

A CASE REPORT- ANATOMICAL VARIATION OF SUPERIOR VENA CAVA WITH HEMIAZYGOUS VEIN DRAINING INTO LEFT SVC

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ABSTRACT-

Abnormalities of the vascular system are always of extreme interest due to its importance in circulation. Duplication of superior vena cava (SVC) is a rare anomaly with the incidence of 0.3% in general population whereas in patients with congenital heart disease it varies between 10-11%. It results from a persistent left superior vena cava, which is due to the embryological failure of regression of the anterior cardinal vein. The left sided SVC drains into the right atrium in 90% of cases via a dilated coronary sinus. Double SVC cases have special clinical importance if the one on the left side drains into the left atrium as it can cause circulatory disturbances and has been linked in cases of septic embolism it is also surgically important in the presence of congenital heart disease. Right sided SVC is seen in 82-90% of cases of double SVC. Usually if the left SVC opens into right atrium then the case is identified accidentally during any other diagnostic and therapeutic procedure as it remains asymptomatic.

AIM- To discuss the importance of cardio-vascular system and discuss the case observed.

CASE REPORT-Here we discuss a case of patient diagnosed with Hodgkin's lymphoma having double SVC where left vena cava is opening into right atrium along with hemiazygous vein opening into left SVC directly, along congenital absence of left kidney.

DISCUSSION- Cardiovascular system is one of the major systems of body and Abnormalities of the vascular system are always of extreme interest. Any abnormality in heart or vessels will lead to a great effect on body. Absence of SVC or embryological persistence of left SVC draining into the left atrium has been associated with many physical disorders/difficulties as it causes mixing of oxygenated and deoxygenated blood. Since this is importance so the asymptomatic case we found in which the PLSVC was opening into right atrium via coronary sinus and was accidentally diagnosed during central catheterization will be discussed.

Keywords:

Left superior vena cava, congenital venous anomalies, coronary sinus, hemiazygous vein, hodgkin's lymphoma, unilateral kidney

Introduction-The superior vena cava (SVC) is a large vein emptying the deoxygenated blood into the heart specifically the right atrium from upper part of the body.¹ Thoracic venous system is complex in embryonic development thus leading to a variety of disorders that can happen in that. There are number of disorders that can occur involving superior vena cava like absence of superior vena cava, its hypoplasia, abnormal opening along with this one important being persistent left superior vena cava and the pulmonary vein. The presence of double superior vena cava can exist alone or with other congenital anomalies.¹⁰ Duplication of superior vena cava (SVC) is a rare abnormality, but the most common thoracic venous congenital anomaly with incidence of is 0.3% in general population and is 10-11% in patients with other congenital heart disease.¹² Superior vein forms by the union of right and left brachiocephalic veins which are formed by the union of corresponding internal jugular and subclavian veins.² During the fifth intrauterine week, three pairs of major veins are seen: the vitelline veins-carrying blood from the yolk sac to the sinus venosus; the umbilical veins- carrying oxygenated blood to the embryo; and therefore the cardinal veins-draining the body of the embryo proper.^{3,4} These three veins form the most venous system . Here the anterior cardinal veins drains the cephalic a part of the embryo, the posterior cardinal veins, drains the remaining a part of the embryo. The anterior and posterior veins further form common cardinal veins entering the proper and left horns of the sinus venosus. Formation of the venous system is characterized by the anastomoses occurring between the left and right sides during a way that the blood from the left side is additionally directed to the proper side. Thus most of the blood from the left side of upper body is directed to the proper.^{5,6} The terminal portion of the left anterior vein is retained because the left superior inter-costal vein. The superior vein is thus formed by the proper common vein and therefore the proximal portion of the proper anterior vein. While , the left common vein and therefore the distal part of the left horn become atretic to forms the ligament of Marshal.^{4,6} If this normal regression of the left cardinal vein fails to occur, it results in occurrence of persistence left superior vena cava (PLSVC).¹¹ So PLSVC is remnant of vessel which exists as an embryological counterpart of the normal right-sided superior vena cava. Presence of PLSVC is symptomatic if it opens into left atrium causing mixing of oxygenated and deoxygenated blood. It may be asymptomatic in case it opens into right atrium via coronary sinus but it interfere and cause problems during various invasive procedures such as pacemaker implantation, central venous catheterization , retrograde delivery of cardioplegia and retrograde left ventricular 3-5 pacing ,during cardiac surgery the presence of PLSVC would be a contraindication for retrograde cardioplegia . It is possible to clamp the PLSVC to avoid retrograde cardioplegia upto the PLSVC and its tributaries so diagnosis of this is important.⁷

