中华医学会核医学分会
技术与继续教育学组

中华医学会核医学分会第十一届委员会
技术与继续教育学组
系列专家讲座

左侧髂骨病变^{18}F-PET/CT显影一例

专家姓名　袁梦晖
专家单位　唐都医院

- 医学博士，副主任医师、副教授，科室副主任
- 中华医学会核医学分会十一届委员会技术与继续教育学组委员、陕西省核学会理事
- 主持国家自然科学基金面上项目1项、陕西省攻关课题1项，在国内外学术刊物上发表文章30余篇，其中第一（通讯）作者发表SCI收录文章5篇。主译学术专著1部。

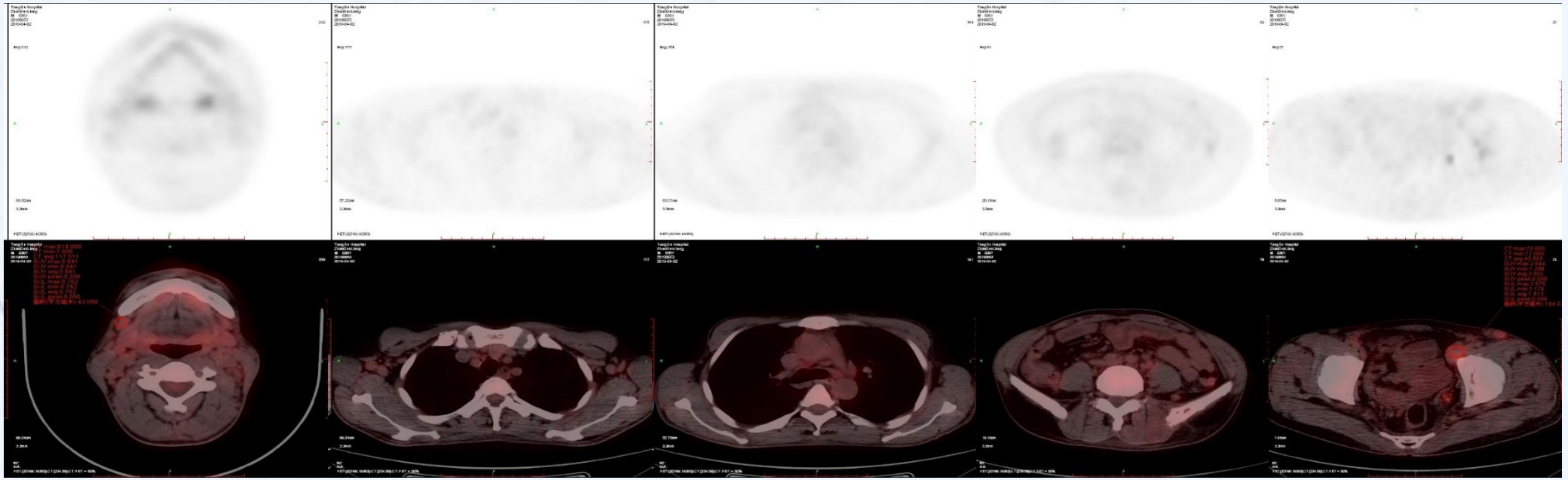
病史摘要

- 男性 36岁
- 主诉：双下肢无力、麻木、行动受限半年余，近期出现上腹部疼痛，无发热等
- 实验室检查：WBC：14.17*10E9/L↑ PLT:205 *10E9/L↑ HGB:157g/L↑

  C反应蛋白：79.30mg/L↑降钙素原：0.54ng/ml↑ 乳酸脱氢酶：212U/L↑

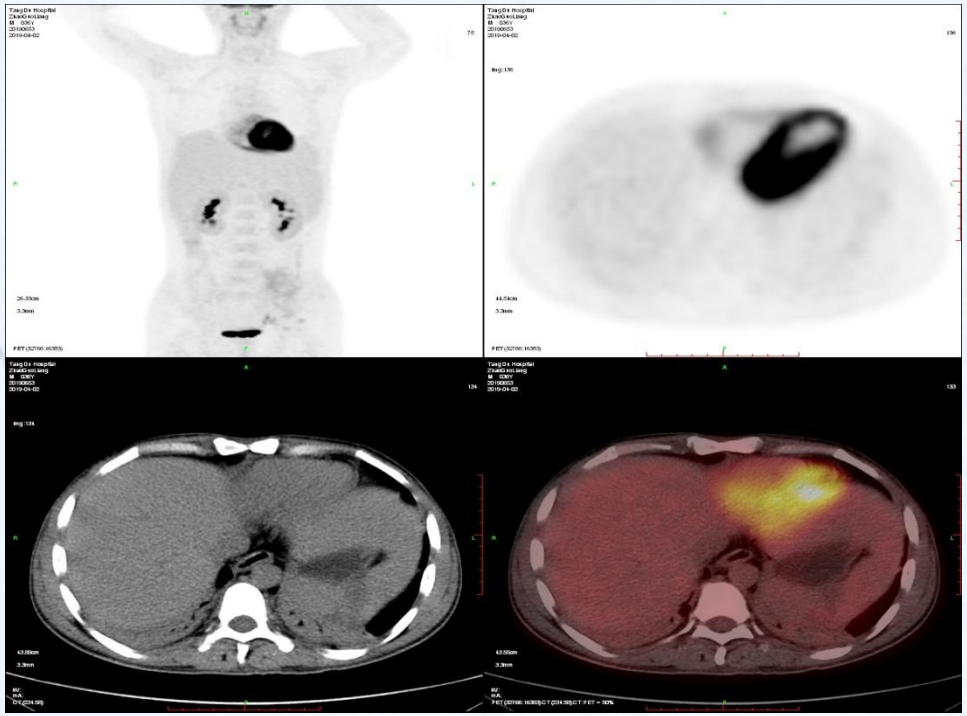
  血清β_2微球蛋白：4.1mg/L↑ 尿便常规、肝肾功、电解质、凝血系列未见异常
- 辅助检查：胸部CT提示双侧胸膜腔少量积液；右肺中叶及双肺下叶炎症；腋窝及纵隔淋巴结肿大

