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CASE REPORT

Agenesis of dorsal pancreas as incidental finding in a COVID-19 85-year-old patient: a case report with a review of literature

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Summary

Introduction: Polysplenia syndrome is a very rare congenital condition with multifactorial inheritance. It is characterized by the malposition of the thoracic and abdominal organs with or without vascular abnormalities. It comprises multiple cardiac, gastrointestinal, hepatosplenic, pancreatic and renal disorders. It is usually an incidental finding on an abdominal ultrasound or computed tomography (CT) scan performed for other reasons.

Patient review: In this case, an 85-year-old male patient is presented, who tested positive for coronavirus disease 2019 (COVID-19) infection, and he was admitted to hospital for bilateral COVID-19 pneumonia. CT scan was performed and revealed polysplenia, various vascular abnormalities, gastrointestinal malformations and agenesis of the dorsal pancreas, so heterotaxy syndrome was diagnosed. Interestingly, this patient did not have diabetes mellitus in spite of agenesis of the dorsal pancreas, neither had he ever have pancreatitis.

Conclusion: This present case shows that the quality of life in patients with polysplenia syndrome does not have to be seriously impaired and that these patients can experience old age. Moreover, the awareness and prior knowledge of anomalies included in this syndrome are crucial in order to avoid complications during surgical procedures and/or interventions.

Keywords: pancreas, polysplenia, syndrome

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INTRODUCTION

The first case of polysplenia syndrome was described in 1781 (1). Polysplenia is a very rare heterotaxy disorder with a reported incidence of 1 per 250,000 live births (2). Additionally, it is encountered in elderly even more rarely (3). Asians have a higher prevalence of heterotaxy syndrome (4) and polysplenia syndrome is more common in females (5).

In complete heterotaxy there is an exact mirror-image of all organs, whereas in partial heterotaxy only some organs are displaced (6). The synonym of heterotaxy is situs ambiguous, and it is usually accompanied by left isomerism (7). Situs ambiguous is defined as the malposition of the thoracic and abdominal organs with or without vascular abnormalities (8). It is subclassified to polysplenia (left isomerism) and asplenia (right isomerism) syndromes (8).

Unfortunately, due to severe cardiovascular abnormalities, a reported mortality for polysplenia syndrome is 75% by the age of five (3) and only those with mild anatomical abnormalities survive into adulthood. It is usually an incidental finding on an abdominal ultrasound or a CT scan performed for other reasons.

CASE REPORT

An 85-year-old male, who tested positive for coronavirus disease 2019 (COVID-19) infection, presented to our hospital with fever and cough. The patient had a past medical history of asthma and arterial hypertension. Blood tests only revealed elevated inflammatory markers. Patient's admission chest radiography demonstrated bilateral opacities throughout the lung fields.

Further evaluation implied computed tomography (CT) scan of the thorax and it showed bilateral ground glass opacities consistent with bilateral COVID-19 pneumonia, but also as incidental finding situs ambiguous was described.

CT scan demonstrated inferior vena cava (IVC) interruption with azygous continuation and suprahepatic segment of IVC drains into the right atrium via azygos continuation **(Figures 1 and 2)**.

Other vascular anomalies include left retro-aortic renal vein, arising of hepatic artery from the aorta, above the diaphragm **(Figure 3)**, arising of lienal artery from the aorta, bellow the hepatic artery and diaphragm **(Figure 4)** and preduodenal portal vein **(Figure 5)**.

Also, levocardia was described and normal atrial situs. In addition, bronchial anatomy is normal in relation to the pulmonary arteries, with eparterial (superior to the main pulmonary arteries) right and hyparterial (below the pulmonary arteries) left bronchus. What is also important, there are four nodular peripherally calcified spleen structures in the left upper quadrant of different sizes **(Figures 6 and 7)**.

