

Imaging Considerations in Polysplenia Syndrome in a Woman with Bilateral Breast Cancer

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Abstract

Case Report

Polysplenia syndrome is a rare congenital condition characterized by multiple spleens, as well as various other associated anatomical anomalies. When combined with a separate medical condition, such as bilateral breast cancer, the management and imaging considerations become particularly important. This article aims to explore the imaging modalities and considerations specific to a woman with polysplenia syndrome and bilateral breast cancer.

Keywords: Polysplenia Syndrome, Bilateral Breast Cancer, CT.

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INTRODUCTION

Polysplenia syndrome, as a subtype of heterotaxy (asplenia is another), is a rare congenital abnormalities usually diagnosed in adults, incidentally. It is initially described by Helwig in 1929 and is frequently associated with a wide range of anomalies in various organs.

In this case report, we presented an interesting case of polysplenia syndrome associated with bilateral breast cancer, in which CT was proved to be valuable in depicting the imaging features of PSS. To the best of our knowledge, the concurrence of polysplenia syndrome associated with bilateral breast cancer in the same patient has not been reported previously.

OBSERVATION

We present the case of a 52-year-old patient, with no significant medical history, admitted to the radiology department of the Mother and Child Hospital in Marrakech for staging after bilateral mastectomy for synchronous breast tumors. She does not present any clinical symptoms. All laboratory data, including tumor markers, were within normal limits. Computed tomography (CT) of the thorax and abdomen revealed multiple round formations at the hepatic hilum with a structure and density resembling that of the spleen, suggestive of polysplenia present in the left upper quadrant. The colon showed a mirror image of a 180° malrotation, with the cecum and appendix in the left hypochondrium, indicative of a common mesentery, agenesis of the dorsal portion of the pancreas. There was

interruption of the inferior vena cava with azygos continuation and direct drainage of the suprahepatic veins into the right atrium. The passage of the right subclavian artery behind the esophagus is consistent with Arteria lusoria, and there was persistence of the left superior vena cava. She received chemotherapy with hormone therapy with good clinical evolution.

DISCUSSION

There are three types of situs dictated by the arrangement of organs that are normally asymmetrical. Situs solitus occurs when these organs are in the normal position and situs inversus occurs when this arrangement is reversed. Situs ambiguus (heterotaxy) is a third and abnormal type of situs in which the relationship of the atria and the viscera is inconsistent [2-5]. Situs ambiguus can be divided into two broad groups: right-sided isomerism (asplenia) and left-sided isomerism (polysplenia); isomerism describes the symmetrical arrangement of the organs that are normally asymmetrical. In polysplenia syndrome, however, there is no single anomaly that is pathognomonic for this condition. The affected patients have a lower prevalence of congenital heart disease (50–90%) and less severe defects than those with situs ambiguus with asplenia [1-5]. The majority of patients with polysplenia syndrome die by the age of 5 years. This high mortality rate is mainly due to the severe cardiac anomalies. In cases with an absence of severe cardiac or visceral abnormalities, as was observed in the current case, asymptomatic survival into adulthood is possible [5]. The anomaly is often incidentally discovered on computed tomography or

magnetic resonance (MR) imaging when these patients are evaluated for other medical reasons.

There is a very low incidence of situs there is a very low incidence of situs ambiguus and furthermore, like the presented case, double breast cancer with situs anomaly is extremely rare. Although this anomaly is not considered to be a premalignant entity, several malignant neoplasms have been sporadically reported in association with situs anomaly [6]. The combination of congenital anomaly and neoplasm has generally been considered to be coincidental rather than a reflection of a common pathogenesis [6].

Polysplenia syndrome is often associated with vascular anomalies, such as interrupted inferior vena cava (IVC) or azygos continuation of the IVC. These vascular variations can impact the venous drainage patterns and affect contrast enhancement during CT scans [7-9]. Radiologists need to be aware of these vascular anomalies to accurately interpret the images and understand the potential impact on surgical planning or other interventions. [7, 8].

CT scans play a crucial role in assessing the spleen(s) in polysplenia syndrome. It helps evaluate the number, size, location, and morphology of the multiple spleens [9]. CT can also identify any associated splenic abnormalities, such as splenic cysts, infarcts, or accessory spleens. This information is valuable for understanding the overall splenic function and potential implications for the patient's health [9].

A second primary breast cancer in the opposite breast can be either synchronous or metachronous. The majority are metachronous. A woman who has had breast cancer has a fivefold increase in risk for a second breast cancer [10]. Additional risk factors include multifocal cancer, lobular carcinoma in situ, and an original cancer at an early age with long survival. Lobular carcinoma in situ is predominantly a marker for the subsequent development of a second primary breast cancer. The incidence of synchronous bilateral cancer is approximately 1% to 2% and that of metachronous cancer 5% to 6% [10].

CT scans are commonly used for detecting and staging cancers, including breast cancer. In the case of a woman with polysplenia syndrome and bilateral breast cancer, CT scans can help assess the extent of the disease in the breasts, regional lymph nodes, and distant metastases [7-9]. The radiologist should carefully evaluate the CT images, taking into account the altered anatomy and potential splenic-related findings that may affect the accurate staging of the breast cancer [7-9].

