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Incidenally finding of dextroxardia situs inversus totalis with polysplenia and Herpetic gingivostomatitis/esophagitis

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Abstract

Polysplenic syndrome with dextrocardia and situs inversus totalis has been reported primarily in children and adolescents; in adulthood they occur sporadically and randomly. Since heart defects are often associated with heart problems early in life, most patients are diagnosed in childhood. The most commonly observed developmental abnormalities include nonzygous or hemizygous continuation into the inferior vena cava, multiple spleens, and cardiac malformations. Our patient was one of the rare cases of an incidental diagnosis in adults. Imaging diagnosed gingivitis and esophagitis with blisters preventing the patient from opening the mouth and eating, multiple spleen and other visceral malformations with dextrocardia. Hopefully, the cardiac ultrasound revealed that dextrocardia was the only cardiac defect and the patient could be safely discharged from the hospital. We report the possible incidental finding of incidental polysplenia or dextrocardia and emphasize that individuals with such severe malformations can lead normal lives; Only future surgical intervention or other instrumental treatment should be considered.

Keyword : dextrocardia ,, situs inversus totalis ,, polysplenia ,, herpetic gingivostomatitis

Introduction

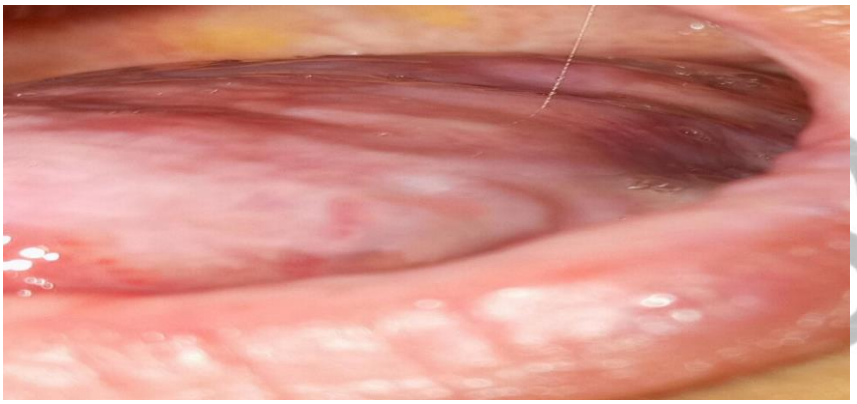
Polysplenic syndrome (PS) situs inversus totalis with dextrocardia is a rare disorder defined as the presence of two or more spleens associated with various anomalies of the thoracic or abdominal cavity that define situs inversus totalis. Situs inversus totalis dextrocardia and polysplenic syndrome are mainly described in children and adolescents, less often in adults [1]. This can be explained by its frequent association with congenital heart defects, which prevent children from reaching adulthood [2]. The other common malformations include vascular, pancreatic, and duodenal abnormalities, as well as abnormal numbers of lung lobes. In adults, PS is usually diagnosed incidentally during imaging tests while

other causes are being searched for.The procedure and prognosis depend on the type of developmental disorder. Adults are usually not prescribed any specific treatment, only information about possible further operations or other treatments. In this article, we describe a rare case of an adult with polysplenic syndrome, dextrocardia situs inversus totalis, diagnosed on computed tomography (CT) for gingivitis and esophagitis. . The primary genesis of this autosomal recessive disorder is unknown; However, it is associated with various factors including mixed twins, cocaine use and maternal diabetes. In 1600, Fabricius first described dextrocardia; However, Severino was the first to present a complete reversal of the situation [3]. The estimated incidence varies between 1 in 10,000 and 50,000 live births [3, 4]. The preferences were the same for men and women.Primary ciliary dyskinesia, a congenital disorder, can also occur in people with this condition. In addition, radiological examination is essential for determining situs inversus totalis and is related to the diseases presented in this showcase.

