

左髋关节滑膜软骨瘤病误诊为色素沉着绒毛
结节性滑膜炎 1 例

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Synovial chondromatosis of left hip joint misdiagnosed as pigmented villonodular synovitis: a case report YUAN Ming-guan, CHEN Kun, ZHU Feng-feng, KANG Wu-lin, YUAN Pu-wei, DONG Bo, and WANG Zhan-kui*. *Affiliated Hospital of Shaanxi University of Traditional Chinese Medicine, Xianyang 712046, Shaanxi, China
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患者,女,26岁,左髋疼痛不适4年余,加重伴活动受限半月余,于2020年7月27日入院。患者自诉4年前无明显诱因出现左髋部疼痛,疼痛为一过性,且发病频率较低(间隔2~3个月),故未予以重视。2年前,患者左髋部疼痛症状加重伴轻度活动受限,因妊娠原因未系统检查及治疗,自诉妊娠后疼痛明显缓解。半月前,或因劳累疼痛加重,伴活动严重受限,故至门诊检查,行骨盆正位DR示左侧股骨头和头颈交界处改变(图1a)。髋关节MRI考虑左髋关节色素沉着绒毛结节性滑膜炎(pigmented villonodular synovitis, PVS),左侧股骨头缺血囊变,双髋关节滑膜炎(图1b, 1c, 1d)。查体见:左髋部旁压痛,叩击痛,左下肢直腿抬高试验40°,左髋关节极度屈曲活动受限。为进一步明确诊断,行髋关节CT示左侧髋关节腔积液伴多发斑点状稍高密度影,符合色素沉着绒毛结节性滑膜炎。左侧髋臼及股骨头斑点状高密度影,考虑继发改变(图1e)。入院后完善相关检查,血沉、血常规、肝肾功、电解质、肿瘤系列等未见明显异常,排除手术禁忌证,于入院后第3天行“左髋关节色素沉着绒毛结节性滑膜炎切开清理,滑膜切除术”。患者取仰卧位,抬高左髋部,麻醉生效后常规手术铺巾。取左髋外侧纵行切口,全长约15cm,依次切开皮肤、皮下组织及筋膜层,仔细分离周围组织,暴露髋关节前侧滑膜及附属物,打开后未见髋臼

