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Inclusion body fibromatosis of the third toe of left foot: Case report 左足第3趾包涵体性纤维瘤病1例

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[关键词] 趾;纤维瘤;包涵体;磁共振成像

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患儿男,3岁,出生后发现左足第3趾包块,8个月前肿块明显增大,挤压两侧足趾。查体:左足第3趾触及突起,无压痛。X线片:左足第3~4趾骨间见软组织肿块。MRI:左足第3趾骨外侧见肿块影,大小约3.2 cm×2.8 cm×2.1 cm,边界清楚,形态较规则,呈稍长T1(图1A)、稍长不均匀T2(图1B)信号,其内见结节状、条索状低信号;增强扫描病变呈明显不均匀强化(图1C),肿块包绕第3趾骨生长,

局部可见骨质吸收,第4趾骨呈受压推移改变。影像学诊断:考虑软组织肿瘤,倾向于间叶组织、滑膜来源。行左足第3趾肿块切除+皮瓣转移术。术中见肿块与第3趾骨关系密切,呈淡黄色,质硬,边界不清。大体病理示肿块切面灰白质韧;镜下见梭形细胞增生形成境界尚清楚的结节样结构,长梭形细胞呈束状或车辐状排列,胞浆中等,其间见小圆形红染的包涵体(图1D)。免疫组织化学染色: β -catenin(+),CD68(+),S-100(个别+). 病理诊断:(左足第3趾)包涵体性纤维瘤病(inclusion body fibromatosis, IBF)。

讨论 IBF是罕见的纤维母细胞/肌纤维母细胞来源良性

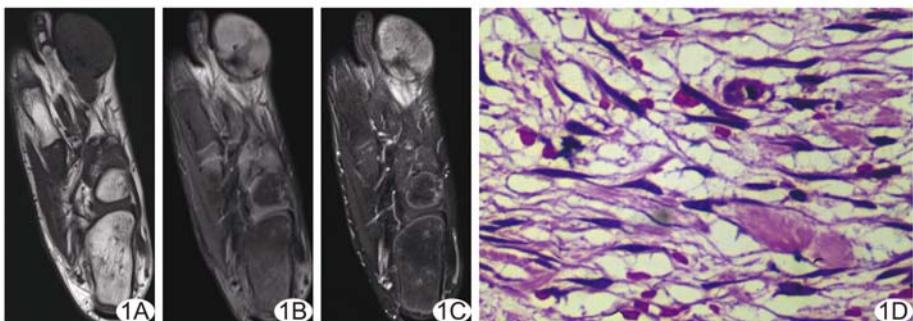


图1 左足第3趾IBF A. T1WI; B. T2WI; C. 增强T1WI; D. 病理图(HE, ×400)

肿瘤,多发生于1岁以内婴幼儿手指或足趾背侧面,30%患儿于出生时即发现此病。IBF由增生的纤维母细胞构成,其内混有肌纤维母细胞和交错排列的细胞外胶原纤维束,特征性表现为肿瘤细胞胞质内大小不一、圆形、淡嗜伊红色包涵体。MRI示IBF呈稍长T1稍长不均匀T2信号,内可见条索状低信号,考虑为增生的胶原纤维束。本病应与其他好发于儿童四肢末端的病变相鉴别,如钙化性腱膜纤维瘤、血管球瘤等。IBF明确诊断需依靠病理学检查,仔细观察并寻找有无嗜酸性包涵体是鉴别诊断的关键。临床多采用肿块切除+皮瓣转移术治疗IBF,但术后复发率高达60%。本例IBF患儿出生后即发现左足第3趾包块,缓慢生长,治疗后18个月未复发。

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