¹⁸F-FDG PET/CT imaging of pseudomyogenic hemangioendothelioma presenting as multiple nodules with right ventricle and pancreas involvement

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Abstract

Pseudomyogenic hemangioendothelioma (PMH) presenting with right ventricle and pancreas involvement is very rare. Herein, we reported a 46-year-old man who presented multiple subcutaneous nodules throughout the body for eight months. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (¹⁸F-FDG PET/CT) revealed multiplehypermetabolic lesions including in the soft tissues, right ventricle and pancreas. Fine needle aspiration of subcutaneous nodule revealed a PMH.



Figure 1. A 46-year-old man presented multiple subcutaneous nodules throughout the body for eight months. The physical examinations showed multiple subcutaneous nodules of varying size with local tenderness. Tumor marker indicated increasing CA-125 (36.6U/ml, normal reference <24U/mL) and NSE (39.7ng/mL, normal reference <20.4ng/mL). Other laboratory examinations had no apparent abnormalities. Puncture biopsy of the nodule in the left thigh indicated tumor-infiltrating lesion. Possible diagnoses included epithelioid hemangioma, rhabdoid tumor and epithelioid angiosarcoma. Thus, ¹⁸F-FDG PET/CT was performed for staging. The maximum intensity projection (MIP) image(A) showed multiple lesions in the soft tissues of extremities and body trunk. The axial CT (B), PET (C) and fusion images (D) demonstrated the biggest soft tissue lesion in the left thigh, with a size of 40 x 35 mm. All of the soft tissue lesions have increased ¹⁸F-FDG uptake with maximum standardized uptake value (SUVmax) ranging from 3.12 to 10.79. Axial CT (E), PET (F), fusion image (G) and axial CT (H), PET (I), fusion image (J) showed two soft tissue mass in the right ventricle and pancreas head with SUVmax of 9.22 and 5.52 respectively. Symmetrical ¹⁸F-FDG uptake in the bilateral neck muscles was considered as physiological uptake. The final pathology of nodule in the left thigh by puncture biopsy supported pseudomyogenic hemangioendothelioma (PMH) with positive desmin, ERG, BRG, IN1, H3K27MES and negative myo D1, SMA, CD34, CD31, P63, CK, EMA, S-100, CD68, ALK detected by immunohistochemical analysis. Pseudomyogenic hemangioendothelioma is an uncommon vascular tumor with the predilection of young males. Pseudomyogenic hemangioendothelioma is mainly involved in the extremities' superficial or deep soft tissues [1], manifesting as multicentric and/or multifocal nodular mass in the dermis, subcutaneous tissue, and muscles [2, 3]. This disease also could cause bone destruction mimicking multiple myeloma or metastasis [4–6]. Other rare sites involving PMH include esophagus [7] oral cavity [8], breast [9], penis [10] and vulva [11]. However, PMH presenting as right ventricle and pancreas involvement is very rare. Pseudomyogenic hemangioendothelioma can have different degrees of ¹⁸F-FDG uptake with SUVmax ranging from 5.3 to 20.7 [2, 4, 5]. It is difficult to differentiate PMH from lymphoma [12, 13], sarcoidosis [14], lgG4-related disease [15], infection [16], metastasis [17]. Our case hints that PMH should be listed as a differential diagnosis when we meet similar appearance on ¹⁸F-FDG PET/CT. Fluorine-18-FDG PET/CT can detect the lesion extent through whole body image and monitor the treatment effect of the disease, like mammalian target of rapamycin (mTOR) therapy, chemotherapy and antiangiogenic agents [2].

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