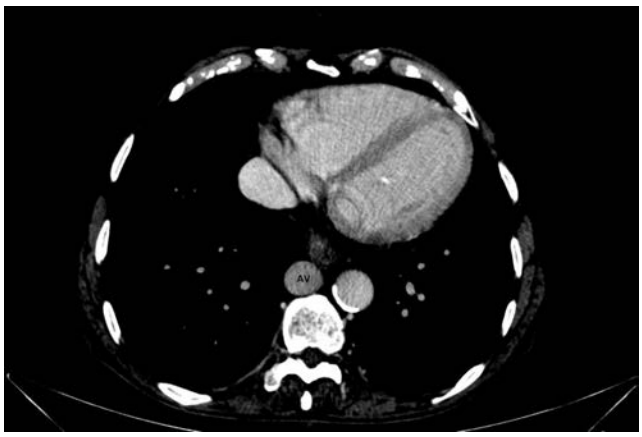

Figure 1. The abdominal computed tomography (CT) scan showing the azygos vein (AV).

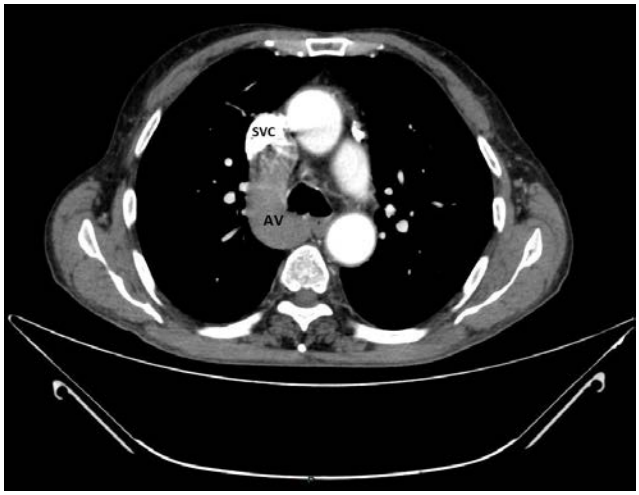

Figure 2. The abdominal computed tomography (CT) scan showing the superior vena cava (SVC) and the azygos vein (AV).

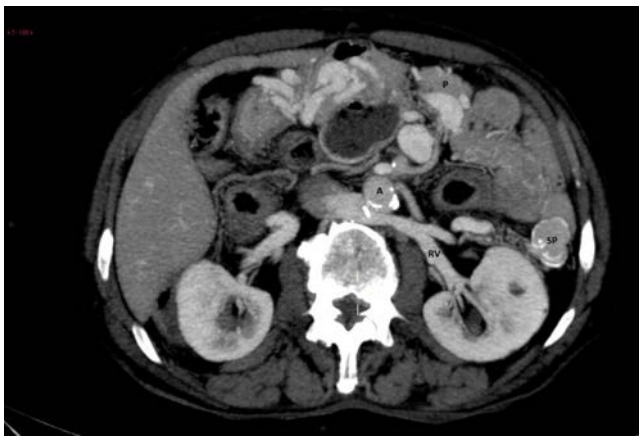

Figure 3. The abdominal computed tomography (CT) scan showing the aorta (A), the spleen (SP), the head of the pancreas (P) and the renal vein (RV).

Moreover, incomplete intestinal malrotation was seen on this CT scan. Furthermore, the stomach is in the right upper quadrant (Figure 8), whereas the liver is located on the right side, with prominent left lobe which extends up to the left hypochondrium **(Figure 9)**.

The gallbladder is located in the midline. Additionally, he was incidentally discovered to have an absent body and tail of the pancreas on CT imaging, but the head

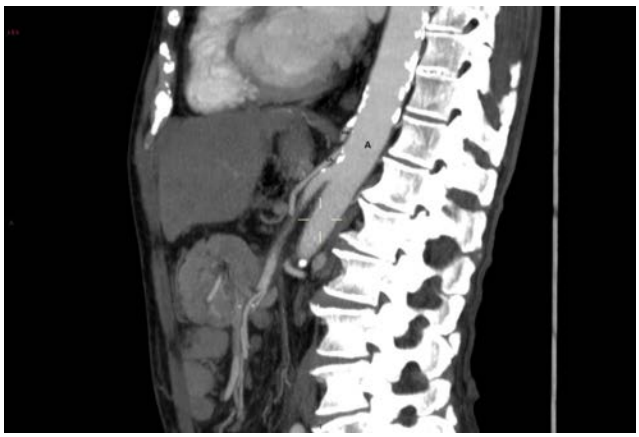

Figure 4. The abdominal computed tomography (CT) scan showing aorta (A), hepatic artery (HA) and splenic artery (LA).

