

Asymptomatic Large Pulmonary Arteriovenous Malformation Masquerading as a Metastatic Deposit in a Patient with Papillary Thyroid Carcinoma

Abstract

Adult cases of congenital arteriovenous malformation (AVM) of the mediastinum are extremely rare, and because of their varied clinical presentations, they pose a diagnostic challenge. There is no reported association of pulmonary AVM and papillary thyroid carcinoma. We describe a 38-year-old female with a large right lung AVM, multinodular goiter, and high serum thyroglobulin (Tg) with papillary thyroid carcinoma. The lung mass was believed to represent a pulmonary metastatic deposit due to high Tg value. Whole body I-131 scan and FDG PETMR imaging were performed as part of the workup.

Keywords: FDG PETMR, papillary thyroid cancer; pulmonary arteriovenous malformation, thyroid cancer, whole-body I-131 scan

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Introduction

Pulmonary arteriovenous malformation (PAVM) is a rare cardiovascular anomaly. Since its first description at autopsy in 1897,^[1] these abnormal communications have been given various names, including pulmonary arteriovenous fistulae, pulmonary arteriovenous aneurysms, hemangioma of the lung, cavernous angioma of the lung, pulmonary telangiectasis, and PAVM. Most cases are congenital, frequently related to hereditary hemorrhagic telangiectasia (HHT), an autosomal dominant disorder.^[2] Isolated cases have also been reported. Rarely, diffuse PAVMs occur in the context of unoperated congenital heart malformations of the type of polysplenia syndrome. Other etiologies include acquired conditions, such as schistosomiasis, tuberculosis, juvenile hepatic cirrhosis, mitral stenosis,^[3] trauma,^[4] Fanconi's syndrome,^[5] and metastatic thyroid carcinoma.^[6] It may mimic a variety of disorders, including neoplasm, infection, inflammation, and vascular abnormality, including some rare conditions of pulmonary circulation. It occurs twice as often in women as in men, but there is a male preponderance in newborns.^[7] In 53-70% of cases, PAVM have been

found in lower lobes, 75 % of them may be unilateral, while 36 % may have PAVMs in more than one site. Half of these multiple AVMs have bilateral localization.^[8]

Case Report

A 38-year-old Indian woman gave a history of a painless anterior neck swelling since 2008. The patient is a school teacher by profession. She was evaluated at a local hospital and was found to have a multinodular goiter on ultrasonography (USG) of the neck. Fine-needle aspiration cytology (FNAC) suggested benign thyroid pathology as per the patient. The neck swelling has slowly increased in size over the years. The patient had occasional cough with mucoid expectoration for the past 2 weeks. The patient presented to the Head and Neck Oncology Department of Amrita Institute of Medical Sciences, Cochin, in August 2019. On local examination, a 9 cm × 7 cm anterior neck swelling, firm in consistency, with the nodular surface was noted. The mass moved with deglutition and the lower border of the mass was felt. The skin over the swelling appears normal with no vessel engorgement. No stridor or pressure symptoms/signs were elicited. The patient was evaluated with baseline blood investigations, such as hemogram,

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thyroid function tests, followed by the USG of the thyroid, and fine-needle aspiration of the thyroid nodule. USG revealed a TI-RADS IV nodule in the left lobe of the thyroid gland, with FNAC suspicious for papillary thyroid carcinoma, Category V as per the Bethesda System for Reporting Thyroid Cytopathology. A total thyroidectomy was suggested and was performed. Histopathological examination revealed follicular variant of papillary thyroid carcinoma, tumour measuring 5.9 cm × 3 cm × 4 cm. No tall cell, oncocytic, columnar, or poorly differentiated cells were seen. Lymphovascular emboli were present. No perineural invasion or any extrathyroidal extension was noted. Tumor abuts the inked thyroid capsule.

The whole-body I-131 scan showed moderate residual thyroid tissue [Figure 1]. Single-photon emission computed tomography-computed tomography (SPECT-CT) of the neck and thorax showed moderate residual thyroid tissue in the thyroid bed. SPECT-CT incidentally detected a lobulated

hilar mass on the right side with a cutoff of the right upper lobe bronchus. There was no abnormal I-131 uptake in the hilar mass. No mediastinal or cervical lymph nodes were seen. There was no evidence of any functioning distant metastases also. Serum thyroglobulin (Tg) was suggested and was found to be high (148 ng/ml off Thyroxine).