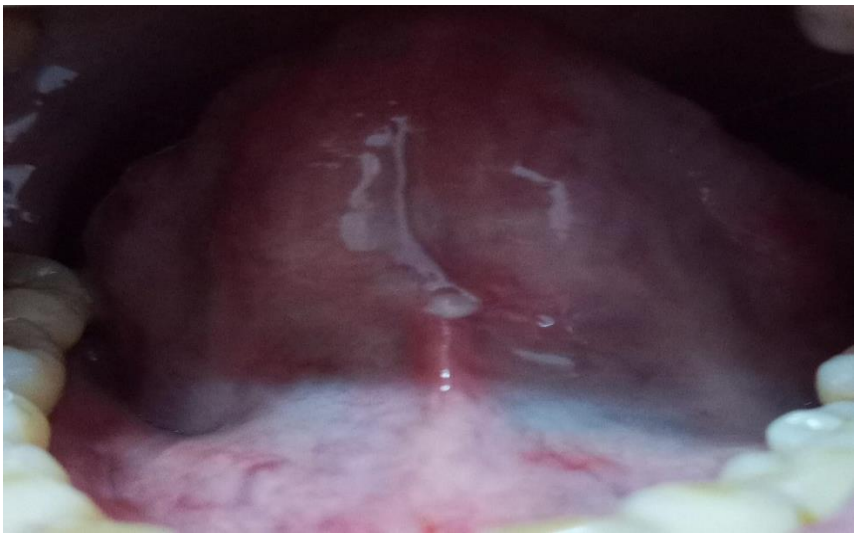
Case summary

A 43-year-old woman, married with a four-year-old son, traveled to Pakistan for a check-up with her gynecologist to try to conceive again and then underwent a tubal examination. After the procedure, the patient experienced abdominal swelling and was given medication. Ten days later, she returned to the UAE and developed a fever with blisters around her lips that spread to her mouth, tongue and the inside of her lips. For the last 2 days she has not been able to open her mouth, eat or drink. Upon arrival at our facility, the patient did not have a fever, but was unable to open her mouth, the buccal mucosa was congested, and blisters were forming. with red and white spots Incedentally , a routine chest x-ray upon admission revealed that the heart on the right side of the chest (dextrocardia). Electrocardiography confirmed dextrocardia with positive R in the AVR lead ,normal axis deviation and positive R in V1. Echocardiography confirmed dextroxardia and revealed no other cardiac anomalies. However, on echocardiography, the liver was found to be on the left side, indicating the possibility of complete situs inversion. CT of the abdomen confirmed the diagnosis of situs inversus totalis and showed (multiple spleens) on the right side. Nasal brush screening for primary ciliary dyskinetic is negative The patient stated that she had no history of sinusitis and was not taking any medications other than

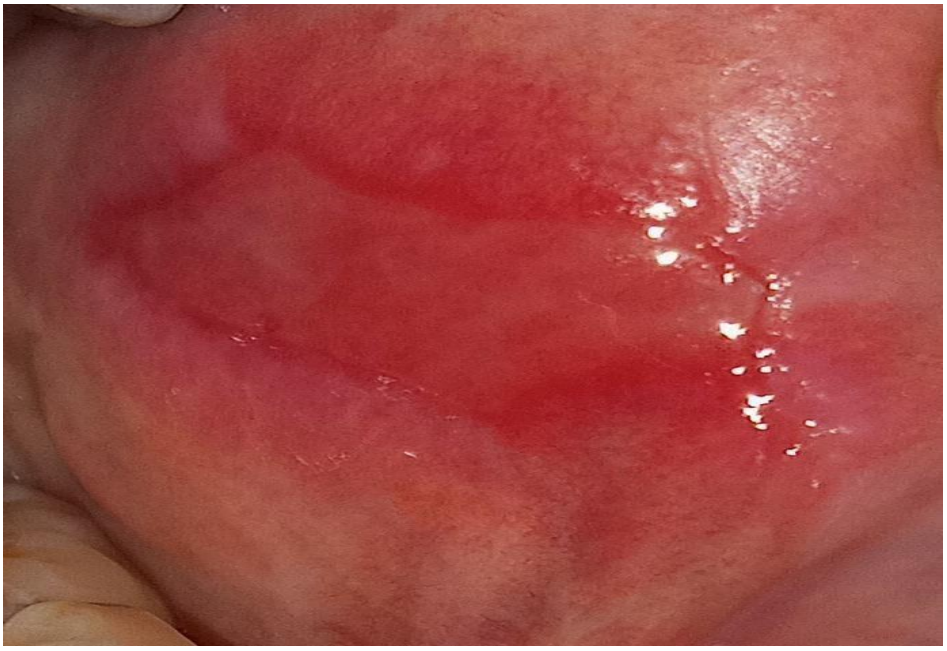
those to control her blood sugar levels. Blood tests revealed a white blood cell count of 15.78 x 10E3/µL, an erythrocyte sedimentation rate of 47 and a hemoglobin A1c of 7.8%, normal electrolytes and minerals, and normal liver and thyroid functions. Serology results were negative for HIV, hepatitis B, and hepatitis C. Urine culture was positive for yeast and white blood cells above 100/HPF and negative for nitrates. The patient was diagnosed with herpetic gingivostomatitis and esophagitis and started on acyclovir. The heart underwent echocardiography and showed dextrocardia without shunt defects, good contractility, and good systolic function.