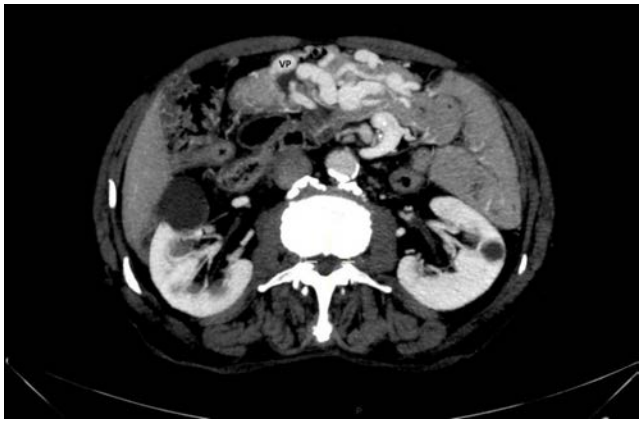

Figure 5. The abdominal computed tomography (CT) scan showing portal vein (VP).

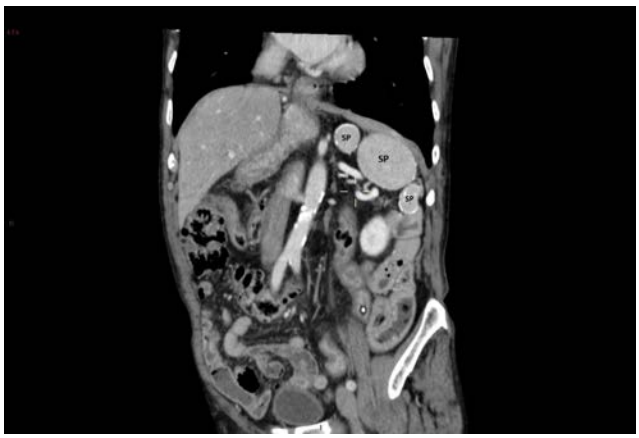

Figure 6. The abdominal computed tomography (CT) scan showing multiple spleens (SP).

of the pancreas was seen, indicating this is a congenital short pancreas defect **(Figures 3 and 7)**. Less importantly, bilateral renal cysts were described.

This morphology indicates polysplenia with agenesis of the dorsal pancreas in heterotaxy syndrome. It is important to emphasize that this patient with such important malformations has had a normal life, considering he is 85 years old. In addition, this patient doesn't have diabetes mellitus as a glycosylated hemoglobin was in normal range, in spite of agenesis of the dorsal pancreas.

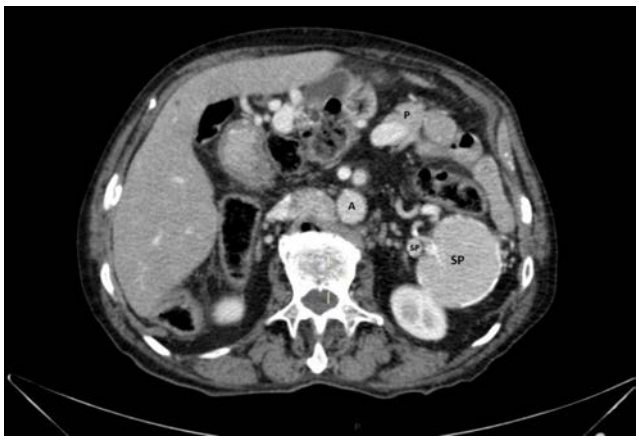

Figure 7. The abdominal computed tomography (CT) scan showing the aorta (A), the head of the pancreas (P) and spleens (SP).

