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CLINICAL CASE REPORT

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# Polysplenia Syndrome With Splenic and Skeletal Muscle Metastases From Thyroid Carcinoma Evaluated by FDG PET/CT: Case Report and Literature Review

## *A CARE-Compliant Article*

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**Abstract:** Polysplenia syndrome (PSS) is a rare congenital abnormality. Metastases to spleen and skeletal muscle from differentiated thyroid cancer (DTC) are also extremely rare. Our case report aims to present an interesting case of PSS associated with splenic metastasis (SM) and skeletal muscle metastasis (SMM) from advanced papillary thyroid carcinoma which was evaluated on fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT).

An 84-year-old Chinese man was admitted with the history of multiple enlarged masses in bilateral neck, right axillary, and inguinal areas for >2 months. The results of ultrasonography examination were highly suggestive of malignancy. The histological results of the following biopsy were consistent with papillary thyroid carcinoma with involvement of multiple regional lymph nodes. He was referred for an FDG PET/CT imaging to evaluate the situation.

FDG PET/CT showed that an intense FDG-avid thyroid mass with widespread regional lymph node involvement and distant metastases in the body. Unexpected sites of metastases were detected in the spleens and skeletal muscles. Most interestingly, FDG PET/CT imaging also described the typical imaging findings of PSS including the 2 right-sided spleens, azygos and hemiazygos continuation of inferior vena cava (IVC), right-sided stomach, middle line liver, a short pancreas, preduodenal portal vein (PPV), and malrotation of gut.

Whole body FDG PET/CT imaging can accurately evaluate the situation of DTC by detecting regional lymph node involvement, common and rare sites of distant metastases which are closely related to staging, management, and prognosis of this disease. Whole-body FDG PET/CT is also valuable in demonstrating the typical imaging features of PSS.

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**Abbreviations**: DTC = differentiated thyroid cancer, FDG = fluorodeoxyglucose, IVC = inferior vena cava, PET/CT = positron emission tomography/computed tomography, PPV = preduodenal portal vein, PSS = polysplenia syndrome, SM = splenic metastasis, SMM = skeletal muscle metastasis.

## INTRODUCTION

Polysplenia syndrome (PSS), 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.[1–8]

Papillary thyroid carcinoma, as a DTC, ordinarily behaves in an indolent manner with good prognosis and long-term survival. Distant metastases are only seen in a minority of DTC patients, and the most common site of distant metastasis is the lung, followed by bone. SM and SMM from DTC are extremely rare, and they are usually ignored in patients with thyroid cancer. However, the detection of rare distant metastases may have significant impact on the staging, management, and prognosis of the patients.[9–12]

In this case report, we presented an interesting case of PSS associated with SM and SMM from papillary thyroid carcinoma, in which FDG PET/CT was proved to be valuable in detecting the common and uncommon sites of distant metastases, as well as in depicting the imaging features of PSS. To the best of our knowledge, the concurrence of PSS, SM, and SMM from papillary thyroid carcinoma in the same patient has not been reported previously.

## CASE PRESENTATIONS

An 84-year-old Chinese man was admitted with complaints of enlarged masses in bilateral neck, right axillary, and inguinal areas for >2 months. He also complained of progressive pain on low back, buttock, and legs for nearly 1 month, ambulation dysfunction for nearly 1 week, and urinary incontinence for 3 days. A Foley catheter was inserted to alleviate symptom. PICC catheter was also inserted to superior vena cava through right side. He had no previous history of splenectomy. On physical examination, masses were palpable in bilateral neck, right axillary, and inguinal areas. In the following ultrasonography, an ill-defined mass in the lower pole of the right thyroid lobe with multiple superficial lymphadenopathies in bilateral cervical and inguinal areas, and right axillary area were found, which was highly suggestive of malignancy. A provisional diagnosis of advanced thyroid carcinoma was made. Biopsy in thyroidal mass and the superficial lymphadenopathies in bilateral neck, right axillary, and inguinal regions were suggested to obtain a definitive histological diagnosis and to exclude other possible malignancies. The histological results of biopsy were consistent with papillary

