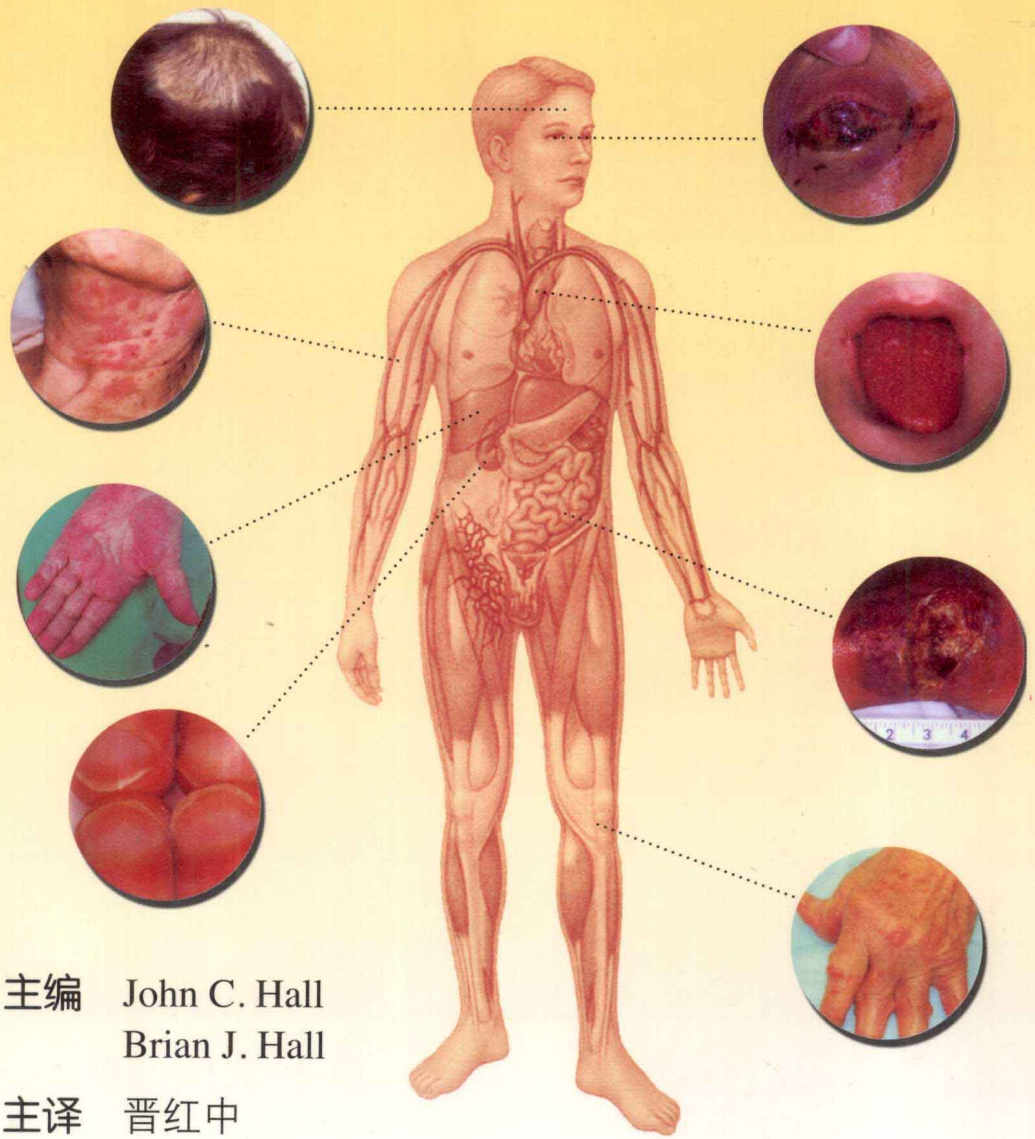
Hall 皮肤病学

——系统疾病的皮肤表现

Hall's Manual of Skin as a Marker of Underlying Disease