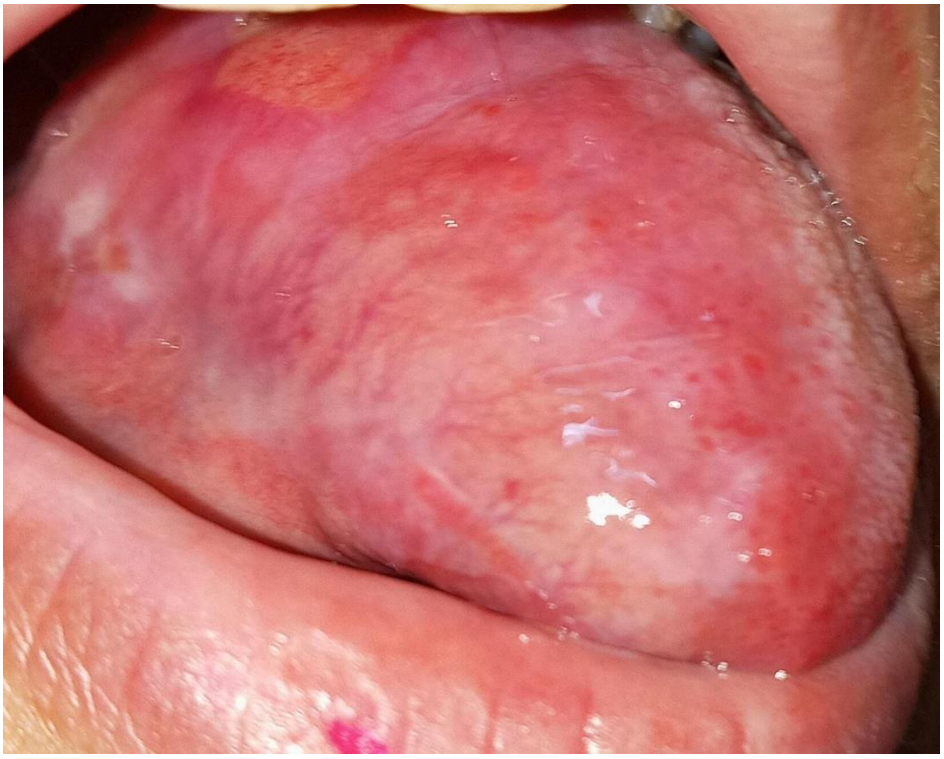
Figur 1- Tongue 1-1



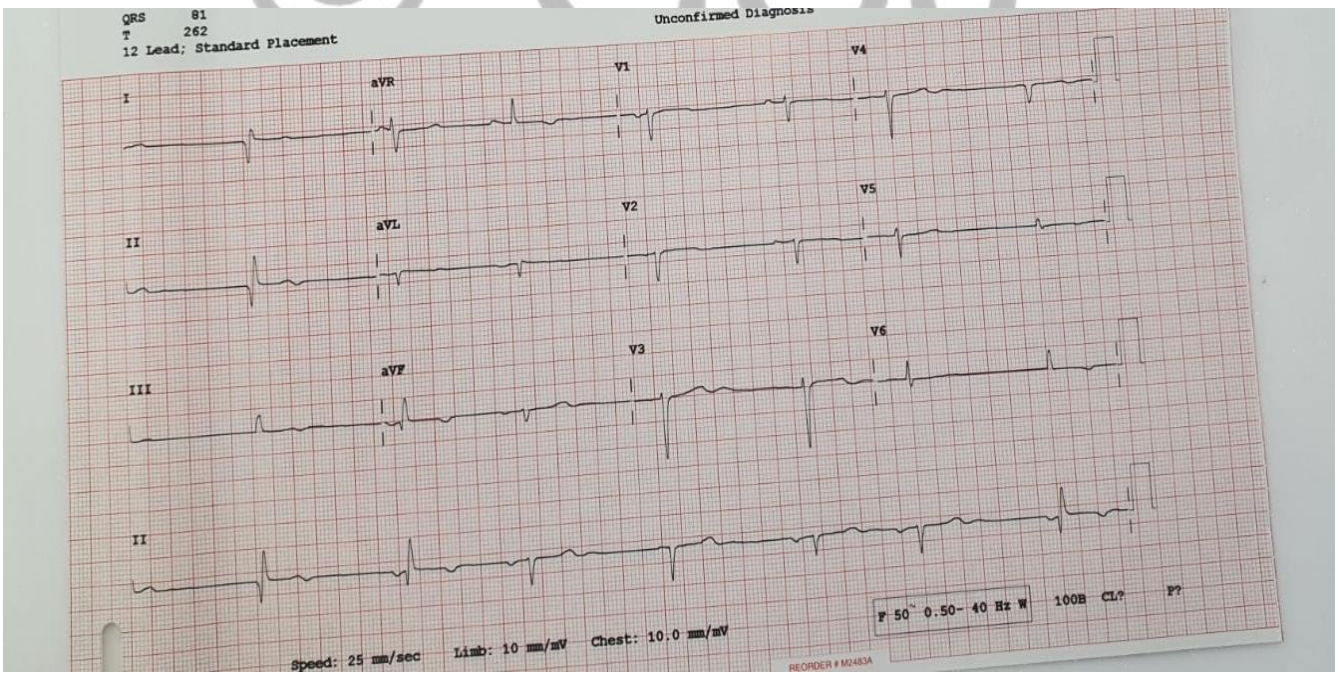
Figur 2 -Under the tongue 1-2