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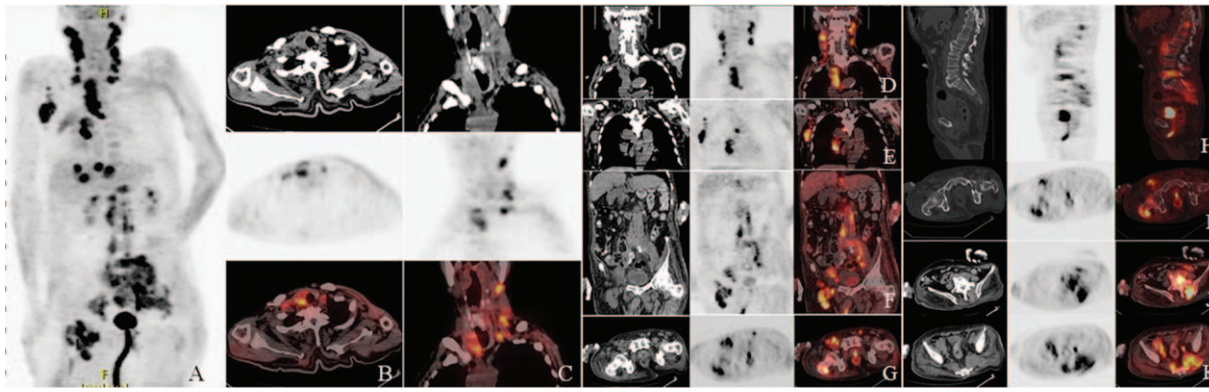


**FIGURE 1.** Whole-body F-18 FDG PET/CT imaging on a patient with advanced right papillary thyroid carcinoma. (A) 3D-MIP images demonstrated widespread increased FDG uptake regions in the body. (B, C) Selected axial and coronal PET/CT images show that an ill-defined hypodensity mass with inhomogeneous intense FDG uptake in the lower pole of right thyroid lobe (red arrow), measuring 2.0 cm × 3.3 cm × 4.0 cm, SUVmax = 7.0. Selected PET/CT images also show that extensive markedly FDG-avid sites in regional lymph nodes (D–G), bones (H, I), and skeletal muscles (J, K). Imaging findings were highly consistent with advanced thyroid carcinoma with widespread metastases in the body. 3D-MIP = 3-dimensional maximum intensity projection, FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/computed tomography, SUVmax = maximum of standard uptake value.

thyroid carcinoma with multiple regional lymph nodes metastases. The patient was then referred to our PET/CT department to further evaluate the situation and search for possible metastatic disease.

The patient fasted >6 h before the tracer injection and received an intravenous injection of ~5.55 MBq/kg (0.15 mCi/kg) of 18F-FDG, with a maximum of 370 MBq (10 mCi). The blood glucose level was measured immediately at the time of FDG injection and it was ~5.7 mmol/L. He was instructed to rest in a quiet dim room without talking during the subsequent 1 h of the FDG uptake phase and was allowed to breathe normally during image acquisition without specific instructions. Whole-body PET/CT scan was performed using a PET/CT scanner (Discovery$^{TM}$ ST-16; General Electric Medical Systems, Milwaukee, WI).

Whole-body F-18 FDG PET/CT images showed an ill-defined hypodensity mass with inhomogeneous intense FDG uptake in the lower pole of right thyroidal lobe (Figure 1 A–C), with widespread FDG-avid regions in regional lymph nodes (Figure 1 D–G), bones (Figure 1 H–I), skeletal muscles (Figure 1 J–K), and 3 intense FDG-avid metastases in 2 equal-volume masses in the right upper abdomen (Figure 2).

