyperglycemia is present in about 50% of affected individuals.

The cause of agenesis of the dorsal pancreas is currently not well understood. It can occur in people with no history of the disease in the family (sporadically) and in some cases, autosomal dominant or X-linked dominant inheritance has been suggested. It has also been reported to occur in very rare conditions, including polysplenia and polysplenia/heterotaxy syndrome [3].

Because agenesis of the dorsal pancreas is considered rare and few cases have been reported in the literature, information on how the disease as a whole might be treated or managed is limited. However, there is current information on how some of the signs and symptoms associated with dorsal pancreas agenesis (such as pancreatitis) can be managed [4].

Conclusion

Dorsal pancreatic agenesis DPA is a rare type of autosomal genetic disease. Nonspecific, persistent, and unexplained symptoms such as bloating or uncontrolled blood sugar may be associated with this disease. Imaging is helpful in the diagnosis. Simple DPA does not require specific treatment, if it accompanies other diseases and physicians can make a clear diagnosis with imaging studies, patients may benefit from differential diagnosis, drug treatment (such as insulin use) and surgery.

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Authors Information: Iken Taha*[1]; Chetoui Ayoub[1]; Remmal Adib[1]; Yazghi Martah Lamyae[1]; Nuiakh Jihan[1]; Omari Tadlaoui Sara[1]; Soulami Fatine[2]; Mohammed Amine El Mouden[2]; Ajana Rabii[3]; Anass Ahallat[2]; Younes Aggouri[2]; Said Ait Laalim[2]; Alaoui Rachidi Siham[1]

[1]Radiology Department, University Hospital Tanger-Tetouan Alhoceima, Tangier, Morocco.

[2]Visceral Surgery Department, University Hospital Tanger-Tetouan Alhoceima, Tangier, Morocco.

[3]Gastroenterology Department, University Hospital Tanger-Tetouan Alhoceima, Tangier, Morocco.

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