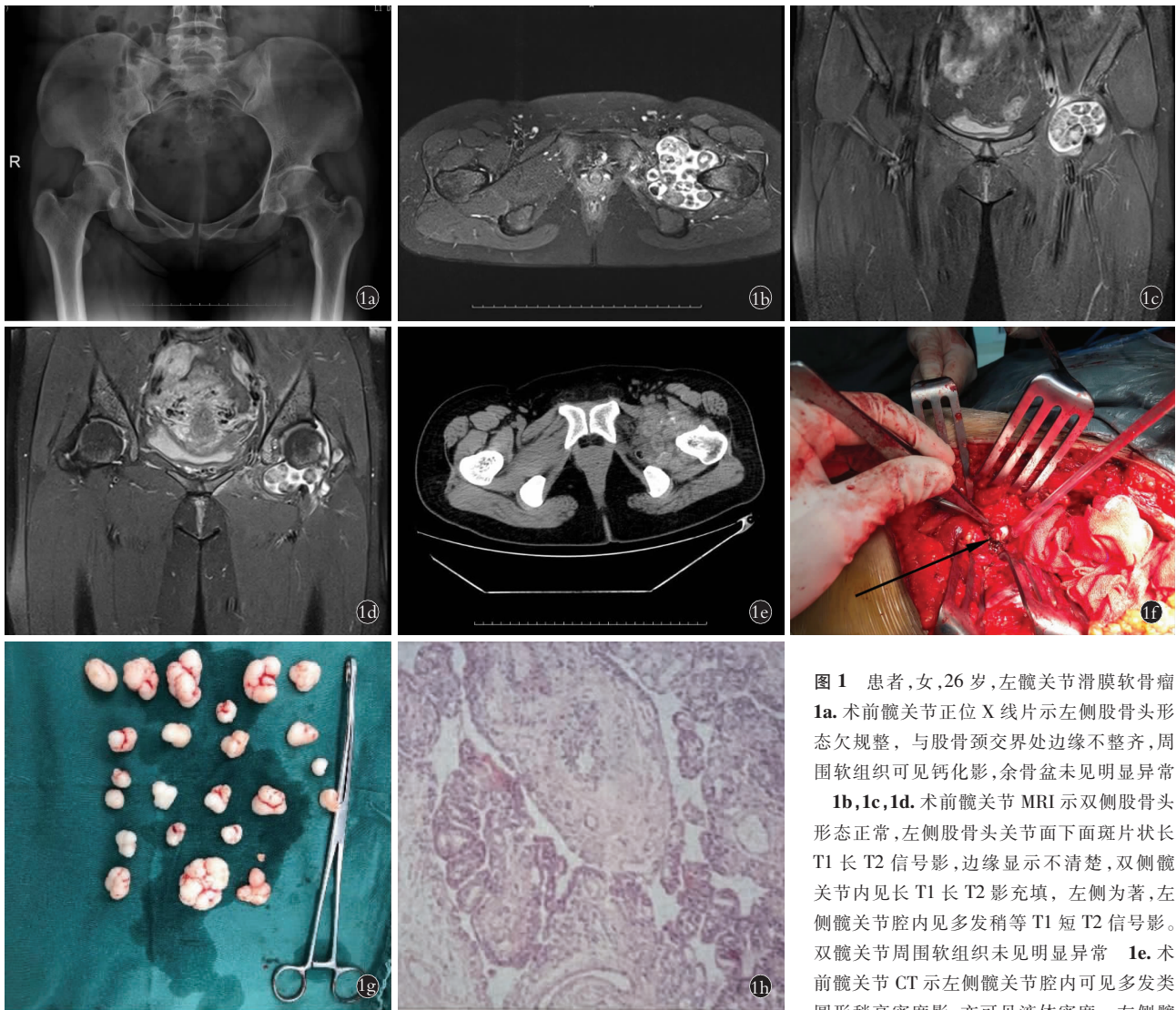
及股骨头局部铁锈色病灶,可见一灰白色游离体(图1f),棉球拭去血液,取出该游离体后,发现关节腔内仍有大量灰白色游离体,仔细完整剥离髋关节滑膜及游离体后,游离体质地稍硬,形状不规则,与周围组织无粘连。完整取出25个游离体(图1g)。清除关节内游离体并用电刀切除滑膜及血管翳。生理盐水反复冲洗伤口3次,仔细止血,查无活动性出血,清点纱块,留置引流条,逆向逐层缝合关闭切口。术中标本送病理学检查。术后病理学检查结果显示:左髋关节表面被覆滑膜细胞呈乳头状生长,间质水肿,纤维结缔组织增生(图1h)。术后修订诊断:左髋关节滑膜软骨瘤病(synovial chondromatosis, SC)。

讨论

滑膜软骨瘤病也称滑膜软骨性化生。本病病因尚不明确,目前多支持化生学说。多是关节滑膜因外伤、炎症等刺激化生为软骨而影响关节功能并出现相应临床症状的一种骨关节疾病^[1]。一般认为滑膜结缔组织内间充质干细胞向软骨细胞方向分化,形成软骨结节,软骨结节继续生长发生钙化、骨化,形成骨软骨小体,小体逐渐离开滑膜,通过一带与滑膜相连而形成悬垂体,之后其完全脱落进入关节腔内形成游离体^[2]。部分学者通过研究发现,少数可能恶变形成滑膜肉瘤或软骨肉瘤^[3-4]。好发于20~50岁,男女发病率约为2:1。此病好发以膝关节为主,其次为髋、肘、肩关节等,常累及一侧关节。本病具有早期诊断率低,临床易误诊,治愈率低的特点^[5]。

本病例发生于左髋关节,而髋关节滑膜软骨瘤病主要表现为髋关节疼痛、活动功能受限、肿胀,活动时关节有摩擦感,体表甚至可触及活动性包块为

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白及股骨头可见点状高密度影 1f. 术中发现灰白色游离体 1g. 完整取出 25 个游离体 1h. 病理学检查(HE×40)示左髋关节表面被覆滑膜细胞呈乳头状生长, 间质水肿, 纤维结缔组织增生

