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Interruption of The Vena Cava: A Rare And Hallmark Association In The Polysplinia Syndrome; A Case Report of A Young Girl.

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Abstract

Polysplenia syndrome (PSS) is a rare congenital anomaly, defined as the presence of multiple spleens, numbering between two and six. Polysplenia syndrome is also associated with multiple congenital variations that involve the heart, digestive tube of the abdominal cavity, or the great vessels. Fewer than 5% are still alive at five years of age due to the associated severe cardiac anomalies. There is no uniform or characteristic clinical pattern in the syndrome of polysplenia, nor is there any typical laboratory finding. The aim of this work is to highlight a rare association and hallmark of polysplenia syndrome.

Materials and Methods

reporting the case of a young girl aged 8 years, without any particular antecedents, admitted to our department for chronic abdominal pain , the clinical examination was without particularity, etiological exploration showed the fortuitous discovery of a polysplenia with incomplete situs inversus, an agenesis of the inferior supra renal vena cava with azygos continuity and interruption of the inferior vena cava with supra hepatic veins which drain directly into the right atrium.

Keywords: polysplenia syndrome, IVC abnormalities, rare anomaly.

Introduction

Polysplenia syndrome, is a congenital and rare anomaly. It incidence is 1 in 250,000 live births[1]. Fewer than 5% are still alive at five years of age due to the associated severe cardiac anomalies [2]. It was first described by Helwig in 1929 [1]. There is great variability in the definition of "polysplenia syndrome" and an often incomplete description of the clinical picture. In addition to the anomalies described in the polysplenia syndrome, other malformative anomalies associated or not with elements of heterotaxia may be encountered.Most patients with polysplenia syndrome die in the neonatal period because of the associated cardiac and biliary complications. [3].

Case report

Female child of 8 years old, without any particular pathological antecedents. Admitted in our department for chronic abdominal pain, which presented since the age of 3 years old chronic abdominal pain of intermittent evolution with epigastric burns, radiating towards the right

hypochondrium associated with regurgitation without pyrosis or vomiting. All this evolves in a context of apyrexia and conservation of the general state. The clinical examination found a child in good general condition with a weight of 32kg and a height of 135cm. The abdominal examination as well as the other organs were unremarkable.

The biological test showed no abnormality. Chest X-ray showed no dextrocardia or mediastinal enlargement, Abdominal ultrasound showed an isoechoic image of 40 mm inter hepatorenal right. An abdominal CT scan showed incomplete situs ambigus (stomach on the right and liver on the left) with polysplenia (presence of two tissue formations in the retrogastric area) without digestive duplication (fig 1) . Oesogatro-duodenal fibroscopy revealed discrete slippery hiatus hernia, congestive pangastritis of pseudo-nodular appearance at the antral level and absence of gastric volvulus. An anatomopathological examination of gastric biopsies showed a chronic antro-fungal gastritis of moderate to severe activity, follicular in nature, with moderate atrophy. Presence of Helicobacter Pylori in significant quantities and absence of intestinal metaplasia or signs of dysplasia. Patient was treated for HP gastritis. Within the framework of the polysplenia syndrome, other associated malformations have been investigated: the abdominal angioscanner showed an agenesis of the inferior vena cava supra renal with azygos continuity, no common mesentery, Transthoracic echocardiography has shown the interruption of the inferior vena cava with supra-hepatic veins draining directly into the right atrium.

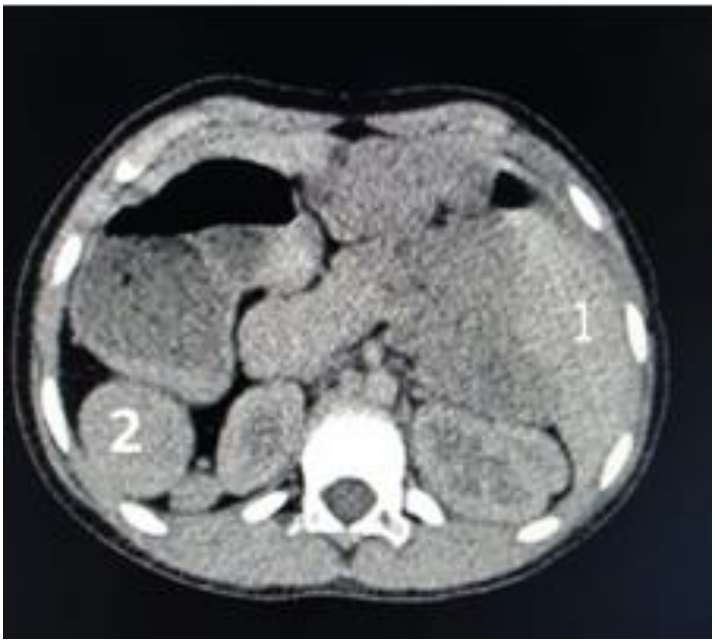

Fig 1: abdominal CT scan showed stomach inversion with polysplenia.

Discussion

Polysplenia syndrome (PSS) is a rare congenital anomaly characterized by the presence of multiple spleens numbering between two and six, associated with congenital variations that may involve the heart, the great vessels or digestive tube of the abdominal cavity [4]. In most cases diagnosed incidentally during other procedures. 5% of patients with PSS stay alive at 5 years of age due to associated severe cardiac anomalies [2]. However, some patients have anomalies in the abdominal organs or mild cardiovascular malformations which are diagnosed incidentally in adulthood [5].

In embryology, during the fifth embryonic week, the initial splenic primordia are created as incisures on the left side of the dorsal mesogastrium; when the incisures fail to fuse, they create two or more spleens [6]..

There is a genetically heterogeneous disorder, which may be associated with chromosome abnormalities such as single gene mutations, balanced translocations, microdeletions, and duplications. Several genes have been

detected in patients with heterotaxy syndrome, including CFC1, ZIC3, LEFTYA, ACVR2B. [7].

Patients with polysplenia have interruption of the inferior vena cava(IVC) with azygous or hemiazygous continuation. In that situation, venous drainage of the lower limbs is achieved through a compensatory dilated vena azygos system. These abnormalities are rarely associated with venous thrombosis, particularly in young patients. [8]. Azygous continuation of the IVC consists of the absence of the hepatic segment of the IVC with azygous continuation [8]. This abnormality is a rare clinical entity characteristic of polysplenia syndrome. [8]. Rameshbabu et al. described in 7 out of 19 cases of his own autopsied series, preduodenal portal vein which is frequently associated with polysplenia syndrome [9] Despite being polysplenic, the spleen is usually considered to be hypofunctional These patients are higher risk for infection, especially from encapsulated organisms, and may need antibiotic prophylaxis. [10].

It is important to recognize the inconstant nature of all these variations: there is no uniform or characteristic clinical pattern in the syndrome of polysplenia, nor is there any typical laboratory finding .[4], that is why the diagnosis is frequently made during surgical exploration for an associated cardiac or digestive anomaly. For other related anomalies common mesentery is found in more than 75% of cases ,biliary atresia in nearly 50% of cases, , and an abbreviated or annular pancreas in 85- 90% of cases.[11]..

Our Case demonstrates the features of polysplenia syndrome with several of the associated congenital malformations , she had an agenesis of the inferior vena cava supra renal with azygos continuity, the interruption of the inferior vena cava with supra-hepatic veins and situs inversus

Conclusion

The polysplenia syndrome is a rare disorder with a spectrum of thoracoabdominal organ rotation abnormalities that varies from patient to patient. The association of a polysplenia syndrome with interruption of the inferior vena cava is a rare and essential entity.

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