¹⁸F-FDG PETMR with gadolinium contrast was performed in view of high suspicion of distant metastases. Inspiratory noncontrast CT of the chest was performed to look for lung nodules. PETMR images showed abnormal diffuse FDG uptake in the thyroid bed region (SUV Max 7.15), suggesting postoperative inflammatory changes. No abnormal FDG uptake was seen in MR detected enhancing lobulated mass measuring 5.6 (AP) cm × 4.2 (T) cm × 4.3 (CC) cm in the anterior segment of the right upper lobe lung [Figure 2]. The lesion was abutting the mediastinal pleura and superior vena cava with the abrupt cutoff of the right upper lobe bronchus. There

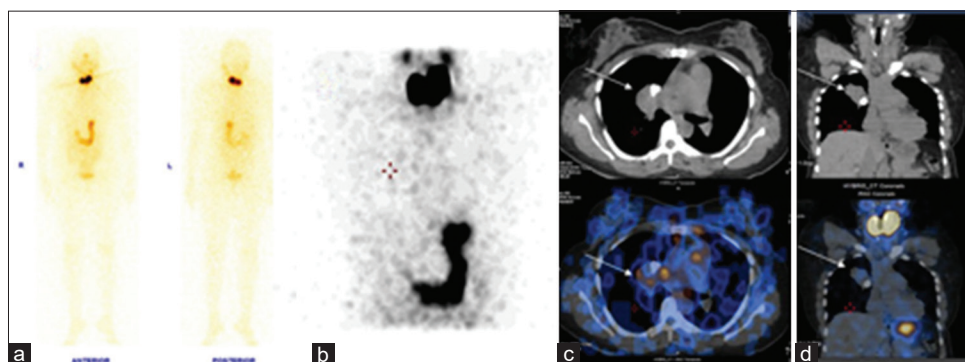


Figure 1: (a) Whole-body I-131 scan in the anterior and posterior projections showing moderate residual thyroid tissue. (b) SPECT image in coronal section showing no abnormal I 131 uptake in site corresponding to the mediastinal mass. (c) SPECTCT in the transaxial section and (d) coronal section showing no significant I-131 uptake in CT detected right hilar mass lesion (arrow) with a focal speck of calcification

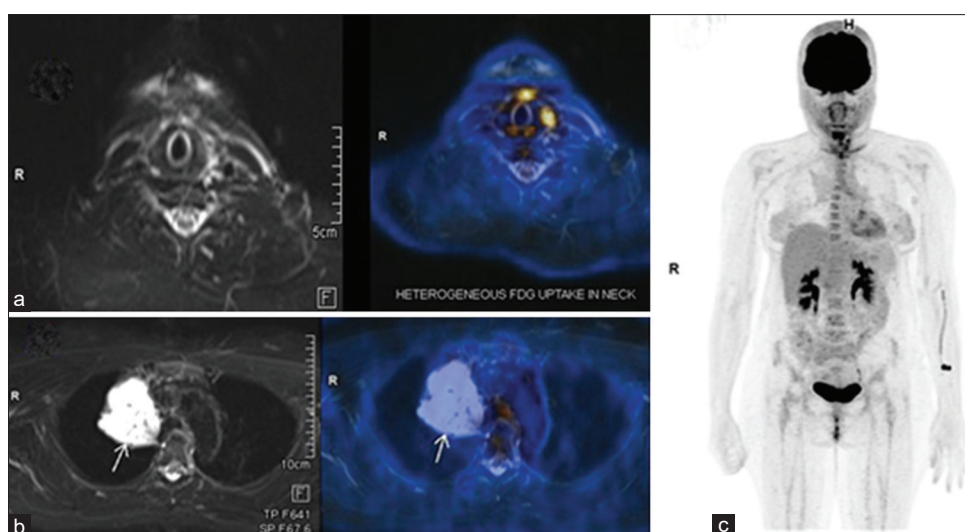


Figure 2: (a) FDG PETMR (MR + Fused PETMR) neck (transaxial) image showing minimal FDG uptake in postoperative site with no residual/recurrent thyroid bed lesion. (b) FDG PETMR thorax (transaxial) image shows no abnormal FDG uptake in MR detected enhancing lobulated mass (arrow) in the anterior segment of the right upper lobe lung, abutting the mediastinal pleura, and superior vena cava with the abrupt cutoff of the right upper lobe bronchus. The lesion showed peripheral restriction on diffusion-weighted images. (c) Maximum intensity projection image showing no FDG avid nodal or distant lesions in the whole-body survey

was subsegmental collapse of the anterior segment. The lesion showed peripheral restriction on diffusion-weighted images. No enhancing nodules in the lung fields and no significant cervical or mediastinal lymph nodes were noted. There was FDG nonavid bilateral minimal pleural thickening, (STIR bright). No abnormal FDG uptake was seen in multiple tiny cystic lesions scattered in both lobes of the liver; few of them showed diffusion restriction and peripheral enhancement, largest one was 1.8 cm × 1.1 cm in Segment V. Findings raised the possibilities of pulmonary metastasis/primary right lung malignancy. Liver lesions were thought to represent cysts/metastases on MR.