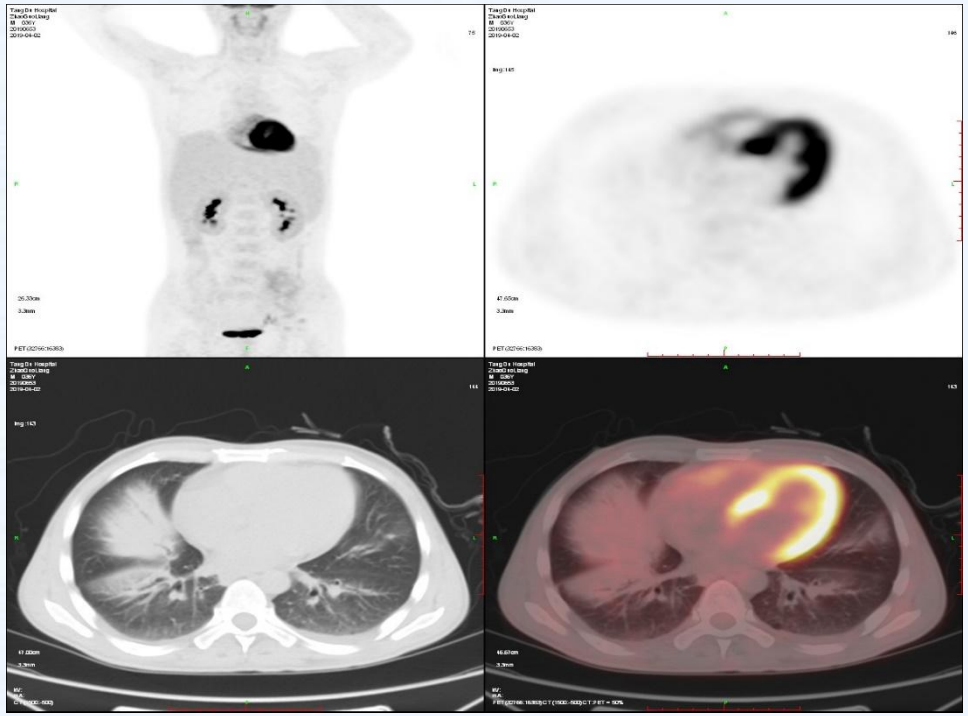
18F-PET/CT图像



双侧颈部、锁骨区、纵隔、腹膜后、双侧髂血管旁、左侧腹股沟区淋巴结肿大，放射性摄取SUVmax：2.04

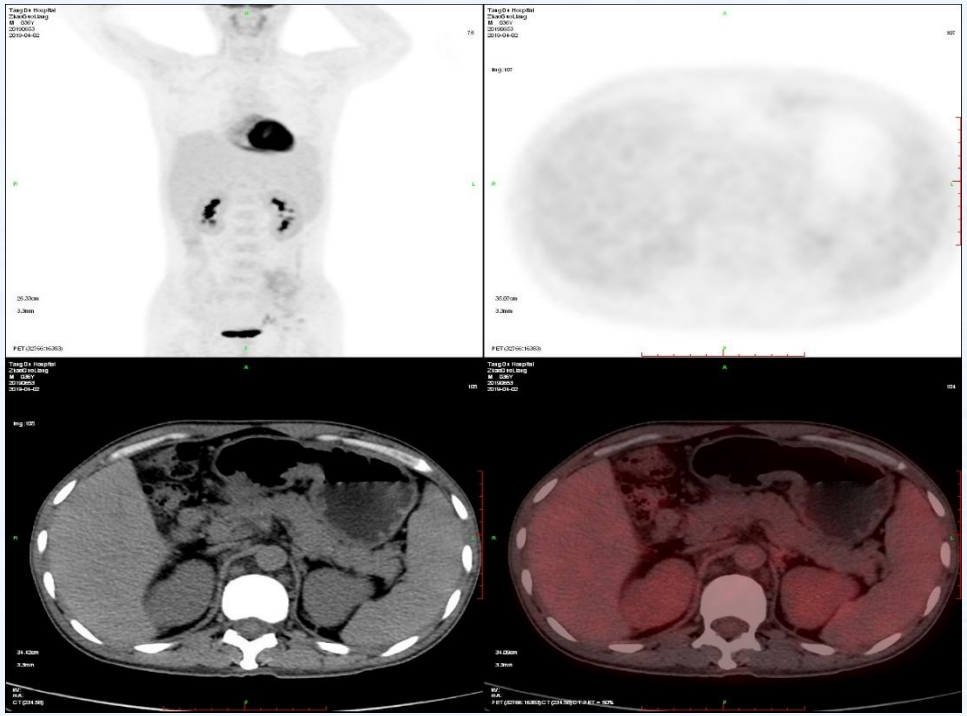
18F-PET/CT图像

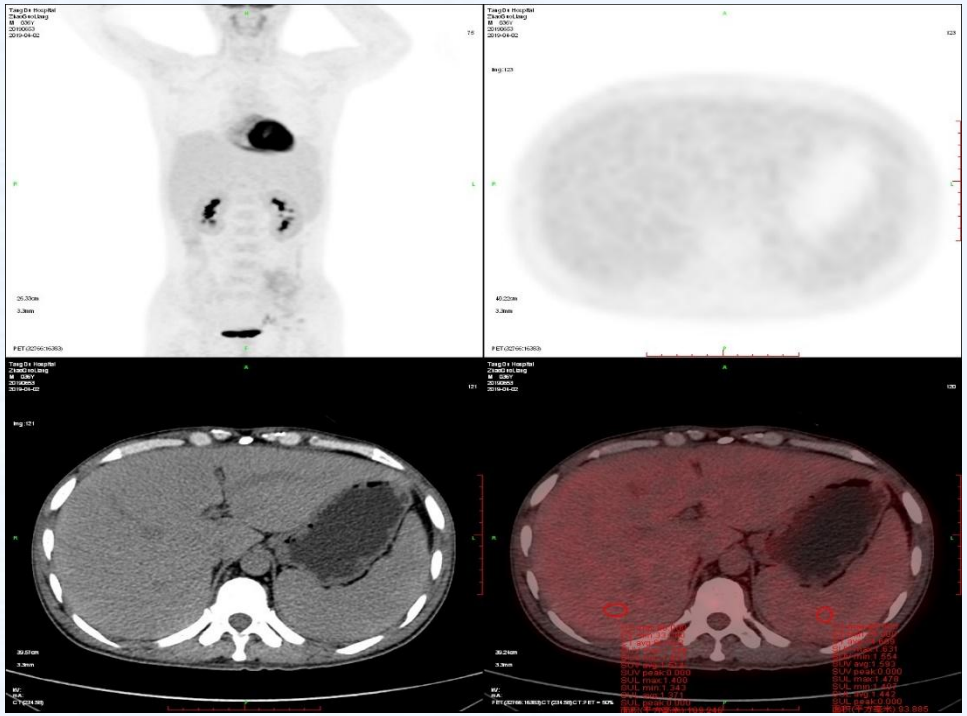




双侧胸膜腔积液，双肺少量渗出

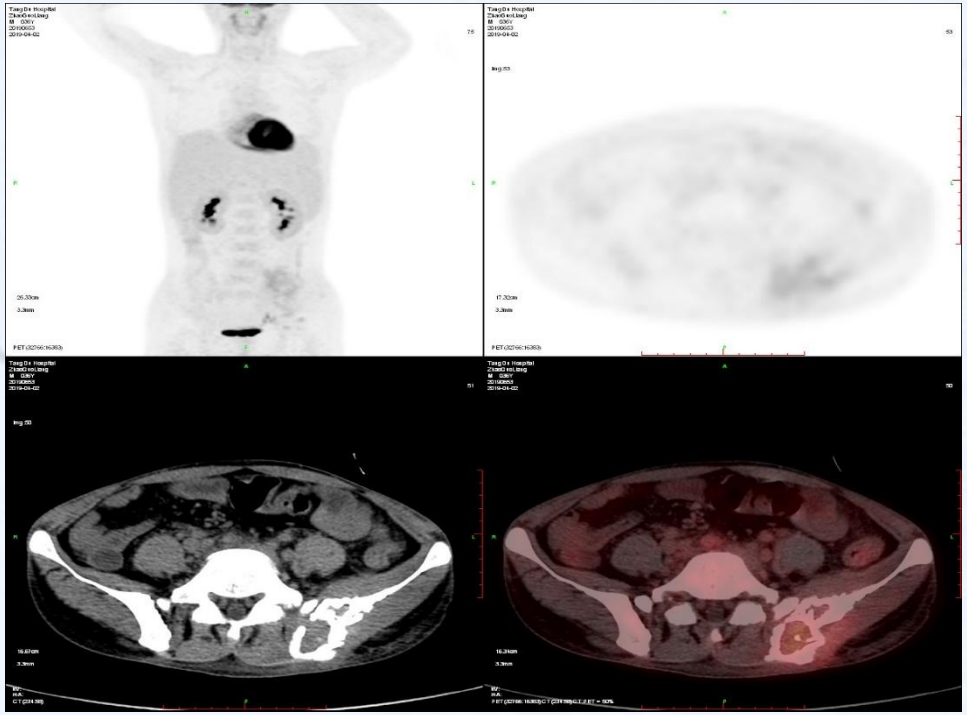
18F-PET/CT图像

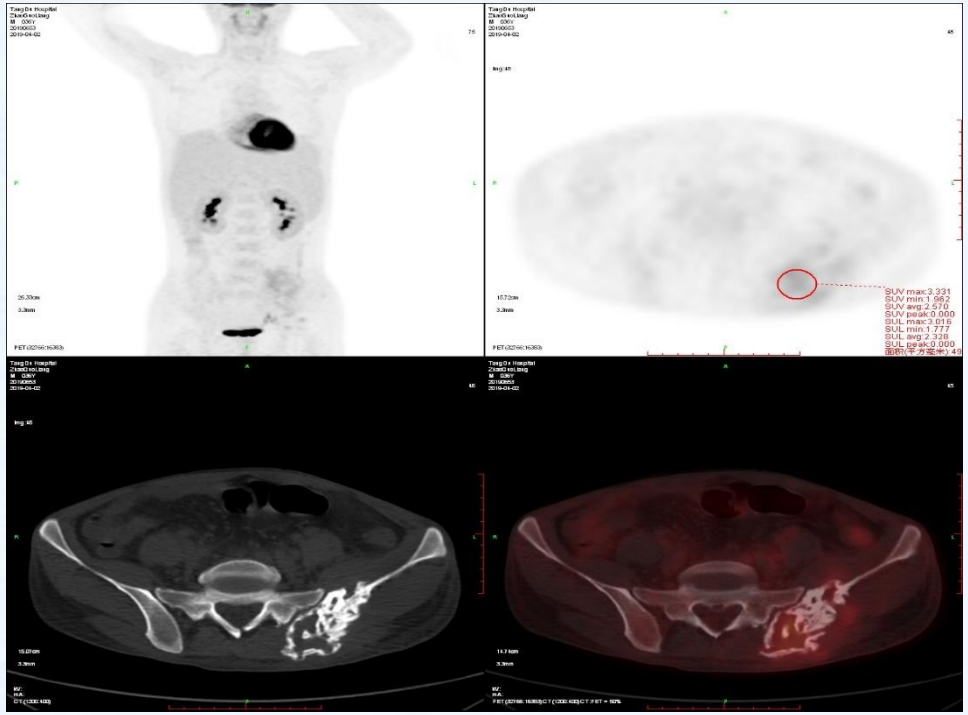




双侧肾上腺体积弥漫性增大，肝脏、脾脏体积增大，肝脏SUVmax：1.54；脾脏SUVmax：1.63

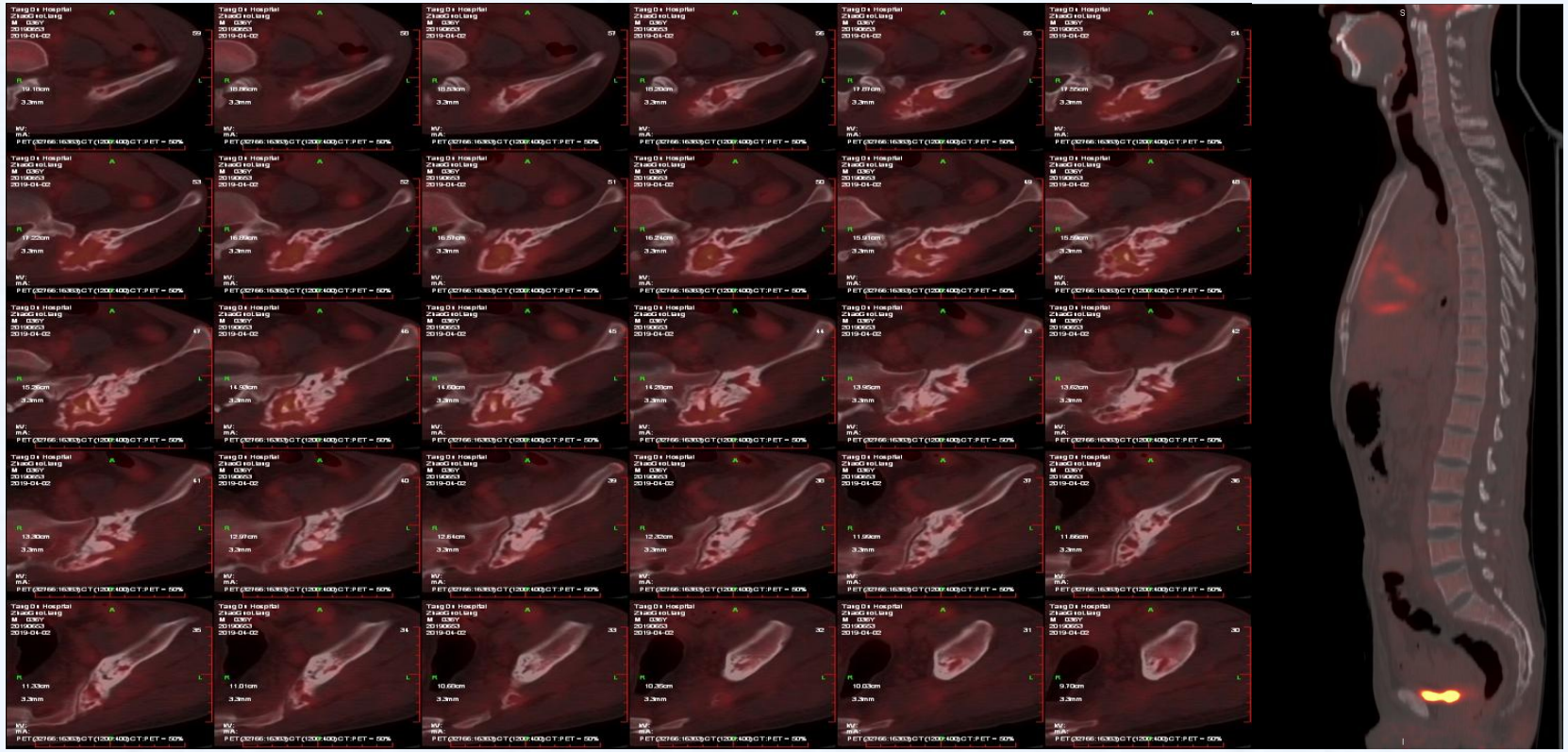
18F-PET/CT图像





左髂骨混合性骨破坏，放射性摄取最高处位于溶骨部位：SUVmax：3.33