Fig.1 A 26-year-old female patient, synovial chondroma of left hip joint 1a. Preoperative X-ray film of the hip joint showed that the shape of the left femoral head was irregular, the edge at the junction with the femoral neck was irregular, calcification shadow was seen in the surrounding soft tissue, and no obvious abnormality was found in the other pelvis 1b, 1c, 1d. Preoperative MRI of the hip joint showed that the morphology of bilateral femoral heads was normal, patchy long T1 and long T2 signal shadow under the articular surface of the left femoral head, and the edge was not clearly displayed. Long T1 and T2 shadows were seen filling in both hip joints, especially on the left. Multiple slightly equal T1 and short T2 signal shadows were seen in the left hip joint cavity. No obvious abnormality was found in the soft tissue around both hips 1e. Preoperative CT of the hip joint showed multiple round like slightly high-density shadows and liquid density in the left hip cavity. Punctate high-density shadow can be seen in the left acetabulum and femoral head 1f. Gray white free body was found during operation 1g. Take out 25 free bodies completely 1h. Pathological examination (HE×40) showed papillary growth of synovial cells covered on the surface of the left hip joint, interstitial edema and proliferation of fibrous connective tissue

主要临床症状。关于本病的诊断,有学者认为只有在术中发现关节腔中的游离体,且经病理检查后方可明确诊断^[6]。而 Milgram^[7]根据病理将该病分为 3 期:第 1 期尚没有关节游离体形成;第 2 期有的带蒂与滑膜相连,有的脱落形成游离体;第 3 期形成多数游离体,游离体逐渐增大或变小,可继发钙化或骨化。

X 线征象:X 线片的表现有关节内及关节外有多数大小不一的圆形或卵圆形边缘光滑致密的钙化

或骨化阴影围绕。关节面和关节间隙一般无明显变化,无骨质破坏及骨质疏松等征象。少数有骨质侵蚀现象,多发生于关节囊滑膜附着处^[8]。

MRI 对本病的诊断尤为重要,对于尚未形成游离体时,无明显症状,且 X 线及 CT 难以对本病明确诊断,而骨化的游离体在 MRI T1 和 T2 加权像上都是低信号。未骨化的游离体和增生的滑膜结节在 T1 加权像上是低信号,而在 T2 加权像上是高信号^[9]。

本病误诊为髋关节色素沉着绒毛结节性滑膜炎的原因:(1)色素沉着绒毛结节滑膜炎是一种相对少见的疾病,发生于关节、腱鞘、滑囊附属纤维组织的滑膜呈绒毛状或结节状增殖的疾病^[10]。PVS 多发于膝关节,其次为髋关节。本病起病隐匿,初期可无明显症状,随着病情发展,可导致疼痛逐渐加重,关节僵硬,活动受限,跛行等症状。PVS 与 SC 在髋关节发病的症状上相似,较难鉴别。(2)本病例中的影像学检查:从 X 线中未观察到明显的圆形或卵圆形钙化及骨化阴影,且关节间隙较右侧变窄,左侧股骨头与颈交界处明显较右侧缩短。而 PVS 的 X 线征象一般以关节间隙变窄,伴有骨质破坏为主要特点^[11]。X 线与 CT 检查往往缺乏特异性,较难鉴别。本例的 X 线表现较符合 PVS,故为误诊的原因之一。MRI 在鉴别中起到重要的作用。PVS 最典型的 MRI 特点是:由于含铁血黄素的沉积,在 T1、T2 像上均表现为关节内低信号的结节性肿块^[12]。而本病例 MRI 的表现与 PVS 相似,故也是误诊的其中因素之一。(3)造成误诊的最重要原因是:因 MRI 及 CT 等影像学检查高度考虑髋关节色素沉着绒毛结节性滑膜炎,所以术前未进行组织病理学检查。而组织病理学检查是诊断 PVS 的金标准,PVS 镜下可见不等量的含铁血黄素沉着。

滑膜软骨瘤病与色素沉着绒毛结节性滑膜炎是临床容易混淆及需要相互鉴别诊断的两个病种,特别是当滑膜软骨瘤病的病变钙化或骨化不全时影像学上很难鉴别,需要病理学活检才能明确诊断。

本病的治疗还是以手术为主,对于少量游离体,可进行髋关节关节镜清理。游离体形成后长期刺激可致使关节软骨的磨损及进行性退变,大量游离体可致使髋部肿痛,活动受限等。所以发现后应尽早行手术治疗,摘除游离体并切除病变滑膜。对于病变滑膜,有的学者认为应切除,有的学者则认为应尽量保留。在本病例中选择了切除病变滑膜,仔细清理关节腔,尽量减少复发的可能。

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