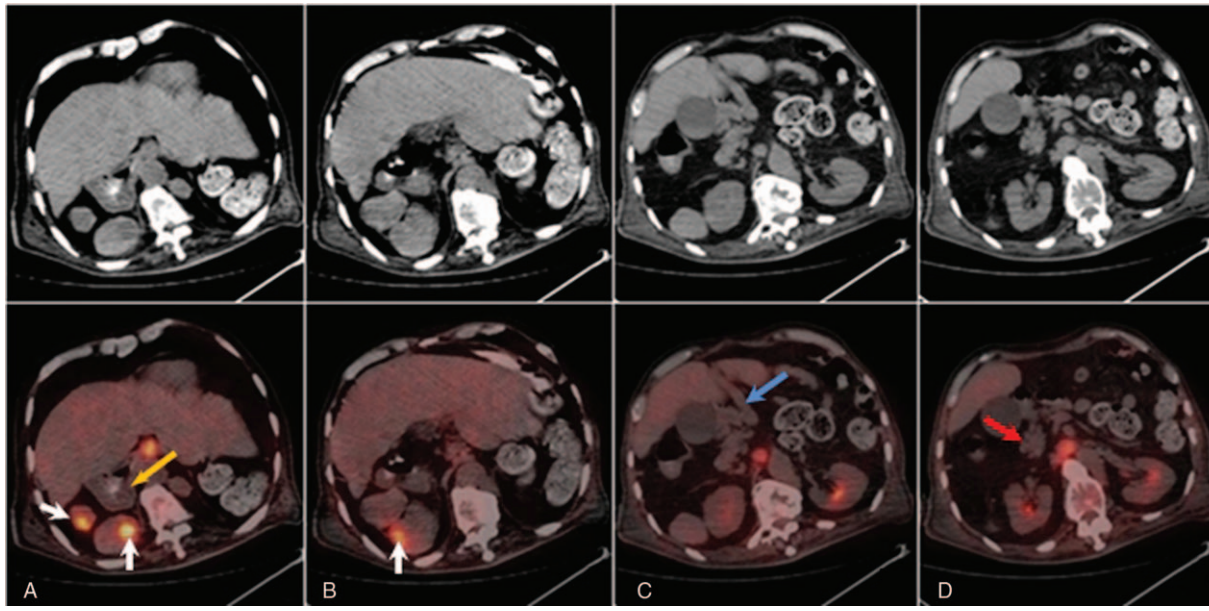


**FIGURE 2.** PSS with multiple splenic metastases was presented. (A, B) FDG PET/CT images showed that 2 spleens were located in right abdomen with 3 hypermetabolic foci (white arrow), suggesting splenic metastases. (A–D) Right-sided stomach (yellow arrow), PPV (blue arrow), a short pancreas in the absence of body and tail (red arrow), a midline liver, and malrotation of gut are also presented. These findings are consistent with the diagnosis of PSS with multiple splenic metastases. FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/computed tomography, PPV = preduodenal portal vein, PSS = polysplenia syndrome.

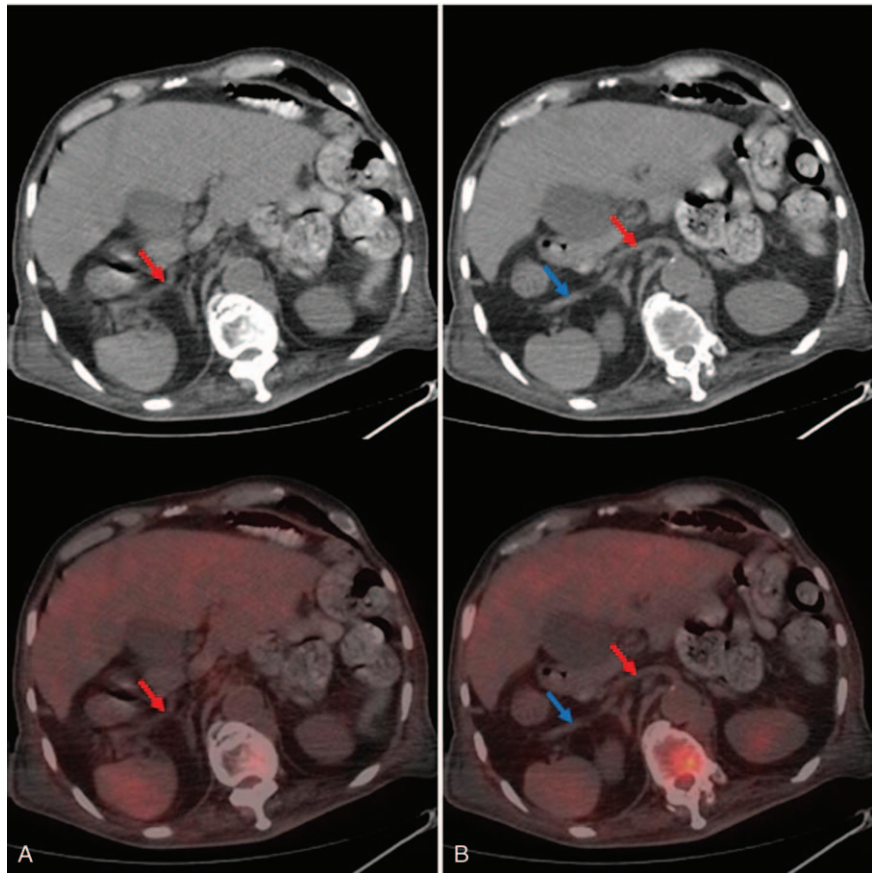


**FIGURE 3.** Splenic vessels were observed on FDG PET/CT. (A, B) Splenic artery (red arrow) and splenic vein (blue arrow) are demonstrated in right upper abdomen on FDG PET/CT images. FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/computed tomography.

