

扁桃体滤泡树突细胞肉瘤 1 例

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摘要: **目的** 探讨扁桃体滤泡树突状肉瘤(FDCS)的临床表现、体征、诊断、病理特征、治疗方法,提高对该病的认识,减少误诊率。**方法** 回顾性分析 1 例扁桃体 FDCS 疾病特征,该患者因右侧口咽肿胀入院,无明显临床症状。入院后行颌面部及颈部 CT 检查,结果示右侧口咽部占位性病变,病理及免疫组化检查结果为 FDCS,遂对该患者行右侧扁桃体及肿物扩大切除术。**结果** 完全切除右侧扁桃体及肿物,术后无出血、感染等并发症。术后予以放疗,随访 3 个月,未见复发。**结论** FDCS 是一种罕见的恶性肿瘤,临床症状表现无特异性,在临床工作中易被误诊,病理学及免疫组化检查是诊断该疾病的主要方法,治疗以完全性手术切除为主,术后可辅以放疗和(或)化疗。

关键词:扁桃体;恶性肿瘤;滤泡树突细胞肉瘤
中图分类号:R739.64

Follicular dendritic cell sarcoma of tonsil: a case report

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Abstract: **Objective** To investigate the clinical manifestations, signs, diagnosis, pathological features and treatment of tonsil follicular dendritic sarcoma, so as to improve the understanding of the disease and reduce the rate of misdiagnosis. **Methods** The characteristics of tonsillar follicular dendritic cell sarcoma were retrospectively analyzed. The patient was admitted to hospital on right oropharyngeal swelling without obvious clinical symptoms. After admission, CT examination of the maxillofacial region and neck was performed. The results of CT examination showed that the right oropharynx was occupied. The results of pathological and immunohistochemical examination were follicular dendrite cell sarcoma; The patient was treated by right tonsillectomy and enlarged resection. **Results** The right tonsil and tumor were completely excised, and had not postoperative complications such as bleeding and infection. The patient was given radiotherapy after the operation, and no recurrence after 3 months of follow-up. **Conclusion** Follicular dendritic cell sarcoma is a rare malignant tumor without specific clinical symptoms, which is easy to be misdiagnosed in clinical work. Pathology and immunohistochemical examination are the main methods for the diagnosis of this disease. The treatment is mainly by surgical resection, and radiotherapy and/or chemotherapy is often used after surgery.

Keywords:Tonsil; Malignant tumor; Follicular dendritic cell sarcoma

滤泡树突细胞肉瘤(follicular dendritic cell sarcoma, FDCS):是一种罕见的来自滤泡树突细胞的恶性肿瘤,是树突细胞肉瘤中最常见的亚型。1986 年 Monda 等^[1]首先报道了起源于滤泡树突状细胞的细胞肉瘤。现报道我院收治的 1 例原发于扁桃体 FDCS 的诊治经过,提高对该病的认识,减少误诊率。

1 资料与方法

1.1 临床资料

患者男,50 岁,因“右侧口咽肿胀半月余”于 2019 年 9 月入院,主诉无咽部异物感、吞咽不畅、呼吸不畅、咳嗽、咳痰及发热等不适,查体可见口咽部

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黏膜充血,右侧腭舌弓向中线推进,未及悬雍垂,表面光滑,扁桃体未窥及,怀疑咽旁肿物。颈部未触及明显肿大淋巴结。病理检查结果示:(咽侧壁)黏膜见大量淋巴组织滤泡样增生,表面鳞状上皮轻中度异型,患者出院等待免疫组化结果,免疫组化结果示:CK(-),CD31(-),CD34(-),Ki67(+10%),SMA(-),ERG(-),F8(-),Vim(+),S100(-),HMB45(-),Desmin(-),CgA(-),Syn(-),CD21(+),CD1a(-),EMA(-),EBER(-),ALK(-),CD30(-),CK(上皮+),Vim(+),LCA(-),P63(上皮+),P40(上皮+),Ki67(+3%),CD68(-)。确诊为(咽侧壁)FDCS。患者再次入院,出现咽部异物感,查体可见右侧软腭及前弓中线向前、向上、向外隆起,向上及悬雍垂根部水平,肿胀较前明显,右侧扁桃体未窥及。

1.2 影像学资料

颌面部 CT 平扫 + 多期增强,颈部软组织平扫 + 多期增强结果示:右侧口咽壁呈肿块增厚,局部突入口咽腔及咽旁隙,大小约 2.8 cm × 2.5 cm,CT 值约为 65 Hu;增强扫描呈中度强化,动静脉期 CT 值约为 71、80 Hu,边缘光整,气管居中,甲状腺体积略大,其内密度不均匀,双侧颈部肌肉组织无异常,双侧颈部 II 区可见稍大淋巴结影,大者位于右侧 II 区,大小约为 1.0 cm × 0.9 cm。双侧颌面部对称,软组织影未见增厚、大小形态密度正常,颌面部脂肪间隙清晰,组成颌面部骨质形态、密度正常,未见明显异常。典型图片见图 1,考虑右侧口咽部占位性病变。胸部正侧位片结果示:左侧第 3 肋间可疑结节影,与既往对比,两肺纹理重。行胸部 CT 结果示:右肺中叶内侧段及左肺上叶舌段陈旧条索影。颈部及腹部多普勒超声未见明显异常。

