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(**收稿**:2013-06-24;**修回**:2013-07-25)
(编辑 张 维)

个案与短篇

文章编号:1000-5404(2013)23-2520-01

系统性红斑狼疮误诊再生障碍性贫血1例

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[**关键词**] 系统性红斑狼疮;狼疮性肾炎;再生障碍性贫血;三系减少
[**中图法分类号**] R593.241 [**文献标志码**] B

系统性红斑狼疮(systemic lupus erythematosus, SLE)是一种全身性自身免疫病。活动性SLE常有全血细胞减少,由于免疫功能紊乱及长期免疫抑制剂的应用,易继发感染,出现发热、腹泻等症状,严重时常危及生命[1]。再生障碍性贫血(aplastic anemia,AA)是一种获得性骨髓造血功能衰竭症,主要表现为骨髓造血功能低下、全血细胞减少和贫血、出血、感染综合征[2]。在临床症状不典型且合并多系统表现时很难鉴别,易误诊、漏诊,现将我科收治的1例SLE误诊AA报告如下。

1 临床资料

患者,中年女性,45岁,因"反复头昏、乏力、水肿2年,腹泻、发热1月"于2013年7月15日入住当地医院血液科。患者2年前因头昏、乏力、心慌、双下肢水肿,入当地医院血液科住院治疗。经查血常规白细胞计数 1.1×10^9/L,红细胞计数 2.20×10^{12}/L,血红蛋白61 g/L,血小板计数 37×10^9/L;肝功白蛋白15.7 g/L;骨髓活检提示骨髓增生减低,诊断"再生障碍性贫血"。复方皂矾丸、十一酸睾酮胶囊服用1个月,其后自行停药未随访,水肿间断出现。院外一直按"再生障碍性贫血"进行治疗,效果不明显。贫血及血小板低下一直未纠正。1个月前,无明显诱因出现全身水肿、腹泻、发热,最高体温39 ℃,无畏寒、咳嗽、胸痛,无腹痛、恶心、呕吐。到当地医院门诊查血常规白细胞计数 3.52×10^9/L、红细胞计数 1.98×10^{12}/L、血红蛋白50 g/L、血小板计数 36×10^9/L。给予对症处理,未再发热、腹泻,但自觉症状及水肿无好转,入住当地医院血液科。查体:慢性病容,贫血貌,颜面眼睑重度水肿,面色苍白,心肺阴性,腹软,肝脾未扪及,移动性浊音阳性,四肢重度水肿;实验室检查:白细胞计数 1.27×10^9/L,红细胞计数 2.10×10^{12}/L,血红蛋白61 g/L,血小板计数 27×10^9/L;尿常规蛋白+++,潜血+++,白细胞++;白蛋白12.1 g/L,总蛋白38.4 g/L。结合既往病史,诊断再生障碍性贫血。给予输注红细胞悬液,水肿进一步加重;血浆白蛋白12.1 g/L,24 h尿蛋白定量5.14 g。患者于2013年7月18日来我科治疗。入院后查补体 C_3 0.11(参考值0.9~2.1),尿蛋白+++,抗核抗体谱SS-A阳性、Ro-52阳性、抗核抗体核型核均质型、核小体阳性、组蛋白阳性、抗核抗体滴度1∶1 000,肾功能正常;腹部超声:腹盆腔积液;肾活检病理结果:狼疮性肾炎Ⅲ型。结合既往病史,诊断"系统性红斑狼疮、狼疮性肾炎"。予以激素免疫抑制(住院期间暂未用环磷酰胺,因其白细胞低,一般情况较差。告知门诊随访据情况加此药)、降尿蛋白、保肾、改善循环。7月30日复查血常规白细胞计数 10.23×10^9/L,红细胞计数 2.35×10^{12}/L,血红蛋白65 g/L,血小板计数 80×10^9/L;肝功白蛋白21 g/L,总蛋白46.9 g/L。患者精神状态明显好转,头昏、乏力症状明显减轻,全身水肿明显消退,体质量较入院时减轻10 kg,于8月7日出院,院外继续治疗,效果良好。

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[**优先出版**] http://www.cnki.net/kcms/detail/51.1095.R.20131104.0957.002.html(2013-11-04)

决策中,应遵循循证医学原则,权衡 PPI 的使用对病患的利弊,慎重考虑是否应用 PPI,以避免 PPI 对抗感染治疗效果造成影响。本研究属于回顾性分析,因为 ICU 患者病情较重,较多的患者都使用了 PPI,故对照组入选病例较少。下一步应更广泛地收集病例,以便明确地分析 PPI 与 HAP 转归的影响。

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(**收稿**:2013-06-19;**修回**:2013-08-07)

(编辑 张 维)

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2 讨论

本例患者表现为贫血、白细胞减少及血小板减少,按 AA 治疗了 2 年,效果不好。经肾活检及抗核抗体谱结果诊断为“系统性红斑狼疮、狼疮性肾炎”,给予激素治疗后效果明显。狼疮患者三系减少常见,主要由狼疮活动细胞毒型变态引起细胞溶解所致[3];少数 SLE 患者存在针对造血干细胞的高效价自身抗体也可致三系减少[4]。SLE 的骨髓象特点:多为增生性骨髓象,骨髓红系及巨核细胞数量增生良好,Ret/Nc 不降低;而 AA 的骨髓象特点表现为:多部位增生减低,三系造血细胞减少,淋巴及非造血细胞相对增多,Ret/Nc 降低。SLE 是一种复杂的自身免疫性疾病,以出现多种自身抗体为特征[5],临床表现多样化,尤其在早期没有特异性特征,可以仅仅表现为 1~2 个系统受损[6],故极易误诊。很多患者早期表现不典型,自身抗体仅表现为未分化型结缔组织病[7]。诊断主要依据详细病史询问、仔细的体格检查及实验室检查,其中自身抗体谱的筛查及追踪尤为重要。因此临床医师应加强业务学习,提高对本病的警惕性,对临床治疗进行全面仔细的综合分析,尤其遇到治疗效果不好的患者,要深究原因,考虑系统性疾病的可能。广大临床医师应重视三系减少的鉴别诊断,特别是血液科医师应了解 SLE 的血液学改变,帮助临床诊断,减少误诊及漏诊。

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(**收稿**:2013-09-24;**修回**:2013-10-15)

(编辑 龙 亮)