The splenic vessels identified by FDG PET/CT images help us to prove that these 2 right-sided masses are actually multiple spleens (Figure 3). Most interestingly, spleen is absent in left upper abdomen, even though this patient had no previous history of splenectomy. PET/CT images also showed the findings of azygos and hemiazygos continuation of the IVC with the absence of the hepatic segment (Figure 4), which was the most frequent finding of vascular anomalies in PSS. Other imaging findings included a right-sided stomach, a middle line liver, a short pancreas in the absence of body and tail, preduodenal portal vein (PPV), and malrotation of gut (Figures 2 and 4). These imaging findings were highly consistent with the diagnosis of PSS.

Although these appearances of PET/CT were not unique to advanced thyroid carcinoma and could also be seen in other malignancies, the histopathological results of biopsy performed before PET/CT imaging excluded other possible malignancy including lymphoma or a second primary malignancy. Based on the results of imaging and the histological results of biopsy, a final diagnosis of PSS associated with widespread metastases from advanced right papillary thyroid carcinoma was made.

Due to the advanced stage of the disease which was successfully evaluated by FDG PET/CT and the poor condition of the patient, he refused any aggressive therapy. The patient was treated with supporting and symptomatic therapy. Then he died from complications probably caused by systemic metastases nearly 3 weeks later.

The institutional review board (Pingjin Hospital) approved this work and informed consent was given by patient. The authors of this manuscript have no conflicts of interest.

## DISCUSSION

Polysplenia syndrome (PSS), as a subtype of heterotaxy (asplenia is another), is a rare congenital abnormalities. It is generally defined as the presence of multiple spleens and various organ anomalies in the absence of a history of splenectomy.[1–8] The precise etiology of polysplenia is unknown yet. Embryonic, genetic, and teratogenic components have all been implicated as causative factors in polysplenia.[13] PSS in adults is usually asymptomatic and diagnosed during investigation for unrelated causes.[2] In our case, even though whole-body FDG PET/CT was performed to search possible metastatic disease, it incidentally reveals that multiple right-sided spleens in the absence of spleen in left abdomen, azygos and hemiazygos continuation of the IVC with the absence of the hepatic segment, and other anomalies including right-sided stomach, a short pancreas, middle line liver, PPV, and malrotation of gut, which were highly consistent with the diagnosis of PSS.

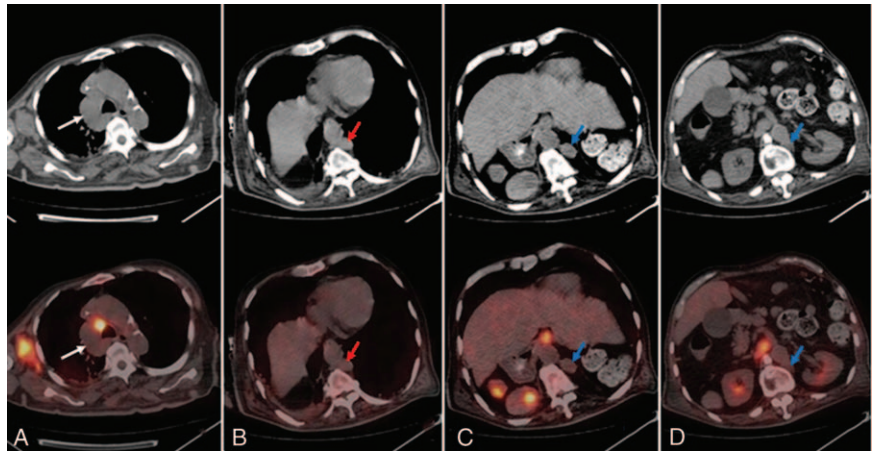


**FIGURE 4.** Azygos and hemiazygos continuation of IVC presented on FDG PET/CT. (A) Enlarged azygos arch is paralleling the aorta as it drains into the SVC (white arrow). (B) Enlarged hemiazygos drains into the azygos vein at approximately the ninth thoracic vertebral level (red arrow). (C,D) Enlarged hemiazygos lies on the left side of the aorta (Blue arrow). The intrahepatic portion of the IVC is not identified. Other anomalies including multiple right-sided spleens, right-sided stomach, a short pancreas, PPV, and malrotation of gut were also noted. FDG = fluorodeoxyglucose, IVC = inferior vena cava, PET/CT = positron emission tomography/computed tomography, PPV = preduodenal portal vein, SVC = superior vena cava.

