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◇ 临床医学 ◇

以谵语为临床表现的儿童烟雾病合并大面积脑梗死 1 例及文献复习

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摘要: **目的** 通过报道 1 例以谵语为临床表现的儿童烟雾病合并大面积脑梗死病例的诊断过程, 提高儿科医生对儿童烟雾病的重视和对儿童烟雾病临床表现的认识程度。 **方法** 回顾性分析 1 例以谵语为临床表现的儿童烟雾病合并大面积脑梗死病例的临床资料与诊疗过程。 **结果** 患儿 12 岁, 男性, 以谵语为主要临床表现。头颅 CT 示左侧大脑半球大面积脑梗死; 颅脑 MRI+MRA 诊断: (1) 烟雾病, 并右侧一向前上纵形畸形血管; (2) 左侧颞顶枕叶大片异常信号灶, 考虑急性缺血、缺氧并梗死改变, 感染性病变不排除。确立诊断为烟雾病, 脑梗死。后转至神经外科行联合血管重建术治疗, 经随访患儿恢复状况良好。 **结论** 儿童烟雾病临床表现以肢体无力、感觉障碍等为常见症状, 以谵语为临床表现较为少见。临床工作中应提高儿科医师对儿童烟雾病的重视程度, 对不明原因谵语、精神障碍、发热、头痛、癫痫发作等表现者, 在排除其他病因后, 应早做影像学检查并行及时有效处理, 尽快的确诊患儿。

关键词: 烟雾病; 脑血管造影术; 脑血管重建术; 谵语; 儿童

Clinical manifestations of delirium in children with moyamoya disease with massive cerebral infarction: a case report and literature review

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Abstract: Objective Reported by 1 case with delirious speech as the clinical manifestations of the diagnosis of moyamoya disease in children with large area cerebral infarction cases process was reported, by which to improve the pediatrician's emphasis on moyamoya disease in children and the awareness of clinical manifestations of children moyamoya disease. **Methods** The clinical data and the diagnosis and treatment of a case of childhood moyamoya disease with large area cerebral infarction were retrospectively analyzed. **Results** The children were 12 years old, male, with delirious speech as the main clinical manifestation. Skull CT showed large cerebral infarction in the left hemisphere. Brain MRI+MRA diagnosis: 1. Moyamoya disease, and on the right side of a longitudinal deformed blood vessel. 2. Large abnormal signal foci of the left temporal occipital lobe, considering acute ischemia, hypoxia and infarction changes, with no exception of infectious lesions. Establish a diagnosis of moyamoya disease, cerebral infarction. The patient was transferred to neurosurgery and treated with combined vascular reconstruction. The patient recovered well after follow-up.

Conclusion The clinical manifestation of childhood moyamoya disease is characterized by weakness of limbs and sensory disturbance. And delirious speech is rare. Pediatricians should be increased in clinical work emphasis of moyamoya disease in children, for unknown reasons delirious speech, mental disorders, fever, headache, seizures and other performers, after ruling out other causes, should do early imaging examination parallel to the timely and effective treatment, the diagnosis of children as soon as possible.

Key words: Moyamoya disease; Cerebral angiography; Cerebral revascularization; Delirious speech; Children

烟雾病(moyamoya disease, MMD)为目前病因未能确定,临床上多以肢体无力、感觉障碍为主要发病表现的一种较为罕见脑底异常血管网疾病。该病存在10~14岁与40岁左右两个发病高峰,临床上以谵语为唯一临床表现的儿童烟雾病较为少见。现对我科确诊的1例以谵语为唯一临床表现的儿童烟雾病的诊断过程进行临床资料回顾,进而分析儿科烟雾病常见临床表现与诊疗过程。经过沟通,患儿父母对本次报道研究已做相应知情同意,本研究符合《世界医学协会赫尔辛基宣言》相关要求。