主编 John C. Hall
Brian J. Hall

主译 晋红中

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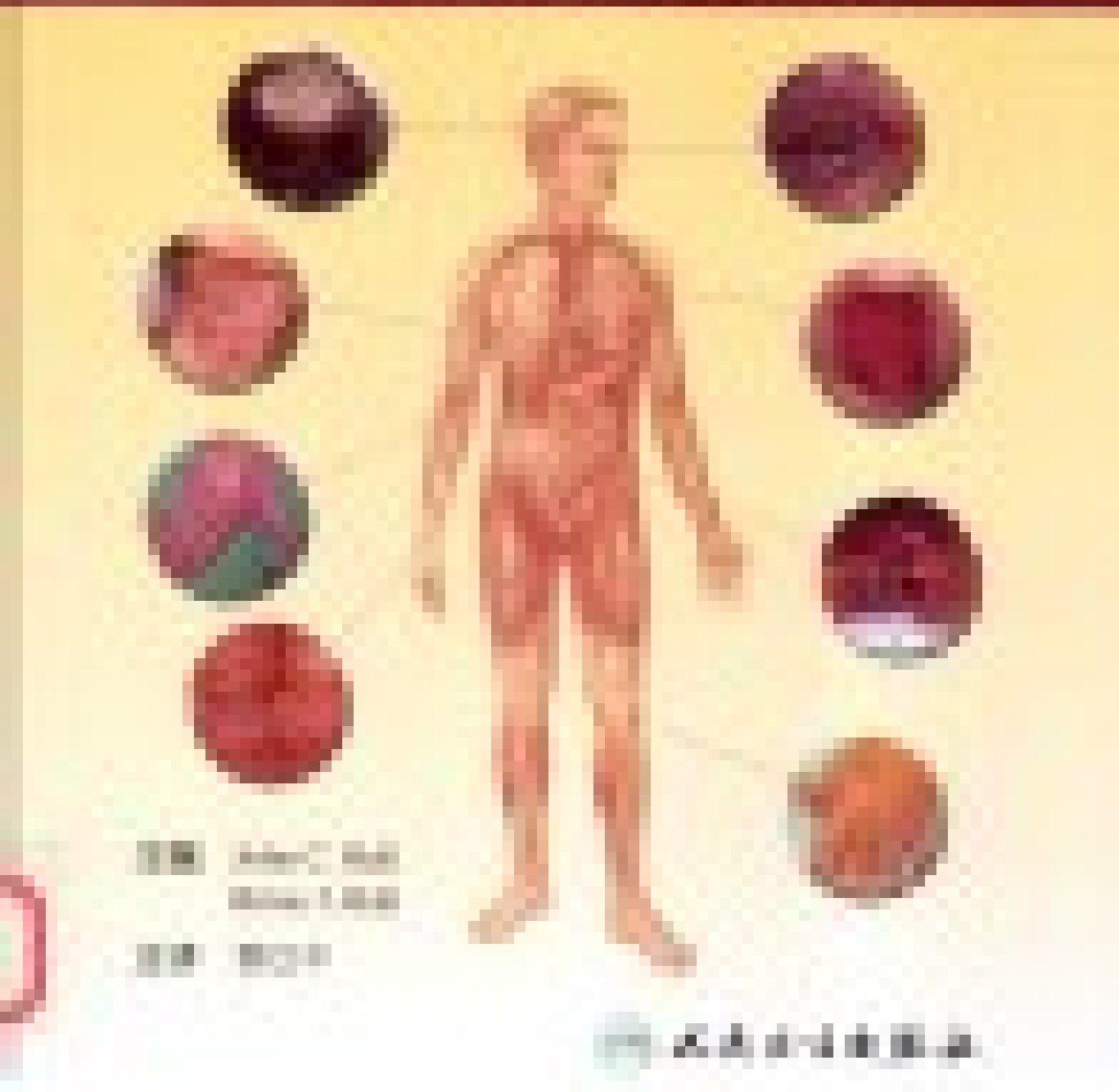
Hall

