ISOLATED ORBITAL NEUROFIBROMA： CLINICAL PRESENTATION AND DIFFERENTIAL DIAGNOSIS

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Isolated orbital neurofibromas without systemic neurofibromatosis are extremely rare. We report one such case and describe our experience with this patient. A 27y/o male patient presented with painless, palpable mass located at the left medial upper orbital area for one year. He had no prior trauma history, family history or other evidence of neurofibromatosis type I. Ocular examination revealed downward displacement of the left globe while computed tomography identified a localized solid mass of the superior orbit. An anterior orbitotomy with tumor resection was performed afterward. Histopathological examination proved the mass to be a localized orbital neurofibroma. The authors emphasize that the unusual occurrence of localized neurofibroma in the orbit with the absence of systemic neurofibromatosis requires careful differentiation from other orbital tumors by radiographic and histopathological studies.

Key words: isolated orbital neurofibroma；localized orbital neurofibroma；differential diagnosis

INTRODUCTION

Orbital neurofibromas are rare, accounting for 0.5 to 2.4 % of all orbital tumors[1]. Isolated neurofibromas of the orbit are relatively uncommon, representing less than 1% of orbital neoplasms[2]. Isolated neurofibromas of the orbit are benign, solitary and slowly growing tumors. They are one of the nerve sheath tumors (NSTs) which are derived from the Schwann and perineurial cells of the peripheral nervous system. The 3 typical ocular manifestations of neurofibromas are plexiform, diffuse and localized[3]. Isolated (localized, circumscribed, or solitary) neurofibromas are distinct from plexi- form and diffuse neurofibromas and are less frequently associated with neurofibromatosis type I (also known as Von Recklinghausen's disease).

We report a rare case of the isolated orbital neurofi-

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