18F-PET/CT图像



左侧髂骨病变及脊柱矢状位融合图像骨窗

术后病理活检及相关检查

右颈部淋巴结活检	• 倾向淋巴结Castleman病（多中心型 透明血管型） • 免疫组化 PAX-5(+)，CD79a，CD20(+)，CD3(+)，CD45RO(+)，CD10(-)，CD5(+)，CD30(-)，CD15(-)，CD68(+)，CD43(+)，BCL-6(滤泡中心+)，MUM1(+)，CyclinD1（-），BCL-2（+），Ki-67（20%+），Lamda（+），Kappa（+），滤泡树突网CD21（+），CD23(+)
左侧髂骨活检	• 支持浆细胞瘤 • 免疫组化 LCA(++)，MUM1(核+++)，CD138(+++)，CD38(++)，Kappa（-），Lambda（++），CD45RO，CD3(-)，CD43(+)，CD5(-)，CD20(-)，PAX-5(-)，CD79a（-），CD23(-)，CD10(-)，BCL-6(核+)，BCL-2（++），CyclinD1（-），TDT（-），Ki-67（30%+）
骨髓检活	• 骨髓细胞增生大致正常，淋巴细胞未见增多，嗜酸性粒细胞易见，纤维组织轻度增生

VEGF基因检测	• >8000.0pg/ml（0-142）
骨髓涂片	• 骨髓增生活跃，浆细胞易见，可见双核浆细胞及三五成丛分布，成熟浆细胞占3.2%
流式免疫分型	• 在CD45/SSC点圈上设门分析，所选抗体组合中未分见明显异常细胞
染色体	• 46，XY[5]，分析5个分裂相，未见克隆性结构及数目异常

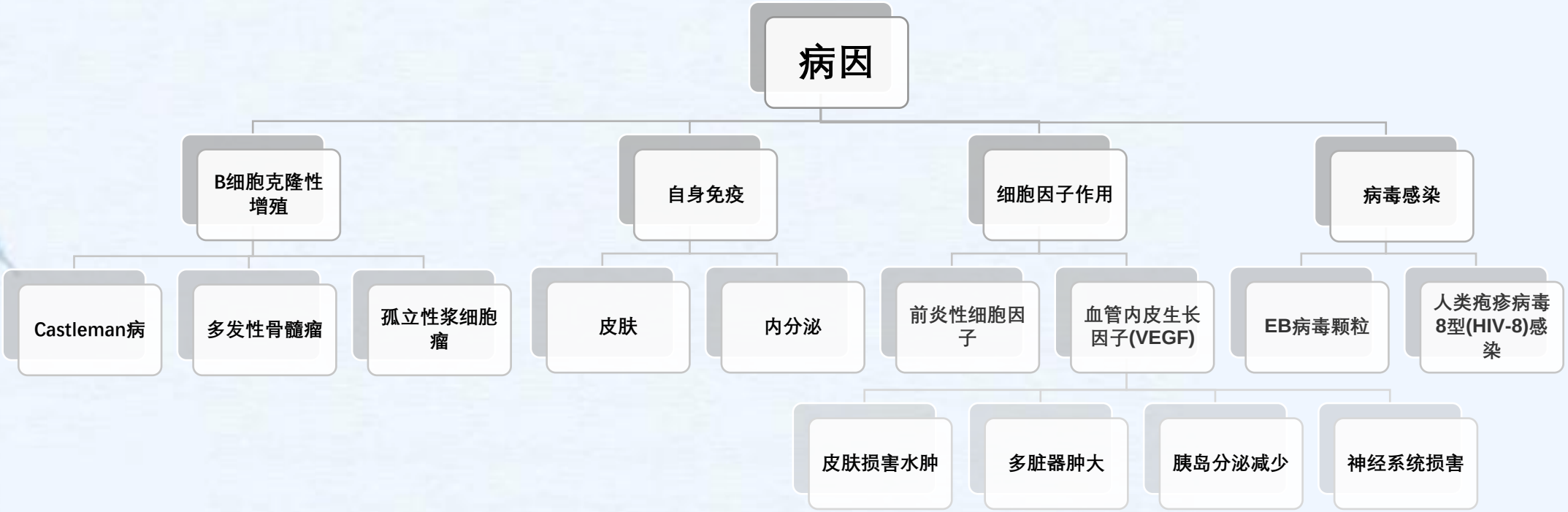
临床诊断：POEMS综合征合并Castleman病（多中心型 透明血管型），浆细胞瘤（左侧髂骨）

POEMS综合征

是一组以多发性周围神经病和单克隆浆细胞增生为主要表现的临床症候群

- **多发性神经(Polyneuropathy)**
- **脏器肿大(Organomegaly)**
- **内分泌病(Endocrinopathy)**
- **M蛋白异常(M protein)**
- **皮肤改变(Skin changes)**