Case report-

A patient named-xyz Age -38years Gender- female
Came to Parul Ayurveda hospital on date 18/12/2020 seeking second opinion for conservative ayurvedic treatment if possible after receiving cancer treatment at Kailash cancer hospital and research centre (Picture 1*). Earlier patient was referred to Kailash cancer hospital and research centre Goraj vadodara Gujarat for the management of lymphoma under Hemato-oncologist Dr Shailesh Lavana on 17/11/2019 after being diagnosed outside with Hodgkin's lymphoma with tumor located at right submandibular region (Picture1,2,3).Patient also had a history of hypothyroidism and was on medication (Picture 4) for the same. While she was being treated for

lymphoma she complained of constant pain abdomen which didn't subside even after analgesics and anti-spasmodics so she was advised for ultrasonography during which we came to know that she has a congenital absence of left kidney, Hepatomegaly with fatty liver, hepatic hemangioma in segment VII and few hypoechoic neoplastic lesions in both lobes (Picture 5). Later on for further chemo treatment central venous catheter was inserted in jugular vein and post insertion x-ray was done to confirm the placement of catheter. In the x-ray catheter was found to be present more towards left atrium in place of right (Picture 6) giving suspicion of having some congenital heart anomaly. So to rule out any other congenital cardiac anomaly CT guided angio and venogram was done which confirmed the presence of functional Persistence left Superior Vena Cava (PLSVC) having partially filling defect which arises from confluence of left subclavian and jugular veins and traverses caudally lateral to the aortic arch (Picture 7,8).^{8,9} In the mid portion the vessel lies anterior to the left hilum and then traverses caudally within the left atrio-ventricular groove to drain into right atrium via a dilated coronary sinus, Hemiazygous vein drains into left SVC at the level of aorto-pulmonary window which normally drains into Azygos vein at the level of 8th thoracic vertebra. The Azygos vein further along with accessory hemiazygous and hemiazygous drains into superior vena cava. Right SVC follows the normal course and there was no communication between right and left SVC. There is a partially filling defect in left SVC signifying partial thrombosis.

Discussion-

Cardiovascular system is one of the major systems of the body and abnormalities of the vascular system are always of extreme interest. The major functioning of the cardiovascular system is maintained by the heart and the major vessels, so any abnormality in the heart or vessels will lead to a great effect on the body. Normally after the complete formation of the heart and vessels there forms a superior vena cava draining deoxygenated blood from the upper extremities, head and neck into the right atrium and one inferior vena cava draining lower extremity into the right atrium from where it further goes to systematic circulation. But absence of SVC or embryological persistence of left SVC draining into the left atrium has been associated with many physical disorders/difficulties as it causes mixing of oxygenated and deoxygenated blood. Presence of left SVC may be asymptomatic if it opens into the right atrium via the coronary sinus as there occurs no mixing of blood but it is clinically important as it can interfere and cause problems during various emergency invasive procedures such as pacemaker implantation, central venous catheterization, and retrograde delivery of cardioplegia and retrograde left ventricular 3-5 pacing, during cardiac surgery. Here the case we found was also asymptomatic as the PLSVC was opening into the right atrium via the coronary sinus and was accidentally diagnosed during central catheterization.

Conclusion-

An anatomical variation is always of extreme interest in the context of surgical branch as it can lead to extreme fatal complications. So the patient was having a persistent left superior vena cava which opens into the right atrium through the dilated coronary sinus, hemiazygous vein draining directly into left SVC instead of the Azygos vein, absence of left kidney congenitally, hepatomegaly with fatty liver, hepatic hemangioma, Hodgkin's lymphoma having tumor located at the submandibular region and hypothyroidism. Though the patient was having functionally PLSVC she wasn't having symptoms related to the clinical condition. There is also a partially filling defect in left SVC signifying partial thrombosis. With no communication between right and left superior vena cava.

