朗格罕细胞组织细胞增生症颅内垂体影像表现及相关临床表现

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摘要 目的:探讨朗格罕细胞组织细胞增生症累及垂体的MR表现及相关临床表现。方法:搜集了6例确诊为朗格罕细胞组织细胞增生症并垂体表现异常的患儿,男5例,女1例,年龄2～11岁,平均(6± 3)岁,对其影像及临床表现进行回顾性分析。结果:临床患儿主要以头面部包块,多饮、多尿等就诊。头颅MR平扫(T1W)表现6例患儿神经垂体高信号全部消失,垂体柄增粗5例,垂体柄著征1例,垂体饱满1例,其中3例治疗后复查垂体及垂体柄均有变化。结论:神经垂体高信号消失,垂体柄增粗或著征为朗格罕细胞组织细胞增生症累及垂体的头颅MR表现。累及垂体者临床几乎都有多饮、多尿表现。目前,MRI是诊断朗格罕细胞组织细胞增生症累及垂体的唯一可靠的影像学检查方法,并对治疗后病情随访有重要作用。

关键词:朗格罕细胞组织细胞增生症;神经垂体;垂体柄;磁共振成像

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The Pituitary Imaging Findings and Relative Clinical Performance for Langerhans Cell Histiocytosis

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ABSTRACT Objective: To study the Langerhans cell histiocytosis invading the pituitary MR findings and associated clinical performance. Methods: We collected six patients who were diagnosed as Langerhans cell histiocytosis and pituitary expression abnormalities in children, 5 males, 1 female, aged from 2 to 11 years, average age (6 ± 3). Their images findings and clinical performance were taken as a retrospective analysis. Results: The main diagnosed reason for children was that they got head and face mass, polydipsia, urine. At the same time, we toke MR scan (T1W1) and found that the high signal of neurohypophysis disappeared, unfounded in six cases, pituitary stalk thickened in five cases, pituitary stalk distinctness in one case, pituitary plumped in one case, and the signal of pituitary and pituitary stalk all changed by MR scan retest after treatment. Conclusion: Pituitary high signal disappears, pituitary stalk thickening or distinctness, are the MR imaging performance for the Langerhans cell histiocytosis invading brain. Polydipsia and polyuria will almostly appear if pituitary is invaded by the Langerhans cell histiocytosis. Currently, MRI is the only reliable imaging diagnosis for the Langerhans cell histiocytosis invading the pituitary, and has an important role for follow-up.

Key words: The Langerhans cell histiocytosis;Neurohypophysis;Pituitary stalk magnetic resonance imaging

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朗格罕细胞组织细胞增生症(Langerhan Cell Histiocytosis, LCH)是以树突状细胞和网状细胞系统增生为特征的一组疾病,缺乏恶性病变的组织学特征,但行为上表现为侵袭性生长[1]。现将搜集的6例确诊为朗格罕细胞组织细胞增生症并累及垂体的情况报告如下。

1 资料与方法

1.1 临床资料

搜集了我院经病理诊断为朗格罕细胞组织细胞增生症并行头颅MRI检查的患儿6例。其中,男性患者5例,女性患者1例,年龄为2～11岁,平均年龄为(6± 3)岁。

1.2 临床表现

全部患者均有多饮、多尿表现,表现为头面部肿块4例,其中2例同时有背部或臀部疼痛表现。1例表现为咳嗽、呼吸困难。神经系统查体正常。患儿发育基本正常,营养尚可。

1.3 检查方法

1) MR应用GE signa excite 1.5T核磁共振机[2],使用头线圈。常规SE T1WI(TR 450ms,TE 15ms)横轴、矢状、冠状面扫描,T2WI (TR 550ms, TE 100ms) 横轴、矢状面扫描。1例行静脉注射钆喷替酸葡甲胺(Gd-DTPA) 0.1mmol/kg后,行T1WI矢状面扫描。层厚:3～8mm,间隔:0.3～0.8mm,矩阵:240× 256,层数5～12层。

2) 2例行头颅CT检查,应用GE signa excite 64排CT机横断螺旋扫描后进行三维重建[3]。

3) 4例分别摄头颅、胸椎、骨盆DR片[4]。

2 结果

2.1 LCH累及垂体的MRI表现

1) 垂体窝后部正常神经垂体高信号消失5例,1例神经垂体纤细,后复查神经垂体消失。其中3例治疗后复查(4-14个月不等)仍未见神经垂体高信号显示。

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2)垂体柄增粗、居中 5 例,横径 4—5mm。垂体柄著征 1 例,横径约 3.5mm。1 例增强 T1WI 显示垂体柄明显强化。其中 3 例治疗后复查垂体柄增粗均有好转。

3)垂体饱满 1 例—1 岁男垂体高约 8.4mm,静脉注射造影剂后见垂体后份大片未强化区。垂体前缘略凹陷 1 例—4 岁男垂体高约 2.9 mm。其余 4 例垂体高约 4.5—5.2 mm 不等。其中 3 例治疗后复查垂体高度均较前有变薄。