皮肤病学



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中文版序

皮肤病不仅仅是皮肤的问题，还常是许多系统疾病的表现之一。正如本书作者所言，皮肤是身体的一面镜子，皮肤是最适宜观察的器官，观察皮肤病变往往能起到事半功倍的作用。《Hall 皮肤病学——系统疾病的皮肤表现》分为器官系统、皮肤附属器、特殊疾病、感染、儿童皮肤病五个部分。在器官系统篇中，介绍了胃肠道疾病、肝脏疾病、肾脏疾病、神经系统疾病、肺部疾病、肌肉骨骼系统疾病、内分泌系统疾病、心血管疾病等主要脏器疾病的皮肤表现。在皮肤附属器篇中，介绍了色素性疾病、毛发疾病、甲病变的皮肤表现。在特殊疾病篇中，描述了白血病和淋巴瘤、糖尿病、代谢综合征、精神障碍性疾病、滥用药物、饮食障碍、虐待儿童、结节病、淀粉样变、自身免疫性结缔组织病、血管炎、遗传性疾病、移植物抗宿主病等疾病或特定课题的皮肤表现。在感染篇中，阐述了严重感染性疾病、性病的皮肤表现。最后介绍了部分儿童皮肤病的系统性表现。本书涵盖了方方面面，总结了多数疾病的皮肤表现，论述严谨，不仅适合皮肤科医师阅读，也可作为内科医师、全科医师、医学生的案头读物。

译者均来自中国医学科学院北京协和医院皮肤科工作、学习过的同仁。目前顾琳博士在清华大学第一附属医院工作；刘永鑫博士在首都医科大学附属北京世纪坛医院工作。

鉴于水平有限，一些不足甚至错误难免存在，恳请医学同道和热心读者批评指正。

北京协和医院皮肤科

晋红中

2011.6.7

原 版 序

“皮肤是身体的一面镜子。探寻疾病在皮肤的特异性表现。”

Wilbult C. Davison, MD

Duke 大学医学中心的创始人、院长

所有的医学都始于观察。皮肤是最适宜观察的器官。在随后的章节中，我们试图发挥每一位内科医生、护士、从业护士和医师助理的最大限度能力，通过评估包绕我们的皮肤组织而发现疾病。

皮肤会告诉我们很多，关于在哪里寻找疾病，疾病有多么严重，以及什么样的治疗对策会受益最多。

同时，皮肤也是最易获取组织进行病理学检查、培养和碎屑检查的器官。这些方法也将讨论并且详细阐述。

我们将皮肤情况按器官角度分类，同时也包括特殊疾病。

停下脚步，与我们一起观察、学习我们从皮肤这面镜子中得到的知识。

John C. Hall, MD

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I

器官系统

第1章

胃肠道疾病的皮肤表现

Brian J. Hall, MD

口腔/黏膜皮肤疾病

念珠菌病(candidiasis)

念珠菌(*Candida spp.*)正常存在于近半数人群中。菌群不受抑制的生长导致鹅口疮(oral thrush)或念珠菌病的发生,常见于新生儿、接受口服皮质类固醇激素或长期抗生素治疗的儿童、包括HIV感染在内的存在免疫抑制的患者、血液系统或其他恶性肿瘤的患者以及机体抵抗力减弱的患者。念珠菌病主要分为系统性和黏膜皮肤/口腔两种亚型。其他口腔念珠菌病亚型包括急性萎缩和伪膜型(见图29.7)、慢性萎缩型、慢性增殖型、慢性皮肤黏膜型、口角唇炎型、正中菱形舌炎(median rhomboid glossitis)型念珠菌病。

婴儿和新生儿的鹅口疮是由于念珠菌入驻胃肠道之前的过度生长所致,表现为红斑基础上或白色和红色质脆皮损上的白色凝乳状斑片。成年男性患者中,慢性增殖型念珠菌病最为常见,表现为边缘围绕红斑的斑块,应与外阴白斑相鉴别。临床中考虑该诊断时,应进行活检以明确病因。免疫低下患者出现黏膜皮肤念珠菌病,同样也表现为慢性增殖性病变,包括那些感染HIV的患者。

老年人多表现为慢性萎缩性病变,常由于应用假牙而出现疼痛性红色斑片。患有慢性黏膜皮肤念珠菌病的免疫低下患者,常伴发皮肤念珠菌病,累及间擦部位、面部、手部,甚至有时表现为更为广泛的播散性分布。

