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· 病例报告 ·

重型血友病 A 合并尿毒症低剂量替代治疗下行巨大假性动脉瘤切除术一例

谢燕燕 闫振宇

Operation of huge pseudoaneurysm with low-dose coagulant factor 8 replacement therapy on a severe hemophilia A patient with uremia: a case report Xie Yanyan, Yan Zhenyu

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患者,男,31岁,体重60 kg,因确诊肾衰竭行规律腹膜透析4年伴左前臂肿物2周于2013年1月2日入院。既往史:3岁时因关节肌肉异常出血就医查凝血因子VIII活性(FVIII:C)1%(参考值50%~150%),诊断为重型血友病A。此后间断出现关节、肌肉、皮肤、齿龈等部位出血,仅大出血时应用血浆、冷沉淀或人凝血因子VIII替代治疗,遗留右膝关节畸形。4年前诊断肾功能不全、尿毒症,予以规律透析,间断予以冷沉淀、人FVIII替代治疗维持APTT 50~60 s。入院查体:生命体征平稳,重度贫血貌,左前臂见一约4 cm×10 cm皮肤隆起,

有瘀斑和皮下出血点,有压痛及搏动感,无破溃及血管杂音。心肺无明显异常。腹平坦,脐左旁12 cm处留置腹膜透析管,腹软,无压痛及肌紧张。右膝关节肿胀,活动受限。辅助检查:血常规:HGB 38 g/L,白细胞和血小板正常。生化检查:尿素氮26.9 mmol/L,肌酐997 μmol/L。凝血四项:APTT 76.4 s(参考值28.0~43.5 s)。左上肢血管彩超:左侧桡动脉旁头静脉与桡动脉交汇处远心侧异常无回声假性动脉瘤伴附壁血栓形成。入院诊断:慢性肾功能衰竭(尿毒症期),血友病A重型,假性动脉瘤伴附壁血栓形成,肾性贫血(重度)。给予规律腹膜透析、重组红细胞生成素以及输血支持治疗。多科联合会诊后于局麻下行假性动脉瘤血管切除术及血管修补术。术前查APTT即刻及温浴2 h 1:1正常血浆纠正试验均为正常范围,复查FVIII:C 1.2%,FVIII抑制物阴性。术前1 d予以FVIII 10 IU/kg,复查APTT 52.6 s;手术当日FVIII 30 IU/kg,每12 h 1次;术后第1天FVIII 15 IU/kg,每12 h 1次,复查APTT 45.9 s;术后第2、3天,FVIII 10 IU/kg,每12 h 1次,APTT 57.9 s;术后第4~7天予以小剂量FVIII维持。术中出血约200 ml,术后患者切口愈合良好。术后第10天后复查APTT 71.9 s。

讨论:本例患者因凝血因子应用受限,在围手术期采用低剂量替代治疗方案,APTT未完全控制在正常范围内,但手术效果仍然较好。

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