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·短 篇·

多脏器受累的血管内皮肉瘤 1 例

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[关键词] 血管内皮肉瘤;CT 引导;经皮肺穿刺活检;胸腔镜术
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1 病案摘要

患者,男,58 岁。因咳嗽、咯痰、咯血(有时痰中带血,有时咯纯血)4 月余,逐渐加重伴体重下降 7 公斤而入院。之前一直从事化学涂料的配制工作 5 年。嗜烟 40 余年,每日 20 支左右,已戒 4 月。既往体健。入院时体温 36.8℃,脉搏 94 次/分,呼吸 26 次/分,血压 112/75 mm Hg,体重 63 kg。慢性病容。全身皮肤及巩膜无黄染、皮疹及出血点。结膜苍白,呼吸浅促。气管居中。双肺叩诊呈清音,呼吸音略粗,右下肺可闻及少许湿啰音。右心界略扩大,左心界不扩大,心率 94 次/分,律齐,未闻及杂音及心包摩擦音。腹部无殊。四肢除甲床苍白外无殊。血气分析:pH 7.48,PaO_2 58 mm Hg,$PaCO_2$ 27 mm Hg;血常规检查:WBC 6.22×10^9/L,中性粒细胞 72.2%,淋巴细胞 22%,单核细胞 3.9%,嗜酸细胞 0.5%,嗜碱细胞 0.2%,RBC 2.20×10^{12}/L,Hb 53 g/L,红细胞压积 19.4%,平均血红蛋白量 24.1pg,血小板计数 320×10^9/L。肝肾功能、糖电解质基本正常。痰细菌培养阴性。血清肿瘤标记物检测:前列腺抗原、癌胚抗原、神经烯醇化酶、甲胎蛋白、铁蛋白、CA 199、CA 153、CA 50 均处正常范围,惟 CA 125 略高(48.8 U/ml)。胸部 CT:两肺弥漫性磨玻璃样密度增高,以及多发圆形结节状阴影,直径约 5~15 mm 不等,双侧少量胸腔积液,纵隔淋巴结肿大,右心房增大。心电图检查未见异常。腹部 B 超提示胆囊息肉,余无殊。心脏彩超未提示异常。支气管镜检查示双侧支气管出血,管腔尚通畅,未见新生物。肺泡灌洗液未找到肿瘤细胞。入院后予糖皮质激素(先后用地塞米松和甲基强的松龙)冲击疗法、垂体后叶素止血、输血、抗感染等治疗,病情一度好转,气急缓解,咯血停止。胸部 CT 复查原两肺磨玻璃样变显著减轻,但胸腔积液增多,双肺结节变化不大,右心及心包占位。经皮肺穿刺活检病理提示双肺含铁血黄素沉着,肺泡增生未见异型细胞。胸腔穿刺引流出血性胸水,脱落细胞检查未查到肿瘤细胞。胸腔镜检查壁层胸膜未见明显异常,脏层胸膜可见血肿样及新生物样结节,直径 3~5 mm 不等。活检病理示血管内皮肉瘤(低度恶性)。后行胸部及腹部增强 CT 示:右心房及心包占位,肺部结节明显增大增多,双侧胸水增多。另见肝脏多发占位,左侧肋骨破坏。左侧肋骨破坏处活检,病理示血管内皮肉瘤,免疫组化提示与肺组织同源,CD31 和 CD34 表达阳性。其后患者反复咯血及血性胸水反复增多,呼吸困难逐渐加重,床边胸片示:两肺病变进展,右心增大显著。入院后 52 天死于呼吸衰竭(由于患方拒绝而未做尸检),距发病时约 6 个月。

2 讨 论

血管内皮肉瘤(angiosarcoma)是一种病因不明、源于血管内皮的恶性肿瘤,临床少见。血管内皮肉瘤常见的原发灶有心脏、皮肤和乳房等,肺原发者少见。转移灶可累及心包、肝、脾、肾、肾上腺、骨和脑等,但同时累及心、肺、肝、骨等多脏器者罕见。侵及肺部的血管内皮肉瘤其主要症状无特异性,有咯血、咳嗽、胸痛、体重减轻等。胸部 X (下转第 234 页)

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(上接第 229 页)线检查提示肺部多发结节、空洞、气胸和胸腔积液(多为胸膜下病灶破裂所致)。无特异性血清学肿瘤标记物,诊断有赖于活检之病理学证据。其治疗策略是:孤立病灶多以手术切除,而多发和转移病灶则以放疗化疗等综合治疗,但其疗效不佳[1-6]。

本例病因不明。有报告氯乙烯可致肝血管内皮肉瘤[7]。根据患者职业史,推测其病因可能与长期吸入化学涂料中的某种化学物质有关。

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