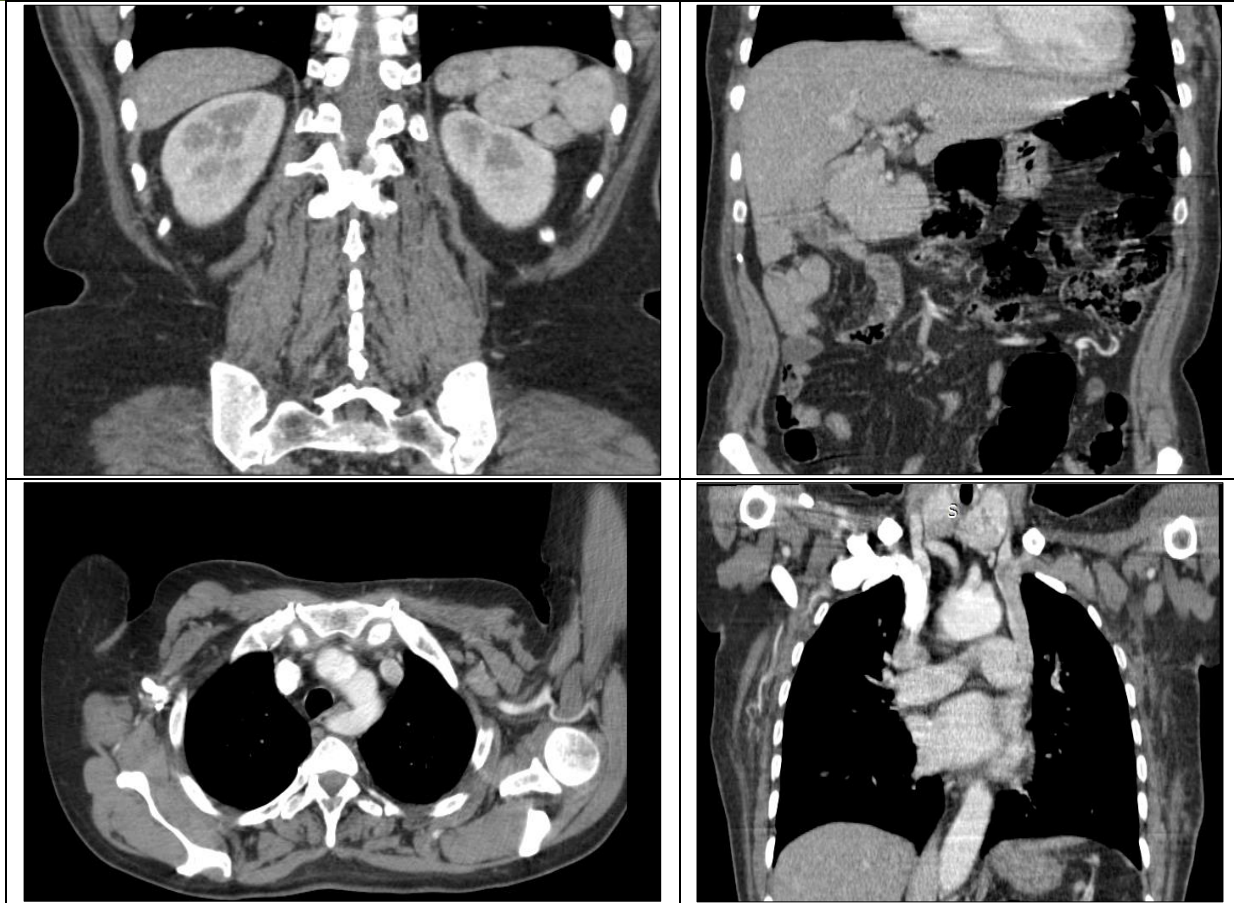
In individuals with polysplenia syndrome and breast cancer, CT scans can aid in surgical planning by providing detailed anatomical information. The scans help determine the relationship between the tumors, breast tissue, and adjacent structures. This knowledge is crucial for surgical oncologists to make informed decisions regarding the extent of surgery and potential modifications required due to the presence of polysplenia syndrome [11].

It is essential for the radiologist and the multidisciplinary team involved in the patient's care to be aware of the unique considerations associated with polysplenia syndrome during CT scan interpretation. Effective communication and collaboration among radiologists, oncologists, and surgeons are key to ensure accurate diagnosis, staging, and treatment planning for individuals with both polysplenia syndrome and bilateral breast cancer [9-11].

MRI of the breast has emerged as the most sensitive radiological modality for breast tumors. Breast MRI has been found to be 94–99% sensitive for the detection of breast cancer [10]. Dynamic breast MRI, attempts to distinguish benign and malignant lesions according to the enhancement kinetics at a high temporal resolution [10]. With this method, both breasts are imaged at the highest possible temporal resolution (about 60 s). In contrast, static breast MRI, attempts to achieve the same goal by evaluating the morphologic features at a high spatial resolution [10]. This method images a single breast at a high spatial resolution. The severe technical constraints of MR units have previously made it necessary to choose between either temporal or spatial resolution [10]. In addition, ultrasonography is widely used to distinguish benign and malignant lesions; breast MRI is usually performed to evaluate the extent of breast cancer after a histological confirmation of malignancy has been made. In this situation, static imaging may be more suitable than dynamic MRI [10].

CONCLUSION

Imaging plays a crucial role in the diagnosis, staging, and treatment planning of bilateral breast cancer in individuals with polysplenia syndrome. Understanding the unique challenges and considerations associated with this rare syndrome is essential for accurate imaging interpretation and optimal patient care. By adopting a multidisciplinary approach and leveraging the strengths of various imaging modalities, healthcare professionals can provide tailored treatment strategies to improve outcomes for these patients. Continued research and collaboration will further enhance our knowledge and refine imaging techniques for individuals with polysplenia syndrome and concurrent breast cancer.



Figures 1: The abdominal coronal and thoracic axial CT scans show multiple round formations at the hepatic hilum with a structure and density resembling that of the spleen, suggestive of polysplenia. Additionally, there is agenesis of the dorsal portion of the pancreas, interruption of the inferior vena cava with azygos continuation and direct drainage of the suprahepatic veins into the right atrium. The passage of the right subclavian artery behind the esophagus is consistent with Arteria lusoria, along with a common mesentery and persistence of the left superior vena cava

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