Later, bronchoalveolar lavage cytology showed no malignant cells. CT angiogram was performed, and middle mediastinal arteriovenous malformations (AVMs) Grade 3 was confirmed. Lung tumor embolization was done in September 2019 through the right common femoral artery and common femoral vein approach. Arch aortogram revealed minimal vascular blush in the mediastinal lesion in the right hemithorax. The selective right intercostobronchial trunk showed multiple tiny branches. The branches were obliterated with 150–300 polyvinyl alcohol particles achieving significant stasis. No feeders from other intercostal or internal mammary arteries were noted. The tumor also showed abnormal blush in tiny upper lobe branch, supplying the inferior aspect of the mediastinal tumor.

6 weeks later, patient was scheduled for High dose I-131 therapy. Patient stopped thyroxine for 4 weeks and was adequately prepared with iodine restricted diet. She was treated with 3515 MBq of I-131 orally. At 3-month follow-up, the patient was asymptomatic clinically. USG of the neck showed no evidence of any local disease or cervical nodes. Follow-up whole-body I-131 scan showed a successful ablation of thyroid tissue. Stimulated Tg was 0.6 ng/ml with anti-Tg antibody of 21 IU/ml (normal <115). Followup I-131 scan and stimulated Tg, findings indicate a good response to high-dose I-131 therapy.

Discussion

AVMs in the anterior mediastinum are exceedingly rare, especially in adults.^[7] They arise from a developmental anomaly occurring at the embryonic vascular system between 4th and 5th week of intrauterine life. They grow slowly over time and are usually asymptomatic unless they produce pressure symptoms to nearby structures due to their gross enlargement. AVM usually consists of multiple direct communicating channels in between the arteriovenous branches, with no intervening capillary network.

Careful evaluation of a presurgical or a medical-based therapy includes a contrast-enhanced CT. This allows (1) the determination of the number and location of PAVMs; (2) the identification of the number, location, and size of supplying arteries and draining veins; (3) the

presence of thrombosis; and (4) location of any upstream side branches useful for anchoring coils.

They can be classified as simple, complex, or diffuse:^[9]

1. Simple type: The most common has a single segmental artery feeding the malformation; the feeding segmental artery may have multiple subsegmental branches feeding the malformation but must have only one single segmental level
2. Complex type: Have multiple segmental feeding arteries (~20%)
3. Diffuse type: Rare (~5% of lesions); the diffuse form of the disease is characterized by hundreds of malformations; some patients can have a combination of simple and complex AVMs within a diffuse lesion and are a particular challenge to treat effectively.

PAVMs can be further characterized according to their radiological appearance. The fistula-type PAVM has a feeding artery directly connected to a draining vein, with an intervening single aneurysmal sac. Less commonly, PAVMs are plexiform with a multiseptated aneurysm or a cluster of vascular channels.

Another older embryological-based classification was proposed by Anabtawi *et al.* in 1965.^[10]

Group I: Multiple small arteriovenous fistulas without an aneurysm

Group II: Large peripheral arteriovenous aneurysm

Group III:

- a. Large arteriovenous aneurysm (central)
- b. Large arteriovenous aneurysm with anomalous venous drainage
- c. Multiple small arteriovenous fistulae with anomalous venous drainage.

Group IV:

- a. Large venous aneurysm with systemic arterial communication
- b. Large venous aneurysm without fistula.

Quiescent AVMs may be managed conservatively using the “wait-and-watch approach.”^[11] If the AVM becomes symptomatic, however, the treatment options are embolization or complete surgical removal. Complete surgical removal of PAVMs may be complicated by the encroachment of surrounding mediastinal vital structures. In such cases, embolotherapy helps in reducing the blood flow within the vascular tumor.^[12-14] In our patient, the mass was considered quite large. However, since the advent of embolotherapy, percutaneous transcatheter embolization with coils has significantly decreased the rate of complications arising from PAVMs. Idiopathic PAVMs are anatomically similar to HHT-related PAVMs, with the notable differences of a greater proportion of solitary PAVMs and a lack of lower lobe predominance. Finally, transcatheter embolotherapy is a safe and effective method for treating patients with idiopathic PAVMs.

Conclusion

Nuclear imaging techniques and I-131 therapy are the established modalities in differentiated thyroid cancer management. There are no reports of association of PAVM and thyroid cancer. A high index of suspicion in this patient led to the identification of asymptomatic PAVM, which is usually challenging, otherwise, it would have prompted further aggressive thyroid cancer management as it mimicked a metastatic deposit. MR is the imaging of choice for mediastinal vascular tumors. In this special situation, FDG PET along with contrast-enhanced MR served as a one-stop shop and was incremental in the final decision-making.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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