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Squamous Cell Carcinoma of the Esophagus Revealing a Polysplenia Syndrome: A Case Report

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Abstract **Case Report**

Polysplenia syndrome is a rare congenital disorder typically diagnosed incidentally during imaging procedures. This syndrome is characterized by multiple spleens along with various vascular and visceral abnormalities. Our case study centers around a 48-year-old woman presenting with a squamous cell carcinoma of the cervical esophagus who underwent a computed tomography for her staging workup revealing fortuitously a polysplenia syndrome. The patient showed good progress post-surgery and chemotherapy, with no signs of recurrence. Her polysplenia syndrome, revealed incidentally in adulthood, remained asymptomatic and required no direct therapeutic intervention. Our objective is to highlight the potential of computed tomography in diagnosing and evaluating polysplenia syndrome, and to underline the incidental detection of this rare condition in the adult population. Additionally, we aim to explore its coexistence with other complex abnormalities and the implications for treatment and prognosis.

Keywords: Polysplenia, left isomerism, IVC, azygos vein, complete common mesentery.

INTRODUCTION

Polysplenia syndrome, or left isomerism, is an uncommon congenital disorder that forms a subtype of the heterotaxy syndrome frequently identified incidentally during an imaging check-up.

The defining features of this syndrome include the existence of multiple spleens often accompanied by a complex spectrum of vascular and visceral abnormalities.

The condition, with a reported incidence of 1 in 250,000 live births, appears to be more prevalent in females [1].

This case study underlines the pivotal role of computed tomography in diagnosing and evaluating the full spectrum of abnormalities linked to this uncommon syndrome, as revealed incidentally in an adult female patient during a staging assessment of squamous cell carcinoma of the esophagus.

CASE REPORT

We report the case of a 48-year-old woman with no particular pathological history, who presented with solid food dysphagia followed by total dysphagia in a context of general health deterioration. The patient underwent an endoscopy with a biopsy that revealed a squamous cell carcinoma of the cervical esophagus. A thoraco-abdomino-pelvic CT scan was performed as part of the staging workup, which revealed no secondary locations; however, it fortuitously led to the discovery of a rare malformation association made of:

Absence of spleen replaced by multiple right nodular formations, dense, homogeneously enhanced after injection of contrast, consistent with polysplenia (Figure 1).

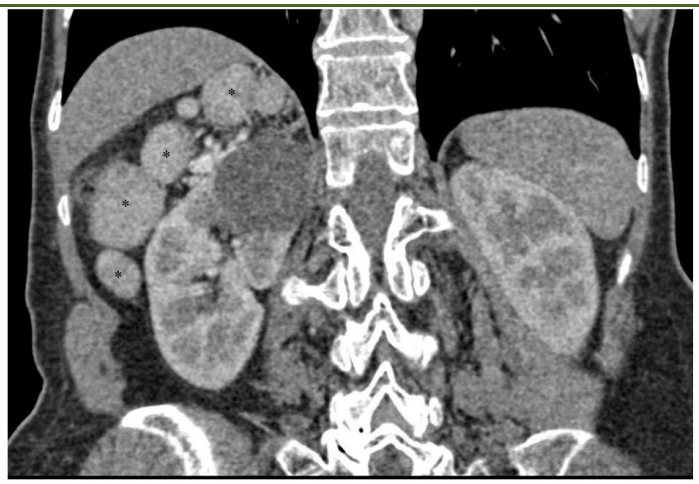

Figure 1: CT scan with coronal view showing multiple right nodular formations, dense, homogeneously enhanced after injection of contrast, consistent with polysplenia

Polysplenia was associated with hepatobiliary and pancreatic abnormalities consisting of a short pancreas (Figure 2) secondary to agenesis of its dorsal bud, a left hepatic isomerism, a median gallbladder and a right stomach (Figure 3).

