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Clinical-Medical Image

Bag of Warms Diagnosis in One Eye of Nodular Plexiform Neurofibroma

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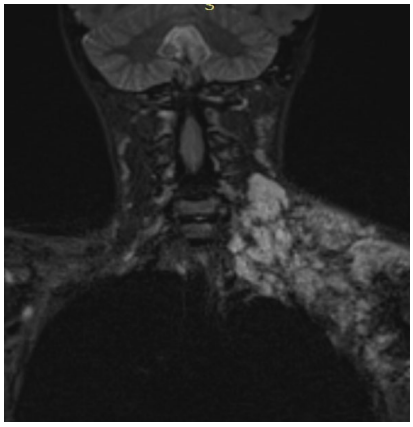

Figure 1: Coronal section MRI showing a serpentine and tortuous aspect of the nerves of the left brachial plexus in T2 FAT-SAT hyper signal giving a worm bag aspect.

Figure 2: Bag of warms

Clinical-Medical Image

Neurofibromatosis is a group of autosomal dominant diseases with two categories: neurofibromatosis type 1 and neurofibromatosis type 2.

Neurofibromatosis type 2 has an incidence of one in 33,000 to 40,000 births, its gene is located on chromosome 22, and is characterized mainly by the presence of bilateral vestibular schwanomas [1]. Neurofibromatosis type 1 or Von Recklinghausen disease is the most frequent of the phacomatoses, with an incidence of approximately one in 3,500 births, its gene is located on chromosome 17. Clinically, it is manifested by cutaneous manifestations (cafe au lait spots, ephelides, cutaneous neurofibromas, lisch nodules), neurological manifestations, namely tumors of the central nervous system, tumors of the peripheral nervous system including diffuse plexiform and nodular neurofibromas, as well as bone manifestations (sphenoidal dysplasia, scoliosis, etc.) The diagnosis of neurofibromatosis type 1 is based on a certain number of criteria according to the Consensus Conference of the National Institute of Health in Bethesda [2].

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Neurofibromas are grouped into three types: cutaneous neurofibromas, nodular neurofibromas and plexiform neurofibromas (diffuse and nodular). The plexiform neurofibromas are pathognomonic of neurofibromatosis type 1, they correspond for the nodular form to a fibro-myxoid proliferation of the sheath of an extended segment of a nerve or its branches which leaks into the fat, along the muscles and into the subcutaneous tissue thus giving a tortuous and serpiginous aspect of the nerve "worm sack aspect" (Figures 1 and 2). Thus, for the diffuse form, it corresponds to an elevation of the skin in plate with diffuse thickening of the subcutaneous plane.

Imaging contributes to the diagnosis of neurofibromas thanks to their characteristic appearance; on the CT scan, the neurofibromas appear isodense in relation to the muscles with weak enhancement after injection of the contrast medium; on the MRI, the appearance is isosignal T1 in relation to the muscles and hyper signal T2 with intense enhancement after injection of the contrast medium [2].

Declaration of interest

The authors declare that they have no ties of interest

Keywords: Benign tumor; Nerve; Neurofibromatosis

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