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CONFLICT OF INTEREST – NIL

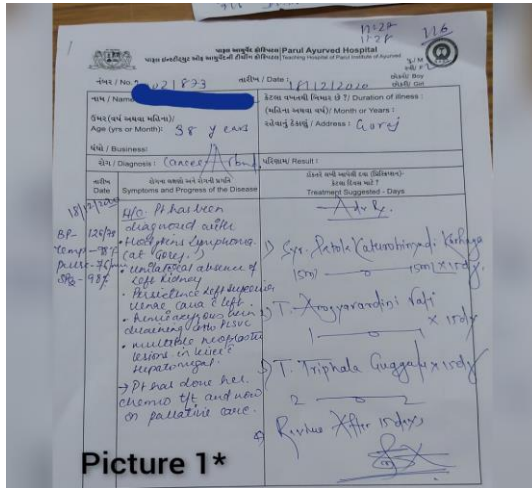
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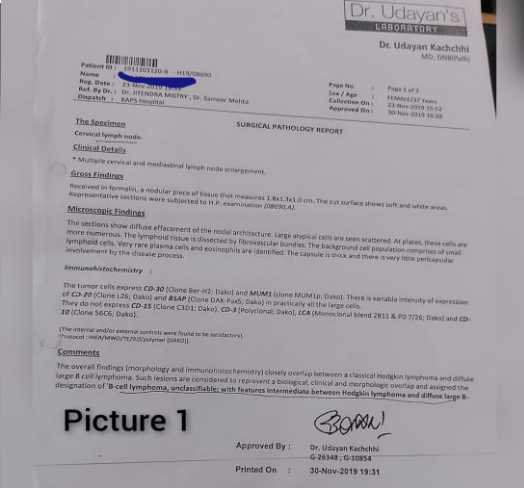
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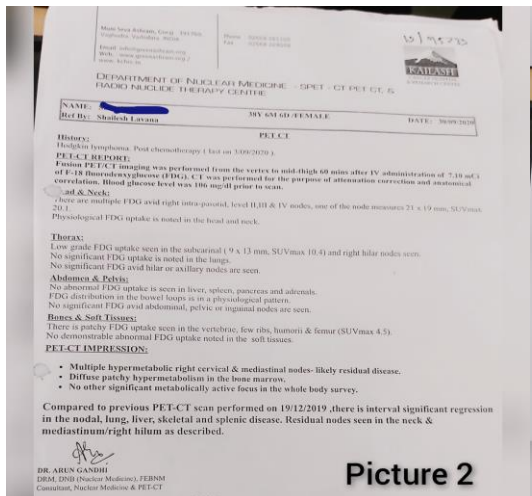
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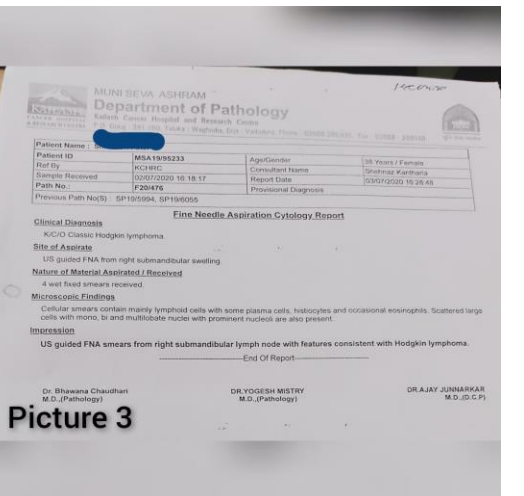
Picture 1*