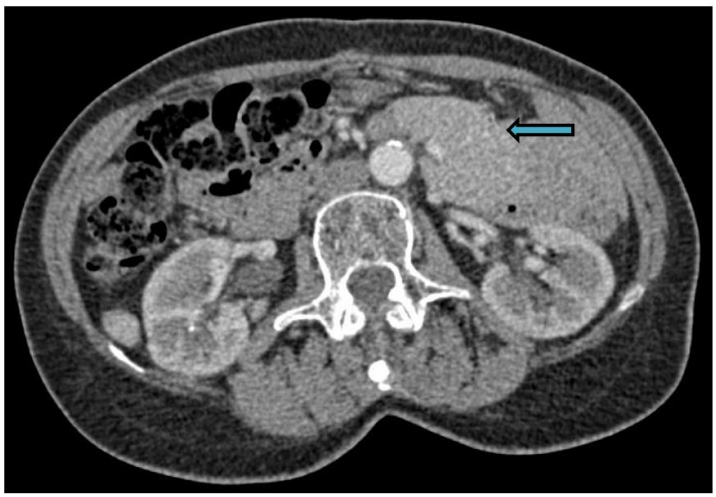

Figure 2: CT scan with an axial view showing a short pancreas with agenesis of its caudal segment

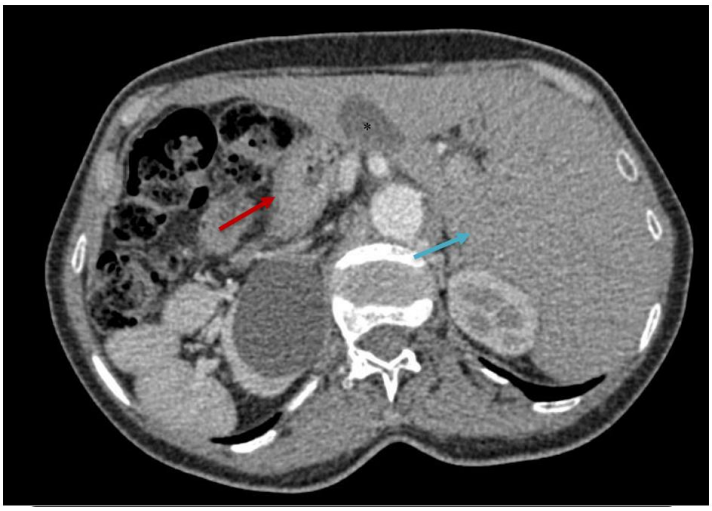

Figure 3: CT scan with an axial view showing a left hepatic isomerism (Blue arrow), a median gallbladder (*) and a right stomach (red arrow)

An abnormal rotation of the digestive tract was also found, which manifests in imaging by the disposition of the superior mesenteric artery behind and in contact with the superior mesenteric vein, consistent with a complete common mesentery (Figure 4).

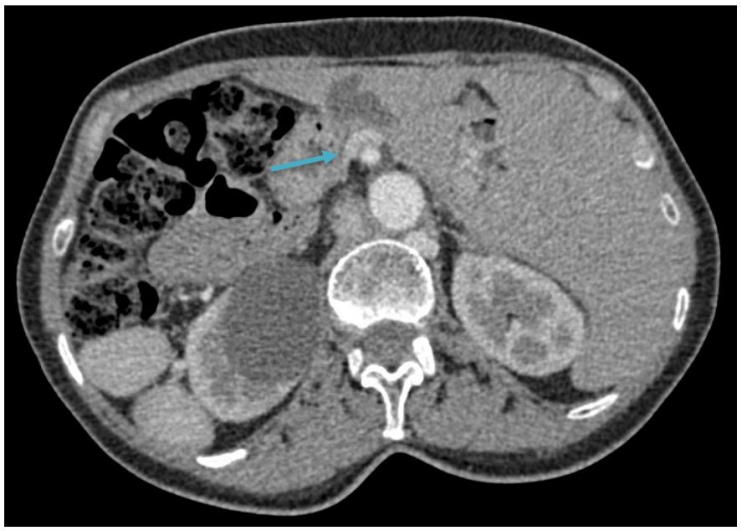

Figure 4: CT scan with an axial view showing transposition of the mesenteric vessels in favor of a complete common mesentery

There was also a vascular anomaly involving an azygos continuation of the IVC; the IVC interrupts above the renal veins with the absence of the hepatic segment of the IVC, and a diversion of the blood flow towards the large azygos vein which itself drains into the SVC (Figure 5). The hepatic veins drain directly into the right atrium.