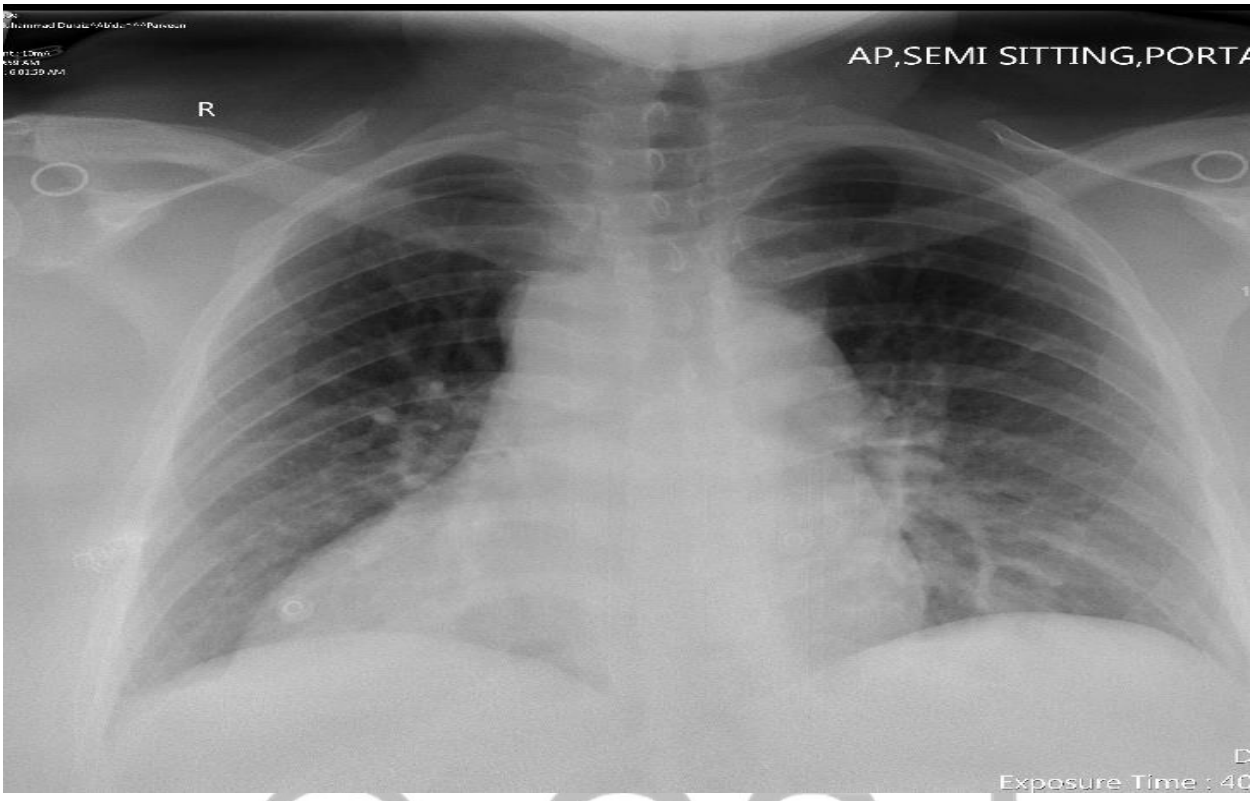
Figur 3 – Right side of the tongue 1-3



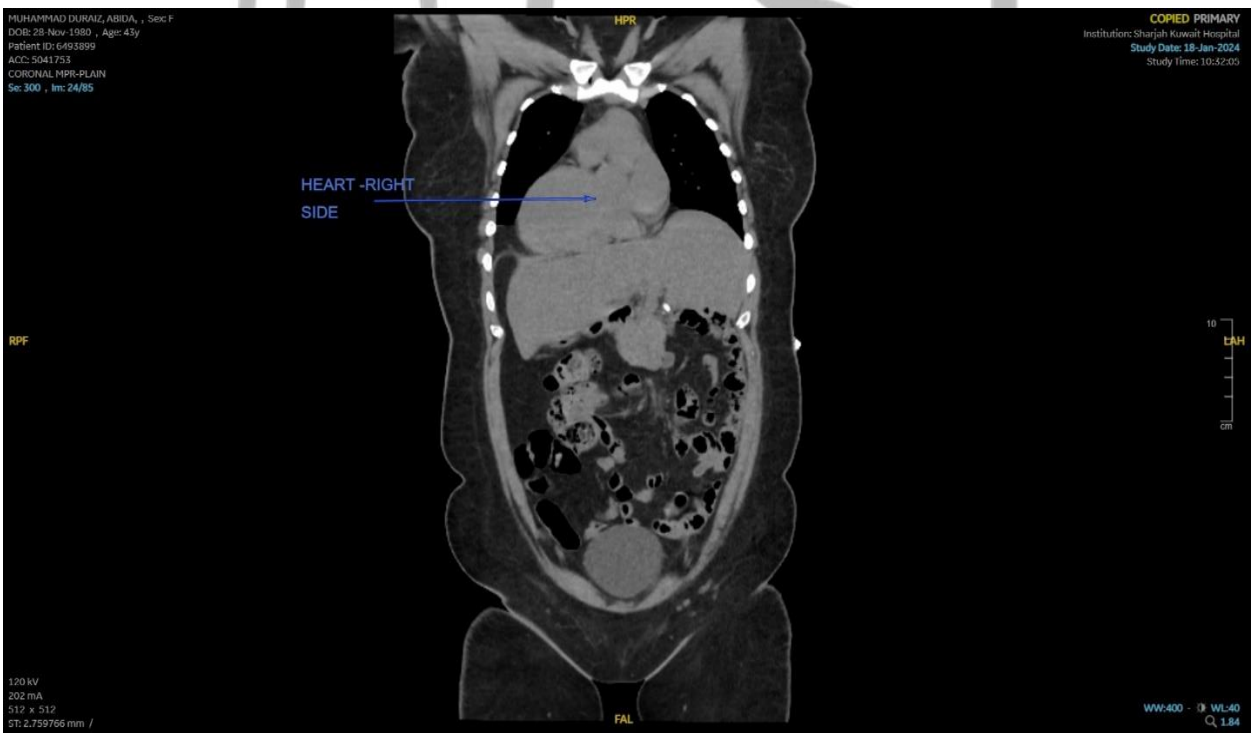