具体病因尚不十分明确，目前有以下四种



2007年诊断标准

强制性主要标准

1.多发性神经病

2.单克隆浆细胞增生性疾病

主要标准

3.硬化性病变

4.Castleman病

5.清或血浆血管内皮生长因子升高

次要标准

6.脏器肿大（脾肿大、肝肿大），或淋巴结肿大

7.血管外容量超负荷（外周水肿，腹水，或胸腔积液）

8.内分泌疾病（甲状腺、垂体、肾上腺、性腺、胰腺）

9.皮肤改变（色素沉着过度，多毛，肾小球样血管瘤，多血症，手 足发绀，脸红，白指甲）

10.视乳头水肿

11.血小板增多症

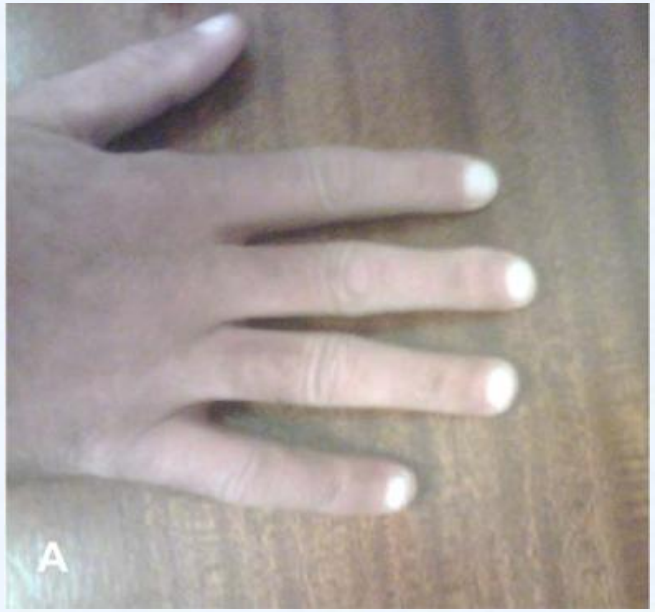
其他症状和体征

肥胖、体重减轻、多汗、肺动脉高压/限制性肺病、血栓形成、腹泻、维生素B12降低

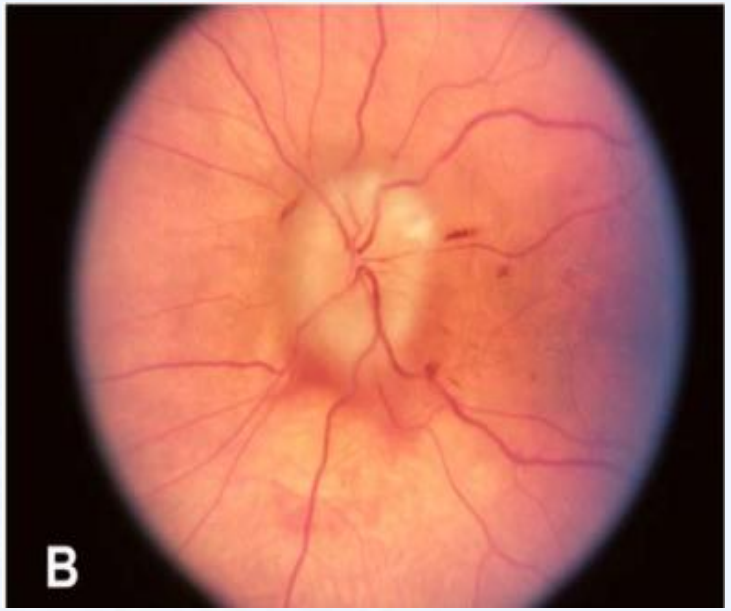
TABLE 1 Criteria for the diagnosis of POEMS syndrome[a]

Mandatory major criteria	**1. Polyneuropathy (typically demyelinating)**
	2. Monoclonal plasma cell-proliferative disorder (almost always λ)
Other major criteria	3. Castleman disease[a]
(one required)	4. Sclerotic bone lesions
	5. Vascular endothelial growth factor elevation
Minor criteria	6. Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy)
	7. Extravascular volume overload (edema, pleural effusion, or ascites)
	8. Endocrinopathy (adrenal, thyroid,[b] pituitary, gonadal, parathyroid, pancreatic[b])
	9. Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomata, plethora, acrocyanosis, flushing, white nails)
	10. Papilledema
	11. Thrombocytosis/polycythemia[c]
Other symptoms and signs	Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B_{12} values

POEMS, polyneuropathy, organomegaly, endocrinopathy, M protein, skin changes.
The diagnosis of POEMS syndrome is confirmed when both of the mandatory major criteria, one of the three other major criteria, and one of the six minor criteria are present.
[a]There is a Castleman disease variant of POEMS syndrome that occurs *without* evidence of a clonal plasma cell disorder that is not accounted for in this table. This entity should be considered separately.
[b]Because of the high prevalence of diabetes mellitus and thyroid abnormalities, this diagnosis alone is not sufficient to meet this minor criterion.
[c]Approximately 50% of patients will have bone marrow changes that distinguish it from a typical MGUS or myeloma bone marrow [45]. Anemia and/or thrombocytopenia are distinctively unusual in this syndrome unless Castleman disease is present.



白指甲



视乳头水肿

Journal of Neurology (2019) 266:1067–1072
https://doi.org/10.1007/s00415-019-09233-z