口腔念珠菌病是最常见的艾滋病(AIDS)相关的口腔病变,也是HIV感染的重要早期标志。患口腔念珠菌病的所有成年人都应筛查HIV。同时,念珠菌病也是许多除前文提及疾病以外的条件致病的重要标志,或与下列情况相关:维生素B_{12}缺乏或其他贫血状况,妊娠期,老年,家族性甲状腺功能低下,库欣病,桥本甲状腺炎及糖尿病。

系统性念珠菌病合并败血症时,微血栓可引起皮肤或口腔黏膜的坏死,导致局灶性的丘疹和溃疡。

不伴慢性基础疾病的患者,局部外用制霉菌素或氯三苯甲咪唑(克霉唑)最为有效。存在免疫低下的患者,需口服抗真菌药物,如氟康唑。

疱疹病毒感染

眼及外生殖器的单纯疱疹病毒(herpes simplex virus,HSV)感染将分别在第7章和第28章中详述。原发性疱疹性齿龈口腔炎(herpetic gingivostomatitis)是单纯疱疹病毒感染的重要口腔表现,常由HSV-1型感染致病,有时也见于HSV-2型。表现为唇(图1.1)、口腔、齿龈或咽部的疼痛性水疱,常伴有刺痛或痒感。90%的原发感染发生于青春期前,皮疹常伴有发热、不适、淋巴结肿大,临床上可能误诊为上呼吸道感染。皮损不需要治疗,1~2周内逐渐愈合,阿昔洛韦或泛昔洛韦能够减轻皮损的严重程度和缩短病程。组织学可见溃疡边缘的HSV感染细胞出现"3M表现",即胞核多核化、染色质边集及核固缩。

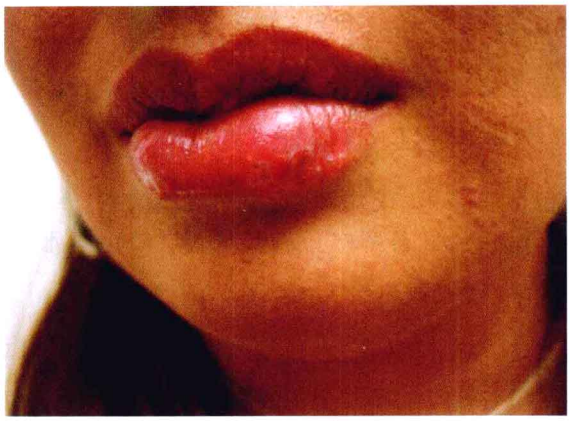

图1.1 原发性单纯疱疹病毒感染,下唇广泛分布成群的疼痛性水疱

临床上,复发性HSV感染常为淡红色,不伴有系统症状。唇疱疹的复发率因来源而异,有文献报道发生率为16%~45%。

带状疱疹(herpes zoster)

带状疱疹(水痘病毒再激活致病)的口腔病变类似阿弗他口炎(aphthous stomatitis),但常伴突然发作的疼痛,有时伴有高热,溃疡呈单侧分布。带状疱疹常是HIV感染的初期表现,若疱疹系复发、病情重或皮疹泛发,应查找潜在的免疫受抑制的疾病(艾滋病、恶性肿瘤)。口服阿昔洛韦或泛昔洛韦有助皮损愈合,剂量为500mg,每日3次,连用7天。合并严重并发症时,每次予阿昔洛韦500mg,每8小时一次,静点10天。有报道证实给予阿昔洛韦1g,每日3次,口服7天能够缩短愈合时间,减少新发皮损。

扁平苔藓(lichen planus)

扁平苔藓是一种非常常见的皮肤和口腔黏膜的慢性炎症性疾病,常伴有剧烈瘙痒。临床中,常用5P来进行描述,即对称、扁平、紫色(紫罗兰色)、瘙痒的多角形丘疹(见图14.8和图14.9)。发病率约为1%,常成人发病(儿童较少见),女性多发。

口腔病变表现为黏膜上白色花边状皮损(图1.2)。约15%~35%的患者仅有口腔病变,约20%的口腔扁平苔藓患者同时存在皮肤病变。多达15%的患者与丙型肝炎相关。临床中应非常重视出现萎缩或溃疡的慢性扁平苔藓,尽管发生率很低,仍有约0.2%~10%的