Figur 4- Left side of the tongue 1-4



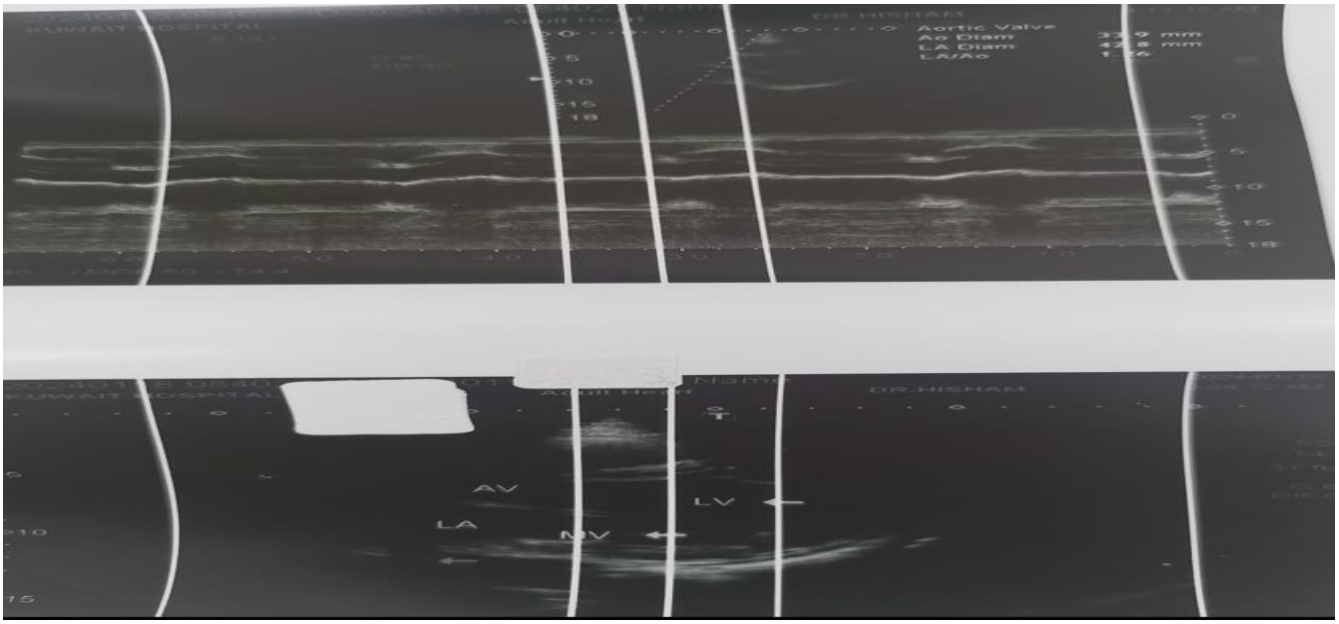
Figur 1-5 ECG: SR, normal axis,upgoing AVR