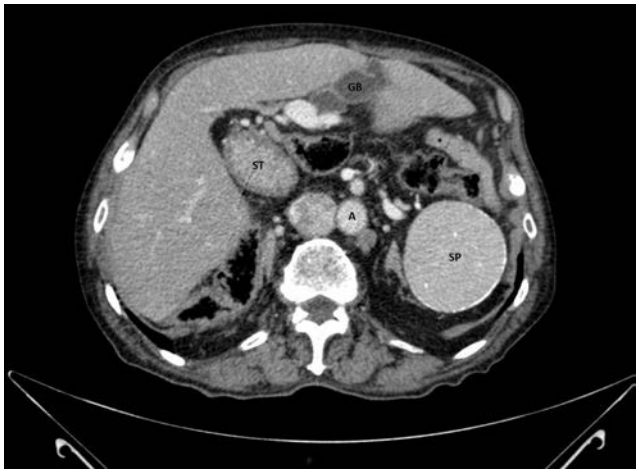

Figure 8. The abdominal computed tomography (CT) scan showing the stomach (ST), the gall bladder (GB), the aorta (A), the spleen (SP).

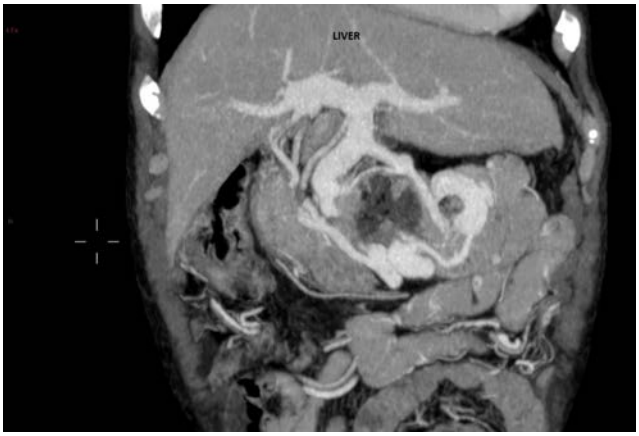

Figure 9. The abdominal computed tomography (CT) scan showing the liver.

In regard to COVID-19 infection, the patient was treated per recommendations of the National Guide for the Treatment of COVID-19 infection, which was based on the guidelines of the World Health Organization. The course of the disease was without any complications, and the patient was discharged from hospital after a few days to continue the COVID-19 treatment at home. An informed consent for this case report was obtained from the above-mentioned patient.

DISCUSSION

Although polysplenia syndrome comprises a wide range of abnormalities, there is no pathognomic abnormality that characterizes it. Abnormalities include multiple spleens and various anomalies such as visceral heterotaxia with a right-sided stomach, a left-sided or large midline liver, right-sided spleens, malrotation of the intestine, a short pancreas, and azygos or hemiazygos continuation of IVC (which is one of the most common) with the absence of the hepatic segments (3). All the spleens in this syndrome are of equal size with the main spleen in the left hypochondrium, which is vascularized by an artery with normal birth (1). What is really interesting, this syndrome can also include a single-lobed spleen or even a normal spleen (9).

The exact cause of this syndrome is still unknown, but it certainly has multifactorial inheritance. There are associations to embryonic and genetic components which are in connection with a disruption of left-right axis determination during early embryonic development and by mutations in some of the 80 genes required for normal asymmetric left-right organ development (10). There are assumptions that genetic inheritance of heterotaxy syndrome could be autosomal dominant, recessive and X-linked recessive inheritance (9).

It is described that dorsal pancreatic agenesis, the anomaly our patient has, is frequently accompanied by hyperglycemia in about 50% of cases, due to the lack of islet cells that are mainly found in the tail and body of the pancreas (11,12). Also, it brings about an increased risk of pancreatitis due to poor drainage from the remnant ventral duct (13). Splenic and pancreatic malformations develop embryonically from the dorsal bud and pancreas develops by the fusion of the ventral and dorsal pancreatic buds, so this is why polysplenia and anomalies of the pancreas, such as short pancreas or dorsal pancreas agenesis, are likely to occur together (14).

