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个案与短篇

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以纤维蛋白溶解综合征为主要表现的主动脉夹层1例

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[**关键词**] 纤维蛋白溶解综合征;原发性;主要表现;主动脉夹层
[**中图法分类号**] R543.16 [**文献标志码**] B

1 临床资料

患者,男性,81岁,因发现全身瘀斑、血细胞减少5 d,发热1 d于2010年8月25日入院。入院前5 d因"突发头晕、呕吐、右上肢乏力"就诊当地医院,查头颅MRI示:多发腔隙性梗死;予丹参酮ⅡA改善循环处理,头晕、呕吐等症状缓解,但出现皮肤粘膜多处瘀斑,查血常规:WBC 8.6×10^9/L,HGB 112 g/L,PLT 76×10^9/L;凝血全套:Fib 0.67 g/L,PT和APTT等指标正常;继续丹参酮ⅡA治疗,皮肤瘀斑范围扩大,并出现发热,体温最高37.5 ℃,无畏冷,复查血常规:WBC 9.3×10^9/L,Hb 77 g/L,PLT 87×10^9/L;遂转诊我院。既往发现"高血压病"6个月,口服降压药治疗,血压控制不详,此次发病前停用降压药物。查体:体温37.5 ℃,脉搏80次/min,呼吸22次/min,血压150/80 mmHg。神志清楚,中度贫血貌,全身皮肤黏膜苍白,双手掌、右侧上臂、左侧臀部可见大片瘀斑。巩膜轻度黄染,胸骨无压痛。双肺呼吸音清,未闻及干湿性罗音。心率80次/min,心律绝对不齐,心音强弱不等,各瓣膜听诊区未闻及病理性杂音。腹平软,无压痛、反跳痛,肝脾肋下未及。双下肢无水肿,双侧病理征均未引出。8月26日辅助检查:血常规:WBC 11.1×10^9/L,HGB 82 g/L,PLT 169×10^9/L,RET 10.2%;凝血功能:PT 14.2 s(对照组11.0~16.0 s),APTT 24.6 s(对照组21.0~37.0 s),FIB 1.86 g/L(对照组2.00~4.00 g/L),D-二聚体:46.20 μg/ml(对照组0.00~0.50 μg/ml)。3P试验:阴性。FDP>20 μg/ml(对照组0~20 μg/ml)。生化检测:ALB 30.7 g/L,TBIL 42.57 μmol/L,DBIL 15.11 μmol/L,IBL 27.46 μmol/L,LDH 240 IU/L,HBDH 259 IU/L。X线胸部平片:心影呈主动脉型,左心缘饱满,向左延伸。蔗糖溶血试验:阴性;传染病3项(包括乙肝病毒抗原、抗梅毒抗体、抗HIV抗体1~2):均阴性;尿Rous试验:阴性;Coombs试验:阴性;骨髓细胞学:有核细胞增生明显活跃,红系增生明显:占68.5%;巨核细胞全片56个,成熟巨核细胞18/25个。提示:增生性贫血。肿瘤标志物:均阴性。血清免疫学检测:均阴性。ANCA:阴性;免疫抗体检查:阴性;抗心磷脂抗体:阴性。肺部CT示:主动脉弓及以下胸主动脉腔内线状高密度影分隔:动脉夹层可能。肺纵膈胸膜CT平扫+增强:扫描所示层面(主动脉弓至双肾上极水平)胸腹主动脉腔可见内膜片分隔呈真假双腔,胸主动脉段双腔增强程度相仿,腹主动脉段双腔呈单腔强化(夹层低于扫描范围)。提示主动脉弓、胸腹主动脉夹层。心脏彩超:①左、右房增大,左室肥厚;②轻度二尖瓣及主动脉瓣返流;③重度三尖瓣返流;④轻度肺动脉高压。9月23日复查凝血功能:PT 13.7 s,APTT 33.6 s,FIB 1.61 g/L。血常规:WBC 7.6×10^9/L,HGB 115 g/L,PLT 147×10^9/L,RET 1.9%。最后诊断:主动脉夹层并发原发性纤维蛋白溶解

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综合征、溶血性贫血。治疗经过:予冷沉淀补充纤维蛋白原,抗纤溶,控制血压、心室率等处理后,血红蛋白、纤维蛋白原升高,皮肤黏膜出血表现明显改善。患者高龄,手术治疗风险大,家属要求保守治疗。随访至今患者多次因纤维蛋白溶解综合征致牙龈出血住院,经抗纤溶、补充纤维蛋白原治疗,病情均缓解。目前继续门诊随访,病情稳定。

2 讨论

患者存在皮肤黏膜出血、血浆纤维蛋白原含量明显减低、FDP水平升高、纤维蛋白碎片D-二聚体增高,结合血小板正常以及3P试验阴性,符合原发型纤维蛋白溶解综合征诊断标准[1]。患者皮肤黏膜有出血表现,存在发热,网织红细胞比例增多、间接胆红素增高,骨髓象提示红细胞系统增生明显,其贫血原因考虑溶血性贫血,Coombs试验阴性,自身抗体谱正常,免疫相关性溶血性贫血证据不足;结合存在纤溶亢进,考虑微血管病性溶血性贫血(MHA)可能性较大,但缺少外周血破碎红细胞证据,MHA尚不能明确。为进一步确定引起纤维蛋白溶解综合征的基础病因,因患者有高血压病,X线胸片示主动脉型心影增宽,予查胸部CT平扫提示:主动脉弓及以下胸主动脉腔内线状高密度影分隔:动脉夹层可能;肺纵膈胸膜CT平扫+增强示:(主动脉弓至双肾上极水平)胸腹主动脉腔可见内膜片分隔呈真假双腔:主动脉弓、胸腹主动脉夹层。该患者最后诊断为主动脉夹层并发原发性纤维蛋白溶解综合征、溶血性贫血。

主动脉夹层是主动脉内膜撕裂,循环中的血液渗入主动脉壁中层形成夹层血肿并沿主动脉壁延伸剥离的严重心血管急症。当中层平滑肌细胞发退变,弹力纤维产生减少和弹力纤维发生囊性坏死时,中层结构受到破坏,其顺应性降低,减弱了主动脉壁对血流切应力的抵抗力,易发生夹层。主动脉壁中层结构的异常,特别是弹力纤维成分的异常,是夹层产生的病理基础[2]。其常见病因包括高血压、遗传疾病、创伤、妊娠、梅毒、血管炎和系统性红斑狼疮等,男性较女性多见。临床表现多以突发剧烈胸、腹、背刀割或撕裂样痛,高血压、心血管症状和神经症状以及压迫症状常见[3]。本例主动脉夹层以原发性纤维蛋白溶解综合征为主要表现较为罕见,且存在皮肤黏膜出血表现,血常规见血小板减少,血浆纤维蛋白原减低,易误诊为弥散性血管内凝血功能障碍(DIC)等血液系统病症。关于主动脉夹层并发原发性纤维蛋白溶解综合征机制尚不清楚,可能与主动脉内膜撕裂形成假腔诱发血栓形成以及组织损伤引发纤维蛋白原等凝血物质消耗有关[4]。因此,对于纤维蛋白溶解综合征且有高血压病患者,应注意主动脉夹层可能。

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