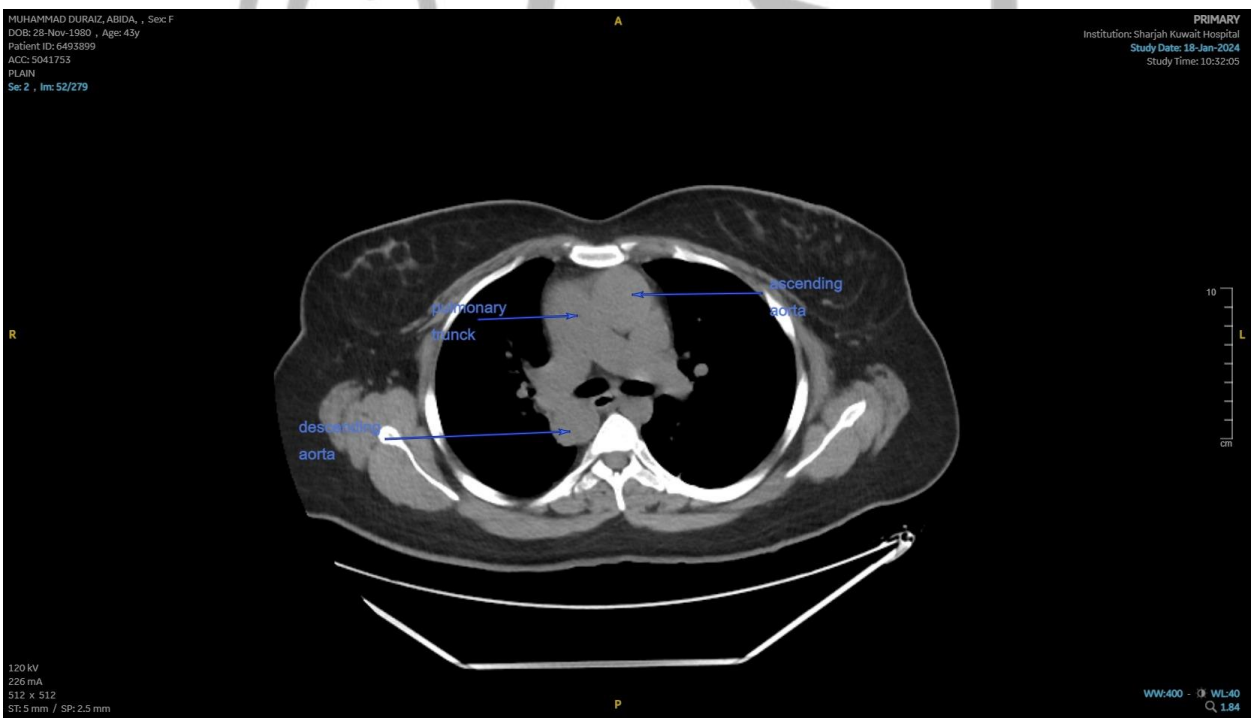
Figur 1-6 Chest x ray – dextrocardia



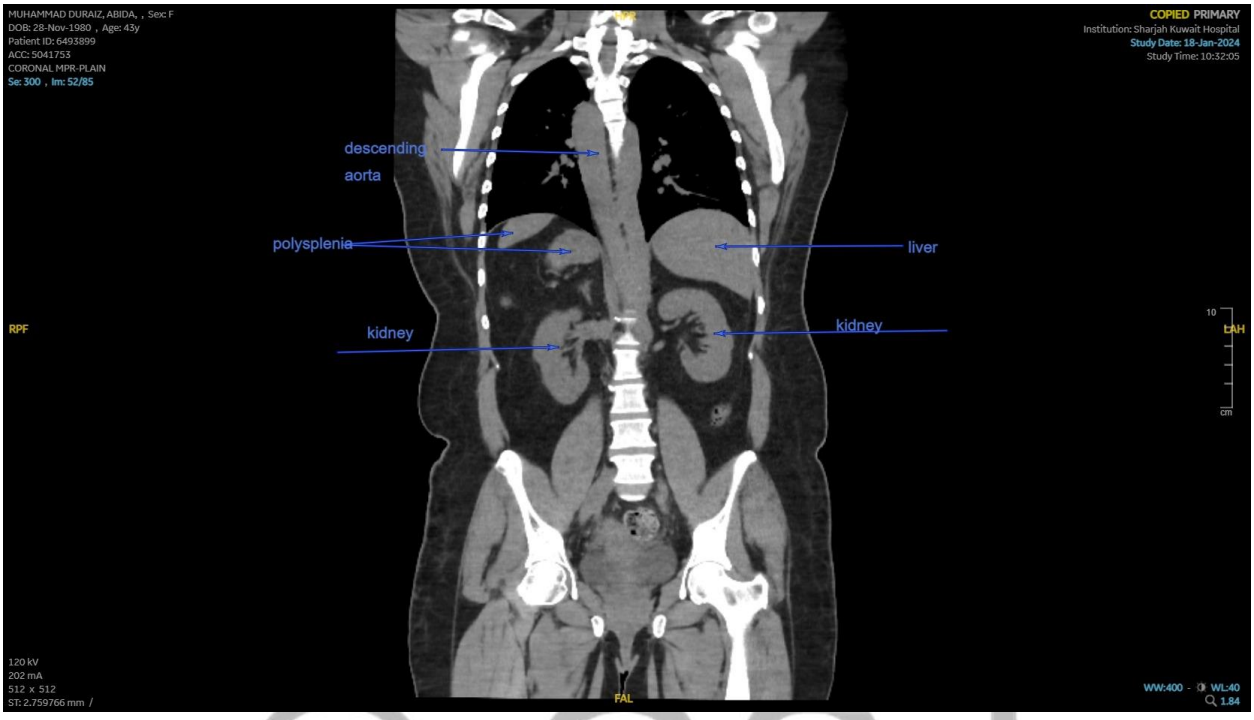
Figur 1-7 Dextrocardia via CT abd -chest