ORIGINAL COMMUNICATION

Check for updates

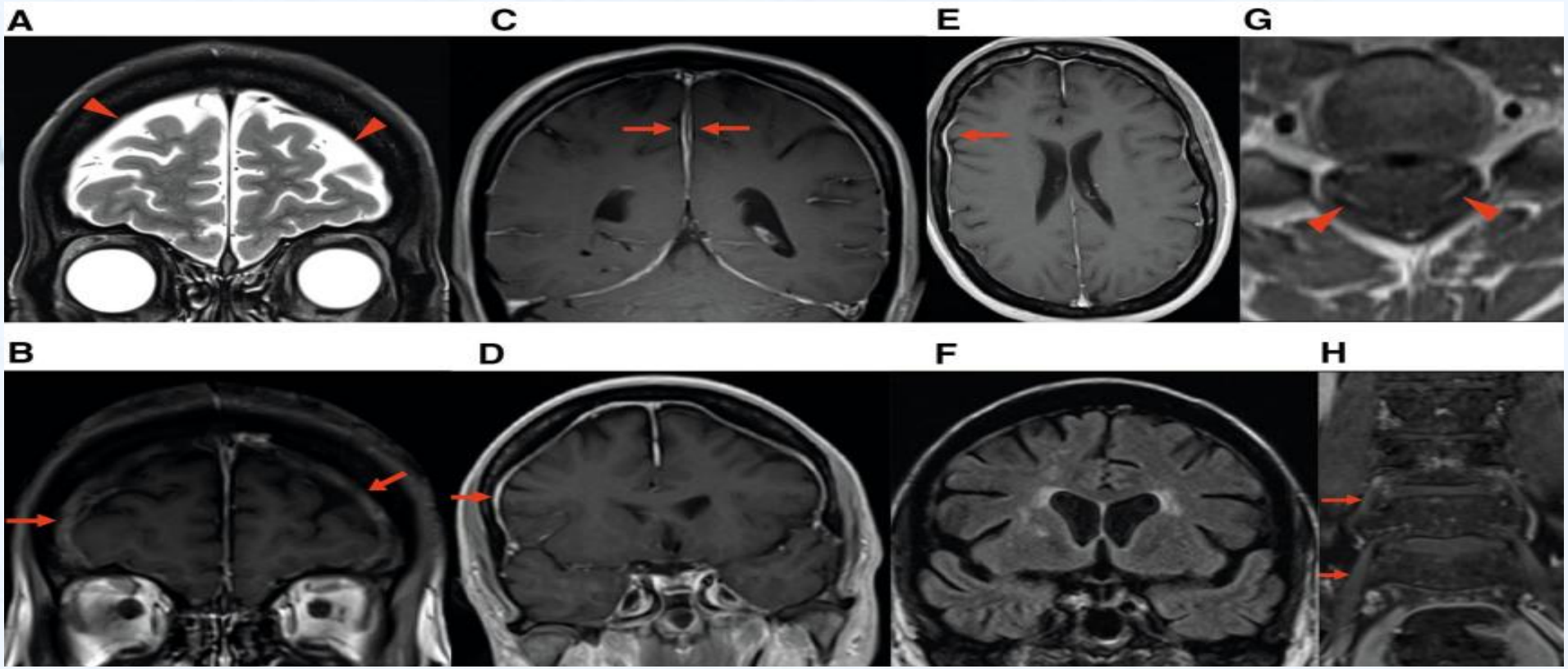
Frequent central nervous system, pachymeningeal and plexus MRI changes in POEMS syndrome

Oliver J. Ziff[1,2] · Chandrashekar Hoskote[3] · Stephen Keddie[1] · Shirley D'Sa[4] · Indran Davangnanam[3] · Michael P. T. Lunn[1,2,5]

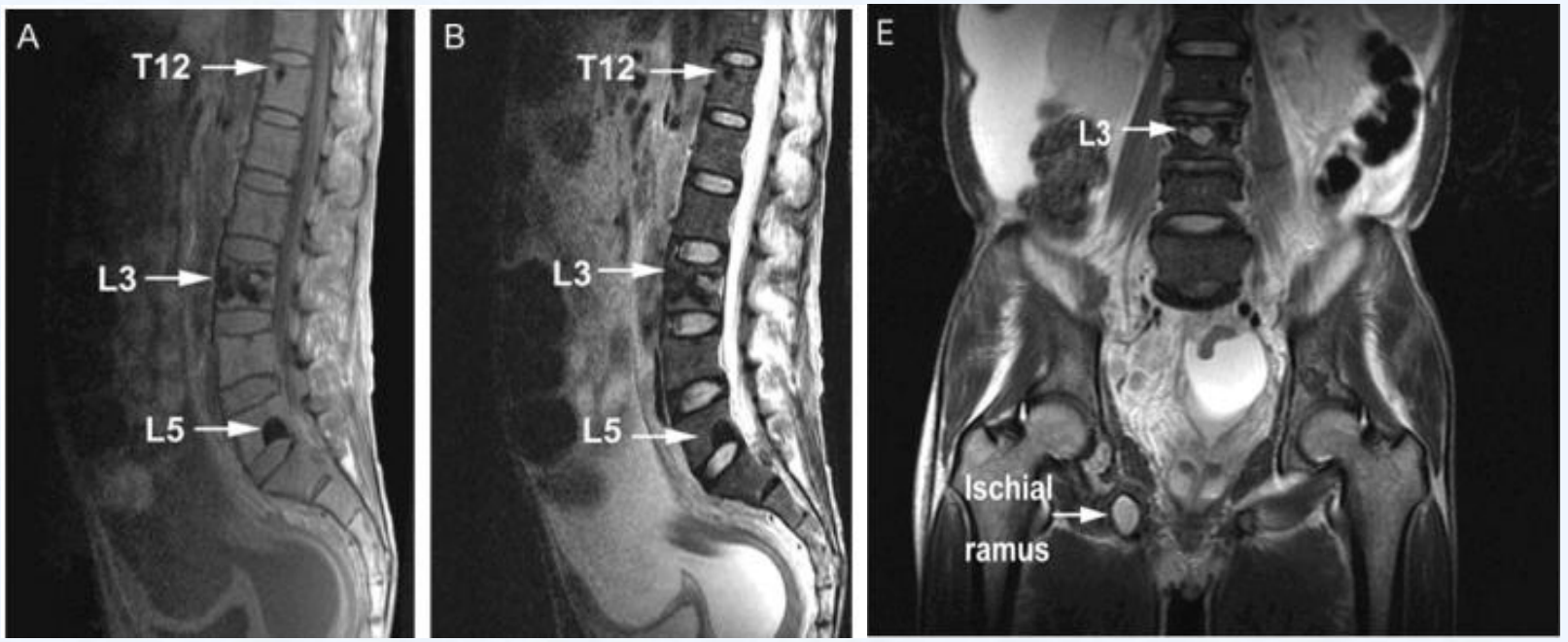
Received: 22 November 2018 / Revised: 3 February 2019 / Accepted: 5 February 2019 / Published online: 12 February 2019

MRI 神经系统改变

POEMS患者外周神经症状发生率为100%，肌电学研究发现传导速度减慢，但其中枢神经系统（CNS）的MRI还没有得到广泛的研究。该文章回顾性评估欧洲最大单中心POEMS综合征患者的CNS MRI。77例经正式诊断为POEMS的患者中，41例为MRI脑扫描，29例为MRI脊柱扫描，以慢性炎症性脱髓鞘性多发性神经病（CIDP）作为对照组，观察硬脑膜平扫及增强的改变情况，发现大脑凸面和大脑镰的硬脑膜呈不均匀性增厚且强化，在T1增强冠状位更易观察到，脊柱的臂丛和腰骶丛软脑膜增厚。