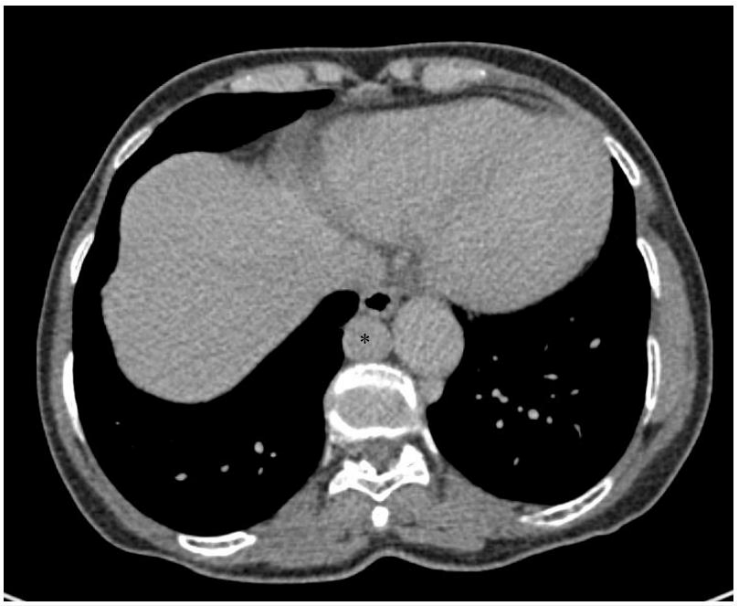

Figure 5: CT scan with an axial view showing a large azygos vein continuating the IVC

The patient underwent surgery and received chemotherapy for the carcinoma of the cervical esophagus with good progression under treatment and no signs of recurrence on follow-up examinations.

No therapeutic management was indicated for her multiple malformation syndrome, which was fortuitously revealed in adulthood and is not symptomatic.

DISCUSSION

Polysplenia Syndrome also referred to as left isomerism is a distinctive form of the heterotaxy syndrome that can be diagnosed during adulthood as well as childhood [2].

It is a rare congenital condition, first described by Peoples *et al.*, in 1781 [3] but the precise etiology of polysplenia remains undefined; although Various theories tend toward embryogenic, genetic and

teratogenic causes as possible contributors to its manifestations [2]. Furthermore, mutations have been identified in the genes of patients with heterotaxia.

This disorder is characterized by the presence of multiple spleens, typically two or more, and an ambiguous spatial arrangement of organs (situs ambigus); on rare occasions, it may be accompanied by a complete transposition of organs (situs inversus). In certain instances of polysplenia syndrome, there may be a single splenic gland, which could be either normal or multi-lobed.

Given that the splenic tissue originates in the dorsal mesogastrium during embryological development, the spleen or splenic nodules are always found along the greater curvature of the stomach, on the same side.

In addition to the presence of multiple spleens, another feature of polysplenia syndrome includes the absence of the supra-renal segment of the inferior vena cava (IVC), with azygos continuation, with the hepatic veins draining directly into the right atrium. It has also been documented that this syndrome may be associated with a range of other cardiac, pulmonary, vascular, and gastrointestinal malformation [4].

The normal pancreas results from the union of ventral and dorsal pancreatic buds, both the dorsal pancreatic bud and spleen form within the dorsal mesogastrium, thus, it is not surprising to find anomalies in both organs in patients diagnosed with PSS [5].

Clinicaly, the PSS is an asymptomatic or minimally symptomatic syndrome and there are no specific biological markers, nor is there an associated hypersplenism attributable to increased splenic volume.

Diagnostic methods, such as contrast-enhanced thoraco-abdominal computed tomography (CT), are critical for detailed evaluation of the anomalies and for devising an effective preoperative plan.

There are no specific therapeutic indications for the PSS, however, during surgery, careful exploration for associated malformations and stringent anatomical control is required to prevent intraoperative mishaps. Preoperative assessment should include thoraco-abdomino-pelvic CT scans with thoracic and abdominal CT angiography, optionally combined with MRI angiography, to ensure a comprehensive understanding of the anatomical variations at play [2].

CONCLUSION

Polysplenia Syndrome, presents as an intriguing confluence of developmental anomalies. Diagnostic modalities, such as contrast-enhanced thoraco-abdomino-pelvic CT, are paramount for detailed evaluation and devising effective preoperative strategies.

This report underscores the importance of being cognizant of Polysplenia Syndrome when evaluating patients with multiple splenic nodules and other visceral abnormalities. As our understanding of this syndrome advances, we are poised to further refine diagnostic criteria and enhance patient care for this complex, multifaceted condition.

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