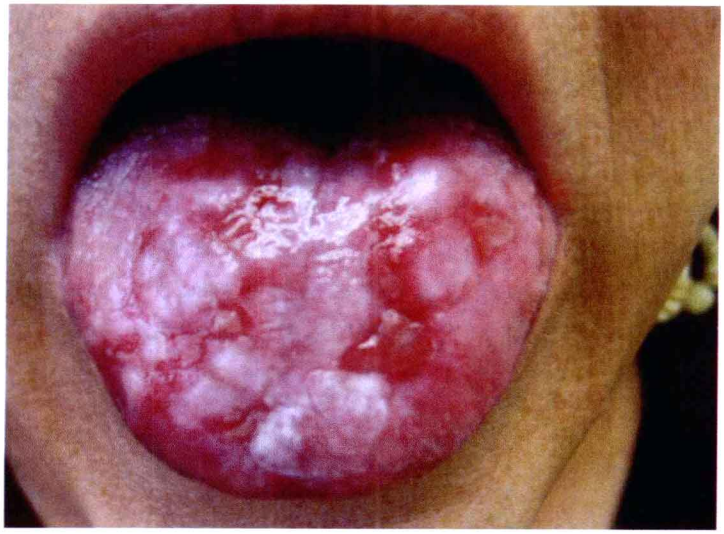

图 1.2 舌部扁平苔藓,红色基底上见白色网状斑块

患者与恶性肿瘤发病率增加相关。治疗包括局部和(或)系统应用糖皮质激素。组织学上,扁平苔藓呈典型的苔藓样皮炎表现,真皮乳头线状(苔藓样)浸润,基底层液化坏死,表皮突呈锯齿状,颗粒层增厚,角化过度。

大疱性疾病(blistering diseases)

大疱性类天疱疮(bullous pemphigoid,BP)

大疱性类天疱疮是一种自身免疫性大疱性疾病,为突出于正常皮肤的紧张性大疱。文献报道 8% ~58% 的患者存在黏膜病变(图 1.3)。近期一篇文献发现,115 例大疱性类天疱疮中 24% 存在口腔受累。偶尔,疾病的发生与潜在恶性肿瘤相关,但这可能仅仅与后者在老年群体中的高发病率相关。

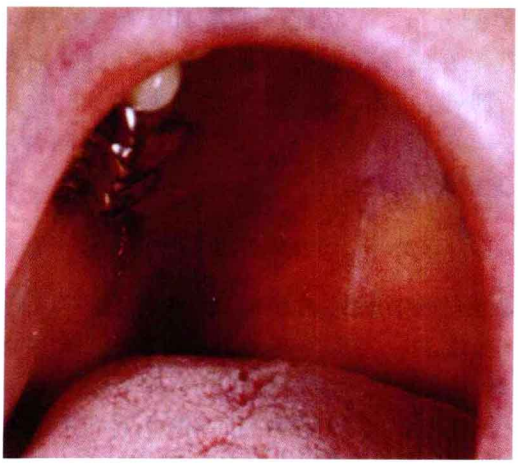

图 1.3 大疱性类天疱疮患者上腭处见小水疱

瘢痕性类天疱疮(cicatricial pemphigoid)

瘢痕性类天疱疮是一种罕见的自身免疫性瘢痕性疾病,受累黏膜(特别是眼和口腔)表面出现糜烂、溃疡和水疱(图 1.4),大约 1/3 的患者同时存在皮肤损害。约 64% 的患者发生眼部损害,这些患者常首先就诊于眼科医生。85% ~95% 的患者发生口腔损害。直接免疫荧光与大疱性类天疱疮一致,80% 病例的基底膜带透明板可见 IgG 和(或)C3 线状沉积,口腔黏膜的检出率高于皮肤。应用系统性皮质类固醇和其他免疫抑制剂进行治疗。