Increased awareness of such anatomical anomalies would prevent serious complications during abdominal surgery or performing invasive diagnostic procedures.

Splenic metastasis (SM) from thyroid carcinoma is extremely rare. Skin melanoma and carcinomas of the breast, lung, ovary, colorectum, and stomach are the most common primary sources of splenic metastases.[14–16] To our knowledge, only 3 cases of splenic metastases from thyroid carcinoma have been reported to date.[17–19] The relative rarity of splenic metastases might be explained by the following reasons: (1) the sharp angle made by the splenic artery, which make it difficult for tumor emboli to enter the spleen; (2) the rhythmic contractile nature of the spleen, which squeezes out the tumor embolus and prevent its lodging in the spleen; (3) the absence of afferent lymphatic to bring metastatic tumor to the spleen; and (4)antitumor activity due to a high concentration of lymphoid tissue in the spleen.[15,16,20,21] In our case report, considering the potential risk of bleeding, as well as the congential anomalies of spleen illustrated by FDG PET/CT, biopsy in the FDG-avid lesions of the spleens was also not used. Therefore, the superior ability of PET/CT in the detection of metastases can also help provide more accessible biopsy sites and help avoid unnecessary invasive diagnostic procedures.

Skeletal muscle metastasis (SMM) is also exceedingly uncommon. The mechanism SMM is not well known yet. It is thought that muscle movement, unsuitable muscle pH, and muscle's capability to remove the lactic acid produced by tumor contribute to the resistance of skeletal muscles for the metastatic process.[22,23] It is reported that SMM occurred more frequently in patients with carcinoma of lung, kidney, colon, ovary, cervix and uterus, malignant melanoma.[24–26] However, the reports of SMM from DTC are relatively rare. According to a retrospective review, there are only a few reports of skeletal muscle metastases from DTC in previous literatures.[10,27–35] In our presented case, multiple intense FDG-uptake metastatic lesions were found in multiple buttock muscles including bilateral piriformis, left erector spinae, and gluteus maximus muscles, as part of multivisceral tumor dissemination in the body. Our findings highly paralleled those previously reported in the literature.[27,33]

Papillary thyroid carcinoma, as a DTC, is frequently characterized by good prognosis and long-term survival. The factors including patient age >45 years and the advanced tumor stage are indictor of poor prognosis and closely related to the management of the disease.[9] Although the common distant sites and regional lymph nodes involvement from DTC usually draw significant concern to us, rare site of metastases are easily ignored because of their scarcity. However, recognizing the patterns of rare metastases has a large effect on clinical decision making and prognosis of DTC patients. Because of its high sensitivity and specificity, molecular imaging with 18F-FDG PET/CT imaging is a well-established valuable noninvasive imaging modality for diagnosing, staging, restaging, treatment monitoring assessment, as well as a prognostic indictor of oncological patients, including DTC.[10–12] As the use of PET/CT imaging become more common in evaluating thyroid carcinomas, SM and SMM may not be as rare as we once thought and may increasingly common. In our case, FDG PET/CT can successfully identify widespread regional lymph nodes involvement and common bone metastases, as well as detect the unexpected SM and SMM. The detection of these common and unexpected distant metastases from DTC can lead to the modification of treatment strategies, have prognostic implications, provide more accessible biopsy sites, and help avoid invasive procedures. Therefore, the patient with thyroid carcinoma should be further investigated with FDG PET/CT to give a more compressive assessment of the disease.

## CONCLUSIONS

Our case presented here highlights that whole-body FDG PET/CT imaging can accurately evaluate the situation of DTC by detecting regional lymph node involvement, common and rare distant metastases which are closely related to diagnosis, staging, management, and prognosis of this disease. It is also valuable in demonstrating the salient imaging features of PSS.

Our case is unique in that PSS, SM, and SMM from DTC are existed in the same patient.

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