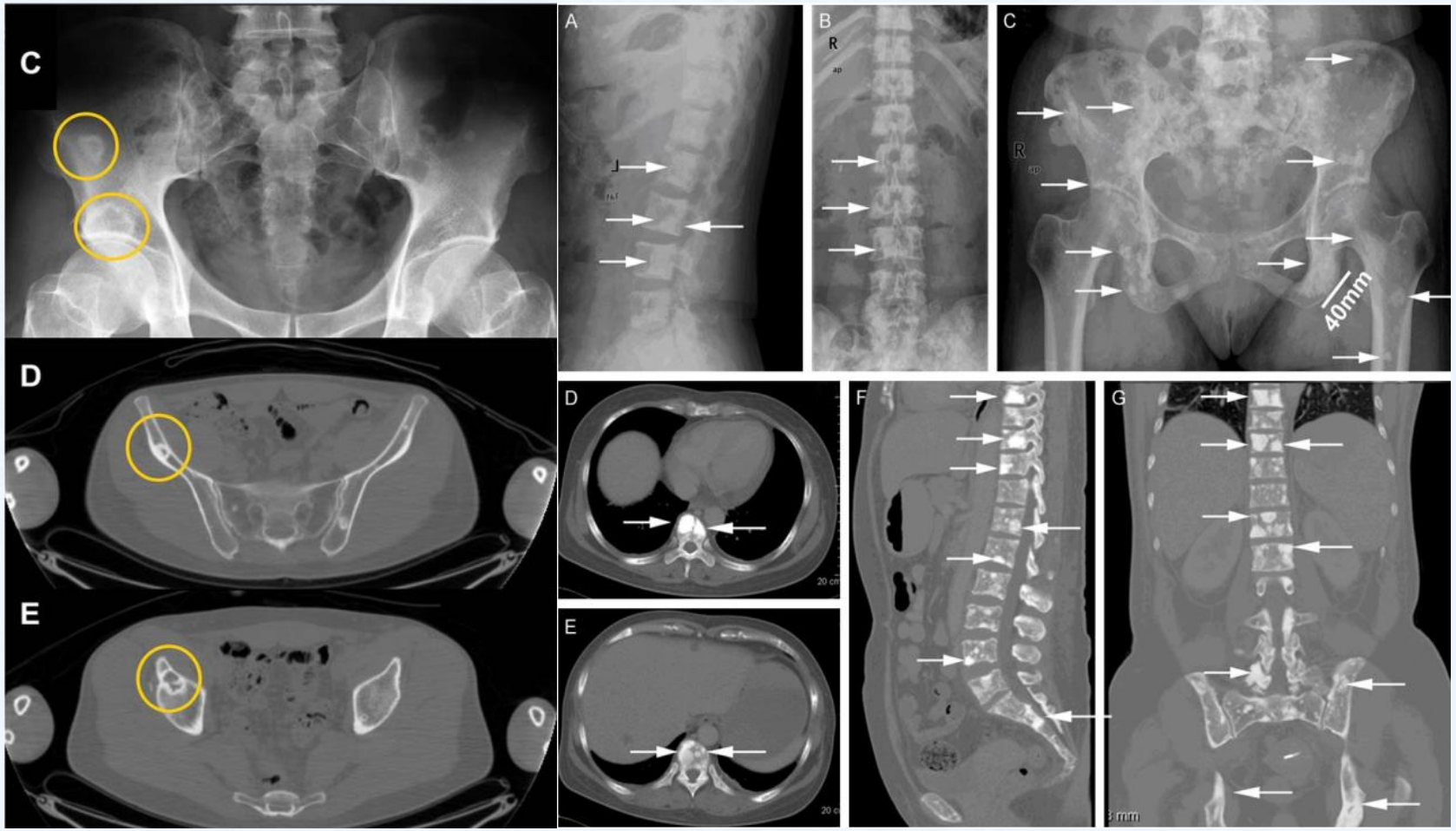


MRI骨骼系统改变



硬化性为主病变T1及T2为低信号　溶骨性病变DWI T2信号增高

X线及CT影像改



X线及CT对四肢骨、颅骨和锁骨病变显示具有优势，可以清晰显示病变的范围和轮廓

PET/CT对POEMS综合征诊断特征

Characterizing POEMS Syndrome with ^{18}F-FDG PET/CT

Qingqing Pan[1], Jian Li*[2], Fang Li*[1], Daobin Zhou[2], and Zhaohui Zhu[1]

[1]Department of Nuclear Medicine, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China; and [2]Department of Hematology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

POEMS (polyneuropathy, organomegaly, endocrinopathy, M protein elevation, and skin changes) syndrome is a rare paraneoplastic syndrome caused by an underlying plasma cell disorder. The patients usually present with multisystemic involvement. Thus, we performed a study to investigate the role of ^{18}F-FDG PET/CT in characterizing POEMS syndrome. **Methods:** Ninety-one untreated patients with proven or suspected POEMS syndrome were recruited to undergo ^{18}F-FDG PET/CT. Features of bone lesions, lymphadenopathy, hepatomegaly or splenomegaly, bone marrow, and serous cavity effusion were examined, and 15 patients were followed up with PET/CT scans 3 mo after therapy. **Results:** Of the 90 patients diagnosed with POEMS syndrome, there were 140 ^{18}F-FDG–avid bone lesions. These lesions were frequently found in the pelvis, and most showed mixed characteristics. Four patients showed enlarged and ^{18}F-FDG–avid lymph nodes. Sixty-five patients had hepatomegaly or splenomegaly. Some of them had hypermetabolic spleen and bone marrow. Forty-six patients had serous cavity effusion. Five male patients had gynecomastia. Three months after therapy, ^{18}F-FDG–avid bone lesions showed decreased metabolism. **Conclusion:** ^{18}F-FDG PET/CT is a useful tool for the evaluation of patients with suspected POEMS syndrome. ^{18}F-FDG PET/CT may contribute to the diagnosis, evaluation, and follow-up of patients with POEMS syndrome by providing systematic findings of bone lesions, lymphadenopathy, liver or spleen involvement, serous cavity effusion, and the metabolic status of the lesions.

Key Words: FDG; PET; POEMS syndrome

J Nucl Med 2015; 56:1334–1337
DOI: 10.2967/jnumed.115.160507

survey and technetium scintigraphy is unsatisfactory (*3*). MR imaging has been used to evaluate patients with POEMS syndrome and can also be used to evaluate the sciatic nerve (*4*). However, the availability of and time required for whole-body MR imaging limits its application. Alberti and Montoriol et al. reported several cases of POEMS syndrome evaluated by ^{18}F-FDG PET/CT, which manifested as ^{18}F-FDG–avid bone lesions (*5,6*). ^{18}F-FDG PET/CT facilitates the diagnosis and follow-up of several hematologic diseases, including lymphoma and multiple myeloma (*7–10*). Considering the multisystemic involvement of POEMS syndrome, the extraskeletal manifestation on PET/CT is still unclear and of interest to researchers. Thus, we recruited a cohort of POEMS syndrome patients and investigated the role of ^{18}F-FDG PET/CT in characterizing POEMS syndrome.