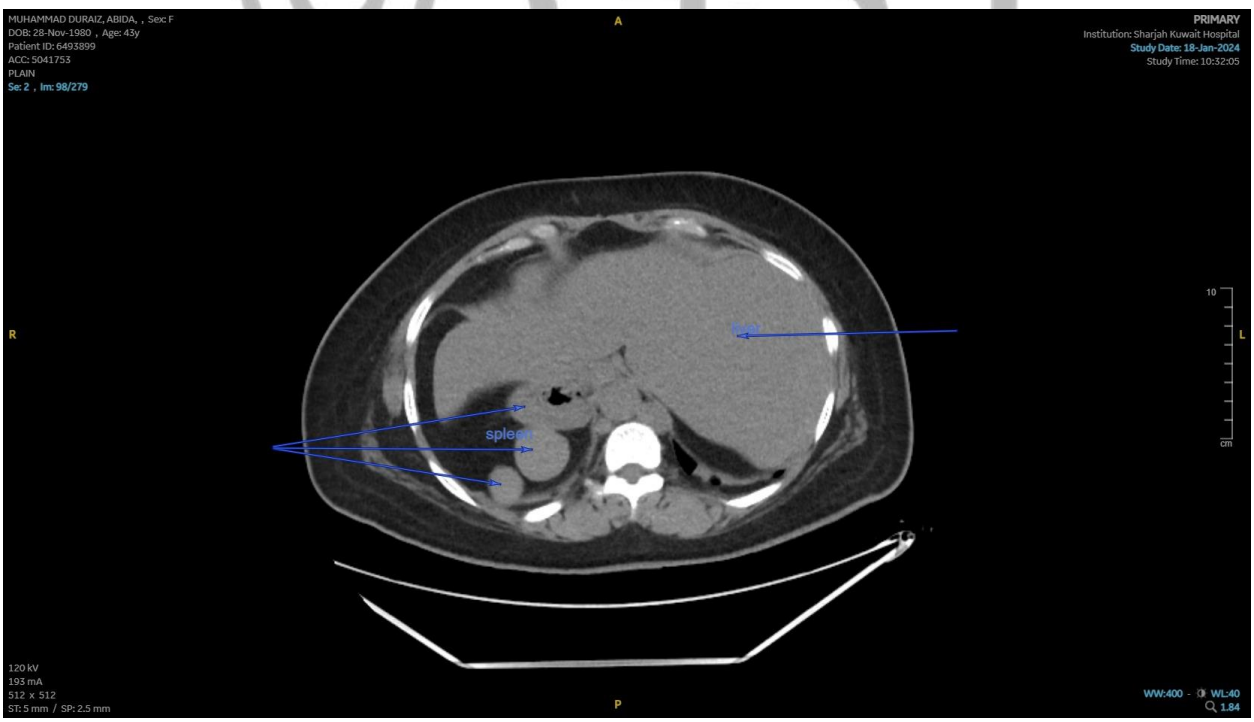
Figur 1-8 Echocardiography : dextrocardia on longtudinal long axis view (normal probe position on left lateral position



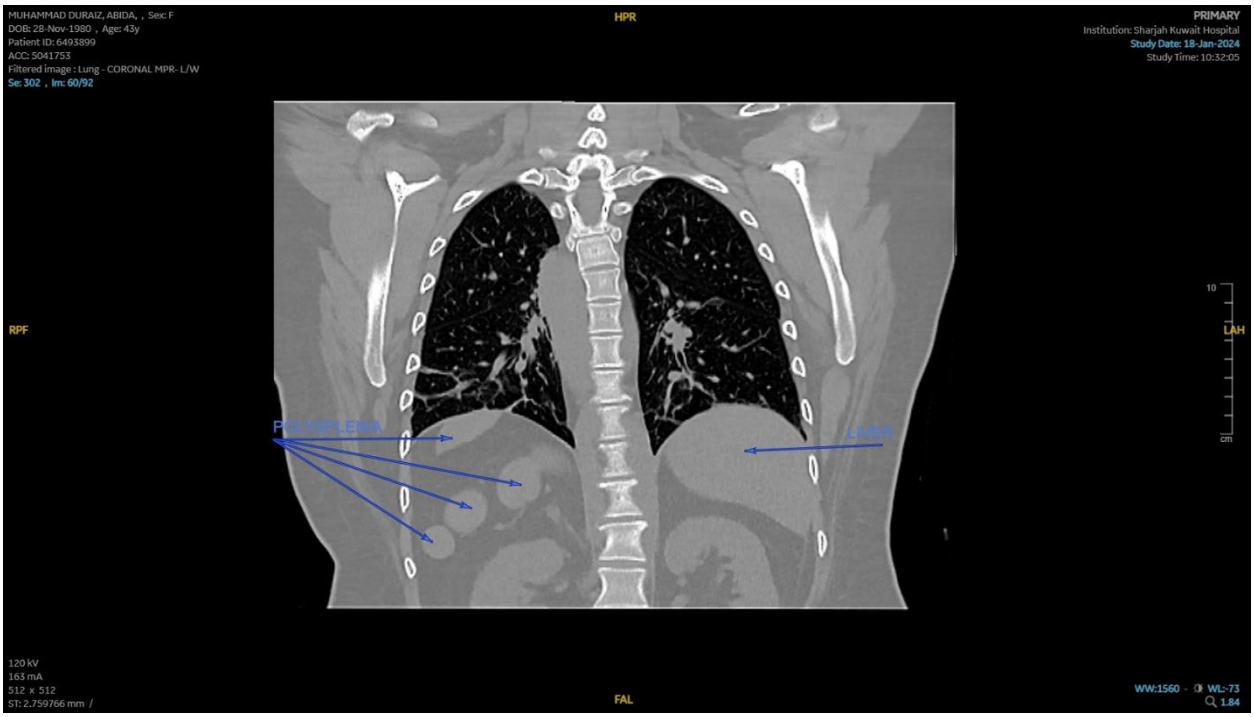
Figur 1-9 HRCT chest – Dextrocardia – right descending aorta