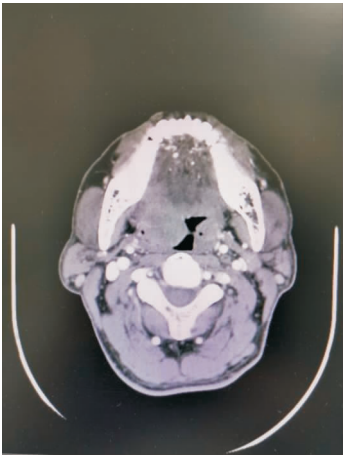


图 1 颌面部 CT 平扫图片

1.3 治疗

完善相关检查后在全麻下行右侧扁桃体及肿物扩大切除术,术中右侧扁桃体表面可见凹凸不平肿物,切开腭舌弓及腭咽弓上方约 1.0 cm 处的黏膜,找到肿物上极,切除扁桃体及肿物和软腭黏膜至悬雍垂根部。肿物与咽旁组织粘连较紧密,界限不清。查无病变残留后,缝合前后弓,术毕。

2 结果

手术过程顺利,完全切除右侧扁桃体及肿物,术后未见出血、感染等并发症。术后标本病理结果符合 FDCS,病理检查见图 2。患者术后 7 d 出院,继续于肿瘤医院就诊,出院后随访 3 个月,局部未见复发。

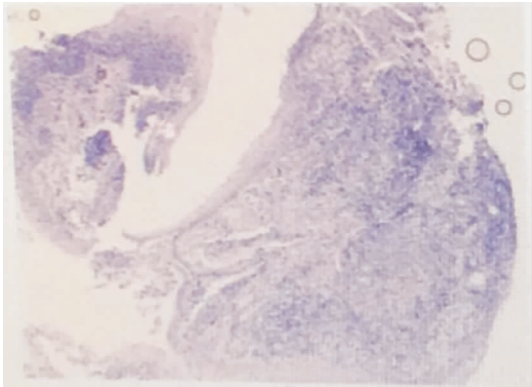


图 2 右侧扁桃体 FDCS 病理图 (HE × 40)

3 讨论

3.1 疾病特点

FDCS 为起源于滤泡树突状细胞的低度恶性肿瘤,可发生于淋巴结内或淋巴结外。该病发病年龄为 17 ~ 76 岁,平均 43.8 岁。男女发病率差异无统计学意义,炎性假瘤样型 FDCS 好发于女性^[2]。Gounder 等^[3]在 47 例 FDCS 的病例研究中发现,其他恶性肿瘤的发病率较高,大多数实体肿瘤发展早于 FDCS,该现象可能与免疫抑制有关。发病机制尚不明确,可能与自身免疫、EB 病毒感染等因素有关,Cakir 等^[4]认为 FDCS 由血管滤泡性淋巴组织增生而来,与透明血管型 Castleman 病有关。肿瘤好发部位为颈部、纵隔、腋窝淋巴结,也可发生于结外部位,如扁桃体、咽喉部、软组织和结直肠等,炎性假瘤样型 FDCS 则多发于肝、脾等腹腔脏器^[5],咽部

FDCS 可发生于扁桃体、鼻咽、口咽、咽旁间隙等部位。肿瘤常呈无痛性生长,部分可表现为压迫症状,咽部可出现鼻塞、呼吸不畅、吞咽异物感等。FDCS 恶性程度较低,但也存在复发和转移可能。FDCS 影像学表现无特异性,主要通过病理学检查及免疫组织化学特征确诊,特征性免疫标记为 CD21、CD35、CD23 和 KI-M4P,还可见 CD68、CD18、CD11a、Vimentin、S-100 蛋白、EAM 及 SMA 的弱阳性表达^[6]。该病发病率低,发病部位多样,临床表现无特异性,需与同属于树突细胞的肉瘤相鉴别,另外还需要与淋巴瘤、肉瘤、异位胸腺瘤、脑膜瘤、恶性纤维组织细胞瘤、淋巴上皮样癌、朗格汉斯细胞肉瘤等鉴别。

3.2 治疗及预后

局部 FDCS 且无远处转移治疗以局部完整切除为主,术后可予以辅助性放疗或化疗,术后予以辅助性放疗或化疗与单纯手术切除病灶预后无明显差异;对无法手术根治和术后复发的病例可予以放疗或(和)化疗,常用的化疗方案包括吉西他滨联合紫杉醇等^[7]。Jain 等^[7]在对 66 例 FDCS 研究中报道对于 FDCS 的靶向治疗,在 2 例患者中发现 *TP53*、*TPEN* 基因突变,将进一步确定基因概况和分子的靶向治疗。影响预后的因素包括:肿瘤体积大(>6 cm),肿瘤发生于腹腔内,未行术后辅助治疗,肿瘤有凝固性坏死,肿瘤细胞的细胞核具有异型性及细胞核分裂指数高(>5/HPF)^[8]。

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