



图1 硬性耳内镜检查 左外耳道口见淡红色新生物; 图2 颞骨CT所示 左侧外耳廓及外耳道软组织影增厚;
图3 术后病理所示 黑色箭头示基底细胞癌,红色箭头示鳞状细胞癌(苏木精-伊红染色 ×100)。

总之,对于可疑基底鳞状细胞癌患者,术前需完善相关术前检查,了解有无转移、扩散现象,尽量做到早诊断、早治疗,术后定期随访,必要时术后辅以化疗,以延长患者生命,改善生存质量。

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以声嘶为首发症状的 Ramsey-Hunt 综合征 1 例

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[关键词] Ramsey-Hunt 综合征;声嘶;误诊
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Ramsey-Hunt syndrome with initial syndrome of hoarseness: a case report

Summary Ramsey-Hunt syndrome is caused by the varicella zoster virus, which mainly affects the facial nerve. The typical clinical features of Ramsey-Hunt syndrome are peripheral facial paralysis and ear herpes. In this case, initial symptoms were hoarseness and coughing, afterwards typical symptom occurred 5 days later which were earache, late-onset herpes, and facial paralysis. Ramsey-Hunt syndrome are difficult to diagnose because of its seemingly unrelated initial symptoms.

Key words Ramsey-Hunt syndrome; hoarseness; diagnostic errors

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1 病例报告

患者,男,65岁,因声嘶伴饮水呛咳3d于2016年9月7日入院。患者主诉3d前无明显诱因出现声嘶,伴饮水呛咳、咽痛、咽部异物感,无咳嗽、咳痰,无吞咽困难、呼吸困难,无发热、全身酸痛,于当地医院给予抗炎药物及激素药物静脉滴注(具体药物不详),症状未见好转。既往高血压、糖尿病6年,自测血压最高达150/97 mmHg(1 mmHg=0.133 kPa),血糖未规律监测,均未规律服用药物治疗。近期无全麻手术史。入院查体:体温36.7℃,脉搏89次/min,呼吸12次/min,血压139/80 mmHg,快速血糖11.5 mmol/L,双侧扁桃体Ⅱ度大,软腭低垂,口腔及咽腔未见溃疡,间接喉镜无法配合。双侧鼻腔黏膜正常,双下甲肥大,未见异常分泌物。双耳廓正常,外耳道清洁干燥,双耳鼓膜完整,骨性标志清。颈部未触及肿大淋巴结。其他各对脑神经功能正常。多功能纤维喉镜示:右侧声带松弛,呈旁正中位,左侧声带活动正常,声门闭合不良(图1)。实验室检查:白细胞 $9.89 \times 10^9/L$,C反应蛋白2 mg/L,中性粒细胞0.686,淋巴细胞0.243,糖12.41 mmol/L,糖化血红蛋白8.1 mmol/L,尿葡萄糖卅。



右侧声带松弛,呈旁正中位,双侧声带表面黏膜无充血、水肿,未见明显新生物,双侧室带黏膜正常。

图1 患者入院后喉镜检查

入院后诊断为“声带麻痹(右)”,给予营养神经药物及激素治疗(甲钴胺10 mg每天1次静脉滴注,甲泼尼龙第1天80 mg,1 d后改为40 mg静脉滴注,每天1次),并给予雾化、化痰辅助治疗。用药后咽痛较前缓解,声嘶、饮水呛咳症状如前。入院2 d后患者自诉右耳痛,查体见右耳廓肿胀,表面泛红,皮温略高,耳甲腔皮肤可见少数散在糜烂面,伴少量渗出,耳痛明显。右侧鼻唇沟较左侧浅,面部静态面容基本对称,示齿时右侧嘴角歪斜,鼓气时未见嘴角漏气,面神经功能分级Ⅱ级。诊断为Ramsey-Hunt综合征。加用抗病毒治疗(阿昔洛韦片0.2g口服,每天5次,持续7 d),入院5 d后出