1 临床资料

男,12岁。因“胡言乱语4 d”于2018年2月9日入蚌埠医学院第二附属医院儿科。患儿入院前5 d无诱因下出现一次头痛,伴有发热,最高体温38℃,无呕吐,于当地诊所给予对症止痛治疗,具体用药不详,用药后体温恢复正常,头痛好转。患儿家属因期末考试成绩较差于入院前4 d有打骂患儿行为,随后患儿出现谵语,唯其母亲陪伴时,谵语情况较为减轻。以“中枢神经系统感染:病毒性脑炎可能,瘵症待排”收治入院。病程中患儿胃纳欠佳,无腹泻、抽搐,睡眠尚安静,大小便正常。既往体健,患儿4~5岁时有车外伤史,无颅内手术病史,否认类似发作史。否认脑血管家族病史,家族成员均未有类似临床表现。体检及实验室检查:体温36.5℃,脉搏90次/分,呼吸25次/分,血压116/86 mmHg,体质量45 kg,神志恍惚,谵语,营养发育良好,头颅无畸形,右侧颞部有一瘢痕,双瞳孔等大等圆,瞳孔对光反射灵敏,脊柱四肢无畸形,四肢肌力V级、肌张力0级,布鲁金斯基征阴性,克尼格氏征阴性,双侧巴彬斯基征阴性。心电图及实验室辅助检查均未见异常。影像学资料:头颅CT示左侧大脑半球大面积脑梗死(图1);颅脑磁共振(MRI)诊断:(1)烟雾病,并右侧一向前上纵形畸形血管;(2)左侧颞顶枕叶大片异常信号灶,考虑急性缺血、缺氧并梗死改变,感染性病变不排除(图2~4)。诊断:烟雾病,脑梗死。考虑患儿脑梗死病情严重,遂转至经外科行联合血管重建术治疗,后经随访患儿恢复状况良好。

2 讨论

烟雾病于1957年首先由日本学者进行报道,并

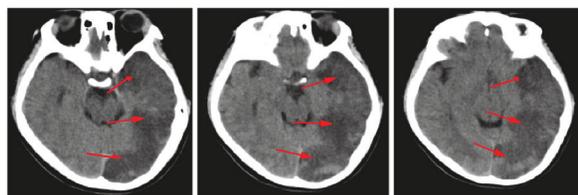


图1 烟雾病患儿头颅CT示左侧大脑半球见大片状低密度影,密度不均,边界尚清,余脑实质内未见明显异常密度影

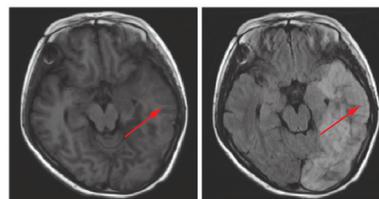


图2 烟雾病患儿颅脑MRI平扫示左侧颞顶枕叶见大片状长T1长T2信号,T2 flair呈高信号

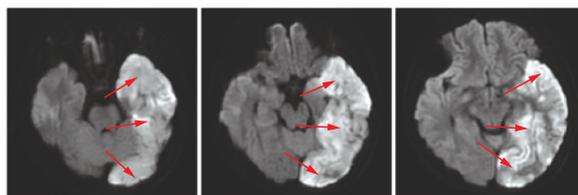


图3 烟雾病患儿病灶DWI呈高信号表现

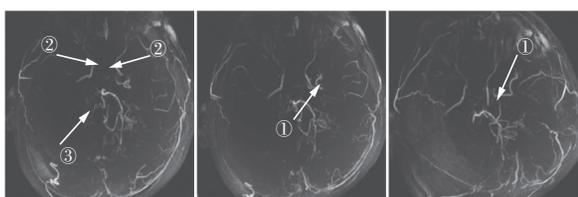


图4 颅脑MRA示双侧颈内动脉末端变窄或闭塞,周围见多发杂乱扭曲血管影,右侧见一向前上增粗纵形延伸血管影(箭头1),两侧大脑前及中动脉闭塞(箭头2),左侧小脑后动脉纤细(箭头3)