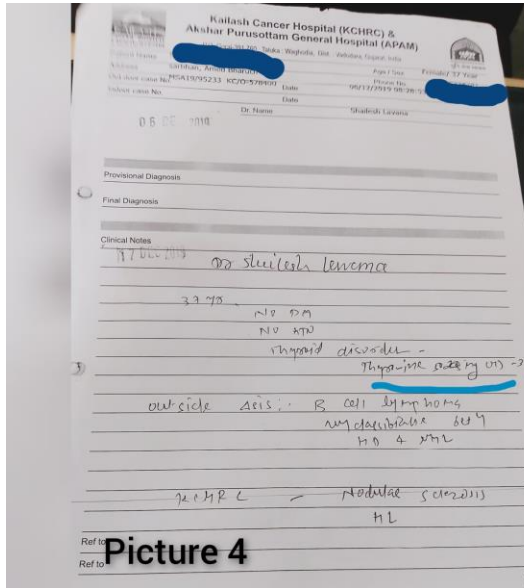
Picture 1



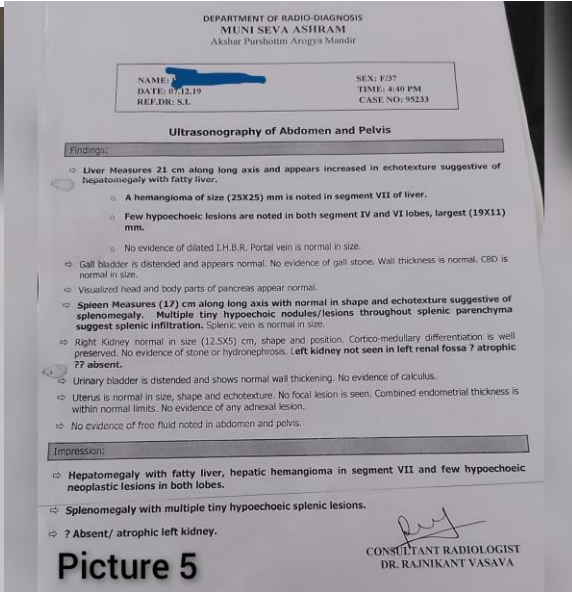
Picture 2



Picture 3



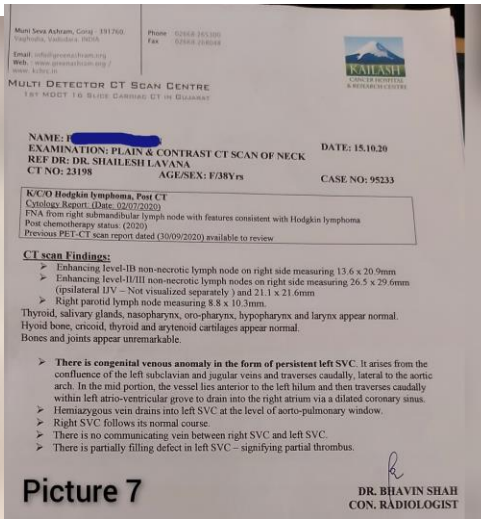
Picture 4



Picture 5



Picture 6



Picture 7



(Picture1* -OPD case of patient from Parul Ayurveda hospital, OPD Of Kaya Chikitsa, image taken on 18/12/2020)

Picture 1- Biopsy report of sample taken from cervical lymph node on 23/11/2019 at Udayan lab, image taken on 22/12/2020 Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 2- PET-CT report of patient done on 30/9/2020 at KCHRC gorej after chemo, image taken on 22/12/2020 at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 3- Report of FNAC done at KCHRC gorej on 2/7/2020, image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 4- OPD case paper of patient at KCHRC gorej at 1st visit on 17/12/2019,Showing multiple diseases of patient seen by Dr Lavana Shailesh, image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 5- report of ultrasound done on 7/12/2019 at KCHRC image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 6-Image of Chest-xray done on 3/9/2020 after Central venous catheter insertion for further chemo treatment at KCHRC gorej, image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 7- image of report of CT-neck done on 15/10/2020 at KCHRC gorej for routine examination during chemo giving suggestion of persistent left svc present,image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

Picture 8- image of CT-neck done on 15/10/2020 at KCHRC gorej,image taken on 22/12/2020at Parul Ayurveda Hospital Kayachikitsa OPD)

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