院,嘱患者继续口服阿昔洛韦片及甲钴胺片。出院2周后随访,患者耳甲腔疱疹已痊愈,声嘶及饮水呛咳症状恢复。

2 讨论

Ramsey-Hunt综合征是由水痘带状疱疹病毒引起,以侵犯面神经为主的疾病^[1],临床典型特征为周围性面瘫伴同侧耳部疱疹,但临床中表现不典型,极易误诊。近年来,因迟发疱疹、异位疱疹、无疱疹而导致误诊的Ramsey-Hunt综合征病例层出不穷。刘冬梅等(1994)报道了以眩晕、剧烈呕吐为首发症状就诊的病例;肖淑芬等^[2]报道1例误诊为下咽癌的不典型Ramsey-Hunt综合征;姜岚等^[3]报道1例患者,初发单侧咽喉部簇状溃疡,3 d后出现同侧耳痛、疱疹、面瘫三联征,合并同侧耳鸣及听力下降。

Ramsey-Hunt综合征主要累及面神经,以面神经运动支受损率最高,约占93.8%^[4]。耳廓和外耳道的感觉神经除面神经分支外,还有迷走神经及舌咽神经,因此水痘带状疱疹病毒除侵犯面神经外,还可侵犯迷走及舌咽神经,故疱疹可出现于上述神经的分布区,如:口腔黏膜、软腭、扁桃体、舌根、喉部及颈部等。另外,带状疱疹病毒还可侵犯到前庭神经、耳蜗神经及三叉神经,表现为眩晕、耳聋及同侧剧痛。

Malin等(1985)对Ramsey-Hunt综合征的定义为:①耳疱疹和(或)同侧软腭溃疡^[5];②同侧面瘫,伴有味觉障碍和泪液分泌障碍或减少;③三叉神经(第一和第三支)分布区感觉障碍^[6];④颈部皮肤感觉障碍(C2~C4分布区);⑤听神经受累;⑥前庭神经受累。还有可能少见的感染范围是:视神经、展神经、舌咽神经、迷走神经、交感神经、脊神经、颅内(脑膜脑炎、脑血管炎、脑血管意外、偏瘫、半身感觉障碍)等^[3]。

本例最初表现为声嘶及饮水呛咳,首先考虑咽喉炎,行喉镜检查示右侧声带旁正中位,需完善相关检查明确声带麻痹原因:①明确全身麻醉手术史,有无全麻术中插管可能导致环杓关节脱位,表现为一侧声带固定;②排除甲状腺、颈淋巴结、肺尖等处的机械性压迫或牵拉;③排除肺部、纵膈等处肿瘤及转移灶可能,特别是肺尖部。排除上述可能后给予营养神经药物治疗无效,应高度怀疑为喉返神经受病毒侵犯所致。患者发病5 d后出现耳痛、耳甲腔典型皮疹、面瘫三联征,考虑为带状疱疹病毒首先侵及迷走神经喉上神经感觉支和喉返神经运动支,继而侵及面神经。由于发病部位特殊,首发症状不典型,属于临床少见病例,容易误诊,给早期治疗带来难度。

回顾文献发现Ramsey-Hunt综合征发病3 d

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后用药痊愈率为 75%，而 7 d 后用药则降为 30%，激素联合阿昔洛韦治疗效果比单独使用激素更好^[7]。除激素与抗病毒药物外，还可辅以外周血管扩张及改善面部血液循环药物。针对面神经功能损伤患者，也可辅以针灸理疗，促进面神经恢复功能，70.4% 的患者面神经功能可完全恢复。部分 Ramsey-Hunt 综合征患者有听力下降症状，平均言语频率提高明显者预示面神经恢复较好^[8]。患者合并全身代谢性疾病，或合并耳蜗和(或)前庭损害或侵及面神经以外脑神经时，预后较差^[9]。

综上所述，临床医生应加强 Ramsey-Hunt 综合征的认识，对于迟发疱疹、异位疱疹，甚至无疱疹的 Ramsey-Hunt 综合征患者，应掌握诊断要点及治疗时机，最大可能地保护受累神经，减少后遗症，改善预后。

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