于1969年正式命名为“moyamoya”(意为烟雾),国内于1976年首次报道该病例^[1]。该病发病机制尚不明确,据目前研究显示,遗传因素在其病因中可能起重要作用^[2]。Kim和Park等^[3-4]认为RNF213基因与烟雾病的发生显著相关,有学者还认为第3、6、8、17号染色体^[5]可能在烟雾病病人中存在特定遗传。此外,在获得性环境因素中,感染因素有着很大影响。我国研究报道部分病例与钩端螺旋体感染有关。国内目前并没有针对烟雾病的统一诊断标准,目前尚以日本烟雾病诊断治疗指南为标准^[6],该病影像学诊断的金标准为数字减影血管造影(DSA)。目前,DSA存在着检查费用较为昂贵和有

创性等缺点。随着CT血管造影(CTA)、磁共振血管成像(MRA)等技术的快速发展,使得CTA、MRA逐渐于临床应用中代替DSA成为烟雾病的主要影像学诊断方式^[7]。

烟雾病多见于儿童及青壮年,存在着10~14岁和40岁左右两个发病年龄高峰。常见的临床表现有:短暂性脑缺血发作(TIA)、脑卒中、头痛、癫痫发作和智能减退等。相比于成年人,儿童烟雾病以梗死型和短暂性脑缺血发作(TIA)型较常见,占儿童烟雾病70%~80%。由于患儿对于TIA症状不易准确清晰描述,故常引发漏诊、误诊,故实际中梗死型较TIA型更多见^[8]。儿童病人的烟雾病临床表现以肢体无力、感觉障碍为常见症状,也可有头痛、癫痫发作、不自主运动、精神症状、智力倒退等^[9-10]。此患儿在校成绩较差,即可能与患有此病具有一定相关性。本例患儿主要临床表现为谵语,重复话语“我要回家”。结合病理生理学依据,该患儿即可能为左侧颞顶枕叶受累较重,病前家属打骂患儿行为刺激加重了已经受损的血管功能失调,致左侧颞顶枕叶缺血加重,从而出现谵语等精神障碍表现,此临床表现在烟雾病以往报道中尚属罕见。虽然烟雾病于儿科疾病中较为少见,但其已成为儿童脑卒中常见的危险因素之一。

目前针对烟雾病的治疗方法以应用血小板抑制剂、抗凝剂、钙离子通道阻滞剂、糖皮质激素等为主的保守治疗和以血管重建为主要方式的外科治疗。其中外科治疗方式包括直接血管重建、间接血管重建及联合血管重建。直接血管重建术于低龄患儿中受到一定的制约,多适用于年龄较大的儿童,且在患儿缺血症状治疗及改善预后方面存在一定的局限性^[11]。间接血管重建术以脑-硬膜-颞浅动脉血管连通术(EDAS)、脑-硬膜-颞浅动脉-颞肌血管连通术(EDMAS)较为常用。联合血管重建术是目前较提倡的儿童烟雾病治疗方案之一^[12]。同时,合理有效的心理护理和康复指导在预防并发症和改善预后方面亦有十分重要影响^[13]。

由于儿童的语言表达能力限制了对自己病情感受的详细阐述,导致儿科医生病史采集困难。部分儿童烟雾病临床表现不明显,也是烟雾病在儿童病人容易被漏诊或误诊的重要原因之一。同时,由于烟雾病并未得到儿科医生的重视以及医生自身经验水平不足,部分患儿则易被误诊为脑炎、急性脑梗死等。

通过本病例的诊治,提示我们临床工作中应提高儿科医师对儿童烟雾病的重视程度,对不明原因

谵语、精神障碍、发热、头痛、癫痫发作等表现者,在排除其他病因后,应早做影像学检查并行及时有效处理,尽快的确诊患儿,减少误诊漏诊可能,争取救治时间,避免耽误病情为后续治疗带来困难。此外,家长对患儿疾病重视程度是争取救治时间的关键因素之一。本例患儿由于成绩原因遭受打骂,其家长在患儿出现谵语时未予以重视,以致入院时病情较重。家庭成员应考虑到儿童语言表达的特殊性,对儿童反常行为应及早关注并采取措施,以免延误病情错过治疗最佳时机。对于儿童烟雾病的预后,早发现、早诊断、早治疗是关键,影像学技术的发展和神经外科治疗手段的不断提升也至关重要。

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