4) 本组 6 例患儿脑实质均未见异常。

5) 其他累及颅骨或头皮软组织者见长 T1 长 T2 异常信号。骨质信号不连续或不均匀。其中 3 例治疗后复查软组织肿块缩小或基本消失,骨质缺损区亦较前缩小。

2.2 LCH 的 CT、DR 表现

1) 2 例行头颅 CT 表现为颅骨或伴眶骨、上颌窦多发不规则大小不等骨破坏区,呈软组织密度,呈膨胀性生长或虫蚀样改变,未见骨质增生硬化。其中 1 例累及眼外肌。

2) 1 例行骨盆 DR 片表现为左髂骨大片骨破坏区及 T6 椎体变薄呈铜钱状,数月后复查破坏区缩小,椎体增厚。1 例行骨盆 DR 片表现为左股骨上端骨密度减低区。1 例行胸部 DR 片表现为双肺多发气囊性病变、囊壁厚薄不均并双侧气胸。

3 讨论

3.1 郎格罕细胞组织细胞增生症 (Langenhans cell histiocytosis, LCH)

原称组织细胞增生症 X,是一组原因未明的组织细胞增殖性疾患。传统依据临床主要分为三种类型,即勒雪氏病 (Letterer-Siwe 氏病),韩雪柯氏病 (Hand-Schuller-Christian 氏病) 及骨嗜酸性肉芽肿 (eosinphilic granuloma of bone)。它们有共同的病理特点,彼此无严格界限,并可相互转化[5]。

3.2 LCH 病因及病理

病因不十分明确,有认为是一种原发性免疫缺陷性疾病,是朗格罕细胞突发性非肿瘤样异常增生和播散所致的单器官或 / 和多器官内的肉芽肿样损害。病理变化分增生期、肉芽肿期、黄色瘤期、纤维瘤期。

3.3 正常垂体解剖及 MRI 表现

1) 垂体前叶、后叶的胚胎学发生和血液供应都是不同的[6]。垂体前叶发生在原始口腔外胚层,是内分泌器官,血供来自垂体门脉系统。垂体后叶由间脑的神经外胚层向下生长形成,血供来自颈内动脉小分支,主要成份是以抗利尿激素为主的神经内分泌颗粒组成。垂体柄是下丘脑和垂体前、后叶间的通道。

2) 正常垂体前后叶在 MRI 上均可清晰显示。垂体前叶的大小、形态和信号强度与年龄和性别关系密切。新生儿期垂体上缘凸起,在 T1WI 上呈明显高信号[7]。从第二个月至成人期垂体上缘逐渐变平,垂体前叶在 T1WI 和 T2WI 呈与白质信号相当。垂体前叶儿童期高度 2～6 mm,青春期垂体高度女性可达 12 mm ;男性可达 7～8 mm。垂体后叶在出生后的第二个月,即表现垂体窝内鞍背前方的 T1WI 点状高信号,神经内分泌颗粒是该组织在 MRI 上 T1WI 图像中形成高信号的基础。说明下丘脑－垂体轴的完整性[8]。垂体后叶信号强弱与性别无关,垂体后叶 MR T1WI 上高信号的强度与年龄呈负相关, 信号强度随年龄的增加呈下降趋势[9]。但值得注意的是,正常人中约 10% 左右神经垂体高信号缺如[10],而呈等信号。垂体柄呈锥形,边缘光滑,自上而下逐渐变细,上份横径约 3 mm,下端横径约 2 mm[11]。

3.4 LCH 累及垂体的 MRI 表现

1) 神经垂体高信号消失。经临床治疗后,该组患儿复查后均未见神经垂体高信号显示, 那么 LCH 患儿经治疗后神经垂体高信号能否恢复还有待探讨。但因为神经垂体高信号缺如可见于 10%左右的正常人[12]及中枢性尿崩症 (CDI) 患者,要注意加以区别。亦要注意和异位神经垂体鉴别。

2) 垂体柄增粗或著明。该组患儿经临床治疗后复查垂体柄增粗均有所好转改善。

3) 本组垂体前叶异常几率较小,仅 1 例,但正常范围内垂体前叶高度在治疗后复查中有所缩小[13],其原因尚不明确。

3.5 LCH 累及垂体的临床相关性

本组患儿男性明显多于女性,临床多饮、多尿与 MRI 表现的神经垂体高信号消失, 其正常分泌抗利尿激素(antidiuretic hormone ADH)功能丧失相印证[14]。垂体前叶高度虽有变化,但下丘脑激素通过垂体门脉系统对垂体前叶作用无障碍,此亦与临床改组患儿无发育异常相符。

3.6 LCH 的特点

LCH 以全身多器官、系统受累为主,具有多发性和此起彼伏特点[15],该组患儿与文献报道一致,未见垂体单一受累者。

3.7 LCH 病人的转归及治疗

LCH 所引起病理改变的临床症状可以较轻, 并可有自限自愈的修复过程, 治疗取决于病变范围和受累器官的数量,合理的化疗对修复更有促进作用[16]。有尿崩症 (DI) 症状者可用 1-去氨 -8-D 精氨酸加压素治疗。

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