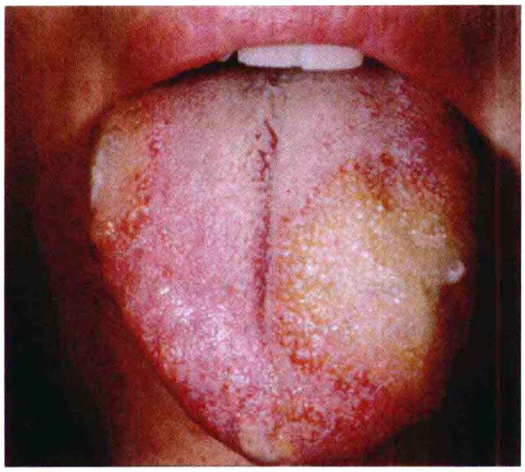

图 1.4　瘢痕性类天疱疮患者的舌上红斑边缘大疱形成

寻常型天疱疮(pemphigus vulgaris)

寻常型天疱疮是一种罕见的水疱大疱性疾病,好发于中年,正常皮肤或红斑上出现松弛性易破裂的水疱(相对于大疱性类天疱疮)。几乎所有患者都存在口腔损害,病变常始于口腔(约 50% ~70% 的病例),经数周至数月后扩展至皮肤。大约 63% ~87% 的患者存在食管受累。

手指摩擦外观正常的未受累的皮肤,导致水疱形成(称为尼氏征阳性)。大疱性类天疱疮与之相反,尼氏征为阴性。皮肤和口腔黏膜上的水疱均易迅速破裂,形成疼痛、新鲜的出血性糜烂面(图 1.5)。

由于水疱常破裂,活检早期皮损对确立正确的诊断十分重要。桥粒核心糖蛋白Ⅲ抗体导致基底层上棘刺松解水疱形成,形成特异性的完整的单层基底细胞,呈墓碑状排列,类似胃肠道绒毛。治疗给予泼尼松(有时为剂量非常大),常为 10mg,每日 4 次;皮损愈合后,渐减量或改用霉酚酸酯。也可应用其他免疫抑制剂。

大疱性表皮松解症(epidermolysis bullosa)

大疱性表皮松解症(EB)是一组罕见的皮肤脆性遗传性疾病,除获得性大疱性表皮松解症外,多发生于婴儿期或出生时。皮肤和黏膜受到轻微创伤即可出现水疱。愈后留有瘢痕,慢性皮损有可能进展为鳞状细胞癌。

四种主要亚型为单纯型、半桥粒型、交界型和营养不良型。营养不良型常表现为口腔糜烂、龋齿、齿龈和胃肠道受累、肛门狭窄、便秘以及食管狭窄,其中口腔糜烂和食管狭窄是最常见的胃肠道并发症。交界型也可出现类似的胃肠道并发症。重症营养不良型和交界型大疱性表皮松解症患者,可由于这些胃肠道并发症继发贫血和生长发育迟缓。幽门闭锁与交界型大疱性表皮松解症相关。

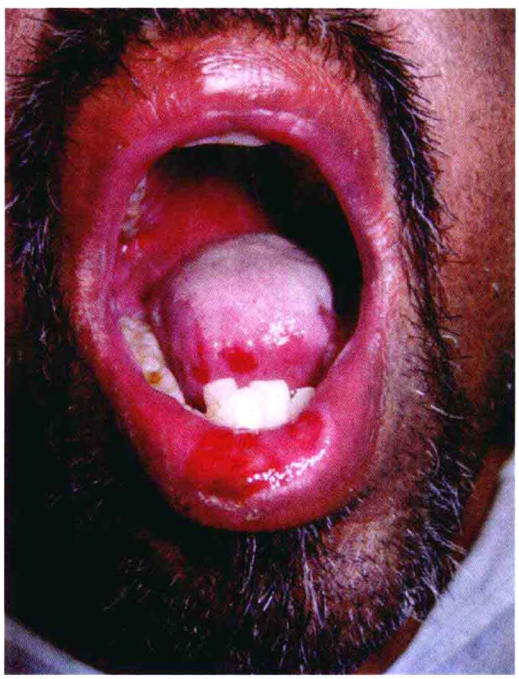

图 1.5　寻常型天疱疮患者下唇上新鲜的出血性糜烂面

获得性大疱性表皮松解症(epidermolysis bullosa acquisita,EBA)

获得性大疱性表皮松解症患者皮损与营养不良型大疱性表皮松解症相同,但不