As already mentioned, this syndrome comprises multiple cardiac anomalies, but our patient has normal atrial situs and levocardia. Additionally, our patient has interruption of the IVC with azygos continuation, which is mentioned to be the most common abnormality associated with polysplenia syndrome (5). Moreover, in this present case, preduodenal portal vein was described on a CT scan, and it is another anomaly that is frequently associated with polysplenia syndrome. This anomaly can cause pressure symptoms on the duodenum and bile duct (5). It is important to emphasize that the presence of preduodenal portal vein, intestinal malrotation and vascular anomalies is very important as prior knowledge of these anomalies would help in order to avoid complications during surgical procedures and/or interventions (5).

CONCLUSION

Polysplenia syndrome is a heterotaxy disorder and it is a very rare condition. Most commonly, this syndrome is an incidental finding on an abdominal ultrasound or a CT scan performed for other reasons. It comprises multiple congenital malformations, including severe cardiovascular abnormalities and only those patients with mild anatomical abnormalities survive into adulthood. In adulthood, it is most important that both the patient and the doctor are aware of the situation in order to avoid complications during surgical procedures and/or interventions. Additionally, if dorsal pancreatic agenesis is present, we must think of very possible hyperglycemia and/or pancreatitis. This present case shows that quality of life in patients with polysplenia syndrome does not have to be seriously impaired and that these patients can experience old age.

Author Contributions: Conceptualization: Marija Branković, Tijana Gmizić, Marija Dukić, Jovana Lalatović and Davor Mrda; Investigation: Marija Branković, Tijana Gmizić, Marija Dukić, Jovana Lalatović and Davor Mrda; Writing—original draft preparation: Tijana Gmizić, Marija Dukić and Jovana Lalatović; Writing—review and editing: Marija Branković, Tijana Gmizić, Marija Dukić, Jovana Lalatović and Davor Mrda; Supervision: Marija Branković and Davor Mrda; All authors have read and agreed to the published version of the manuscript.

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AGENEZIJA DORZALNOG PANKREASA SLUČAJNO OTKRIVENA KOD 85-GODIŠNJEG PACIJENTA OBOLELOG OD KOVIDA 19: PRIKAZ SLUČAJA SA PREGLEDOM LITERATURE

Marija Branković[1,2], Tijana Gmizić[1], Marija Dukić[1], Jovana Lalatović[1], Davor Mrda[1]

Sažetak

Uvod: Sindrom polisplenije je veoma redak kongenitalni poremećaj sa multifaktorskim nasleđivanjem. Karakterizovan je malpozicijom organa u grudnom košu i trbuhu, sa ili bez abnormalnosti krvnih sudova. Uključuje više poremećaja srca, gastrointestinalnog trakta, pankreasa, jetre, slezine i bubrega. Najčešće je slučajni nalaz ehosonografije trbuha ili CT pregleda koji su bili učinjeni iz drugih razloga.

Prikaz slučaja: U ovom slučaju, prikazuje se 85 godina star muškarac, pozitivan na Kovid 19, koji je bio hospitalizovan zbog radiografski opisane obostrane upale pluća. U sklopu evaluacije bolesti, učinjen je CT pregled grudnog koša na kom je opisana obostrana pneumonija, ali uzgredno i polisplenija, nekoliko abnormalnosti krvnih sudova, malformacije gastrointestinalnog trakta i agenezija dorzalnog pankreasa, te je postavljena dijagnoza sindroma polisplenije. Uprkos ageneziji dorzalnog pankreasa, ovaj pacijent nema dijabetes melitus, niti je ikada imao pankreatitis.

Zaključak: Ovaj prikaz slučaja ukazuje na to da navedene anomalije ne moraju značajno uticati na kvalitet života pacijenata, kao i da se može doživeti duboka starost. Ipak, neophodno je dijagnostikovati ovakvog pacijenta na vreme kako bi se izbegle komplikacije tokom eventualnih hirurških procedura i/ili intervencija.

Ključne reči: pankreas, polisplenija, sindrom

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