MATERIALS AND METHODS

Patients

From January 2013 to December 2014, 91 patients (34 women, 57 men; mean age ± SD, 49.09 ± 10.93 y) with proven or suspected POEMS syndrome were recruited at Peking Union Medical College Hospital to undergo ^{18}F-FDG PET/CT. Among them, 90 patients were finally diagnosed as having POEMS syndrome according to the 2007 Mayo Clinic Criteria (*11*). Fifteen patients underwent additional PET/CT scans 3 mo after treatment, and serum vascular endothelial growth factor (VEGF) levels were also evaluated. Biopsy revealed that 1 patient with suspected POEMS syndrome actually had amyloidosis. The institutional review board of Peking Union Medical

在诊断标准中，硬化性骨病变、脏器肿大、浆膜腔积液可通过PET-CT成像方式显示

18F-FDG在骨破坏具有优势，骨盆多发，混合型多见（可见髓腔扩大），其次为硬化型，骨溶解型最少但摄取更为明显

骨骼病变治疗后葡萄糖代谢的降低早于CT变化改变

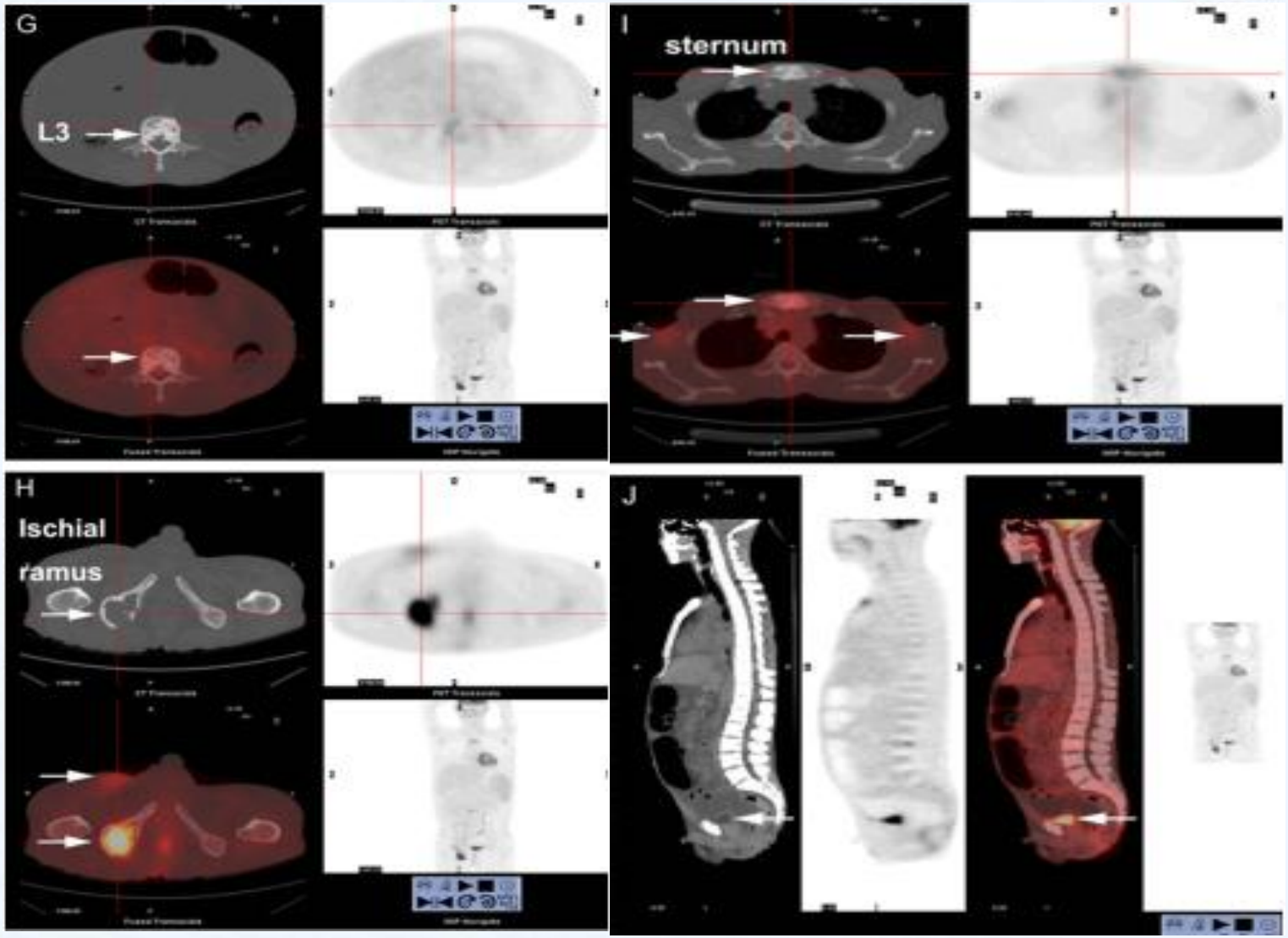
中轴骨代谢增高的原因是反应性的改变，与多发性骨髓瘤的浆细胞直接浸润不同

淋巴结Castleman病，
最多的为透明血管型

随着血清VEGF降低，18F-FDG摄取值降低，表明完全或部分缓解，提示18F-FDG PET/CT可能在疾病严重程度的早期评估中起作用

当18F-FDG PET-CT显影同时有
骨病变、淋巴结病、肝脏或脾脏肿大、多浆膜腔积液特征，高代谢性脾脏和中轴骨，鉴别诊断应包括POEMS

POEMS 综合征18F-FDG PET/CT骨显像



治疗：自体外周血造血干细胞移植是目前治疗POEMS综合征的有效方法

小　结

当^{18}F-FDG PET/CT显影同时有硬化性骨病变、淋巴结肿大、肝脏或脾脏肿大、多浆膜腔积液特征，高代谢性脾脏和中轴骨，且临床症状包括周围神经系统病变、皮肤及内分泌异常时，鉴别诊断应包括POEMS综合征。

参考文献

1.Ziff O J , Hoskote C , Keddie S , et al. Frequent central nervous system, pachymeningeal and plexus MRI changes in POEMS syndrome[J]. 2019.

2.Dispenzieri A . POEMS syndrome: 2011 update on diagnosis, risk-stratification, and management[J]. American journal of hematology, 2011, 86(7):591-601.

3.Shi X F , Hu S D , Li J M , et al. Multimodal imaging and clinical characteristics of bone lesions in POEMS syndrome[J]. International Journal of Clinical and Experimental Medicine, 2015, 8(5):7467-7476.

4.Pan Q , Li J , Li F , et al. Characterizing POEMS Syndrome with 18F-FDG PET/CT.[J]. Journal of Nuclear Medicine Official Publication Society of Nuclear Medicine, 2015, 56(9):1334.