Figur 1-10 CT abdomin -situs inversus totalis -polysplenia, normal bilateral kidneys



Figur 1-11 Situs inversus totalis with clear multiple spleen (splenules)



Figur 1-12 Situs inversus totalis with polysplenia (multiple splenules), left liver position

Discussion :

Polysplenia syndrome is a rare congenital disorder that is generally diagnosed in early childhood owing to various severe cardiac abnormalities. Most polysplenia patients (75%) die by the age five due to severe cardiovascular anomalies (1,2). However, an isolated occurrence without severe cardiovascular anomalies may go unnoticed during early life and may be detected incidentally during radiologic examination or vascular interventions (3). The spectrum of abnormalities observed in patients with this syndrome is broad. In addition to multiple spleens, there are often cardiac anomalies, interruption of the IVC with azygos continuation, bilateral hyparterial bronchi, and abdominal heterotaxia (1,2,4). This case included polysplenia syndrome in an adult patient with situs inversus totalis,dextrocardia without interrupted IVC or azygos continuation.

“Heterotaxy” broadly refers to a wide spectrum of anomalies involving malposition and dysmorphism of the thoracoabdominal organs and vessels across

the left-right axis of the body (5,8). In a narrow sense, heterotaxy is a somewhat ambiguous entity that includes situs ambiguity and can be classified into several conditions including asplenia (right isomerism) and polysplenia (left isomerism) syndromes. Right isomerism involves bilateral trilobed lungs, bilateral eparterial bronchi, and bilateral right atria, with asplenia. Left isomerism involves bilateral bilobed lungs, bilateral hyparterial bronchi, and bilateral left atria with multiple spleens (5,8). However, our patient presented with polysplenia , situs inversus totalis and dextrocardia without definite isomerism.

Polysplenia is usually accompanied by abdominal-organ abnormalities, including renal agenesis or hypoplastic kidneys, double ureters, and a short pancreas (6). These gastrointestinal abnormalities can cause various symptoms or diseases, such as abdominal pain and pancreatitis. However, in our case, only situs inversus totalis was observed in the abdominal area. Although most of these anomalies were detected incidentally, knowledge of the presence of vascular anomalies and situs inversus could be important if the affected individual needs to undergo catheterization of vessels coursing to the right side of the heart, electrophysiological studies, cardiopulmonary bypass surgery, IVC filter placement, temporary pacing through the transfemoral route, or arterial angiography (4,7). Recent advances in MDCT technology have led to the incidental diagnosis of these complex anomalies.

In our case, numerous splenules and dextrocardia were associated with situs inversus. The kidneys appeared normal. His right aorta and left IVC showed no vascular abnormalities. Dextrocardia with situs inversus, as seen in our case, is associated with a lower incidence of congenital heart disease (10%). The reason varies depending on the associated malformation.Situs inversus is associated with asplenism, various sphenoid syndromes, ectopic kidneys, horseshoe kidneys, duodenal atresia, and various pulmonary and vascular anomalies (4).

Conclusion :

In summarize , we describe a rare case of a 57-year-old woman who presented with a viral infection causing gengipho-stomatitis and esophagitis. It was discovered incidentaly that the woman had polyslenia and dextrocardia situs inversus totalis. Insulin was used to control the patient's blood glucose levels while she received antiviral treatment in the form of oral vitamin B complex,

injectable acyclovir, and oral triamcinolone paste. The patient's condition has improved with oral antiviral acyclovir after five days of safe release at home. She was also informed about her rare condition and the information she needs to provide to her physician and surgeon in the event that she needs any future surgery.

Patient consent :

The patient provided informed consent and anonymity and confidentiality were maintained.

Acknowledgment :

We are appreciative to the patient and patient husband and individuals for giving assent for this case report to be composed

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