Case Report

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Cervical and Spinal Plexiform Neurofibroma: A Case Report

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ABSTRACT

Plexiform neurofibroma is a rare benign tumor of the peripheral nerves at the expense of perineural connective cells. It is pathognomonic of neurofibromatosis type 1 (NF1 or Von Recklinghausen disease). MRI is of great help in the diagnosis of this pathology. Anatomopathological confirmation is sometimes necessary, especially in the absence of a context suggestive of NF1. We report the observation of an young boy with a cervical plexiform neurofibroma revealing a neurofibromatosis Type 1.

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Introduction

Neurofibromas are benign tumors that develop from the roots and plexuses of spinal nerves. They may be uni or bilateral, sometimes stepped, superficial or deep. Neurofibromas are either cutaneous, diffuse or plexiform. Plexiform neurofibromas correspond morphologically to a more or less long segment of tortuous dilatation of a nerve and its branches taking on a worm sack appearance. Plexiform neurofibromas are pathognomonic of NF1. They are generally slow-growing tumors. Their symptomatology is variable depending on their topography. We report the case of a 14 year-old boy who consulted for two lumbosacral and left cervical masses.

Patient and Observation

A 14-year-old boy, without any particular pathological history, consulted us for two left latero-cervical and lumbosacral masses of progressive evolution, which were aggravated after a trauma to the cervical and lumbosacral spine complicated by the appearance of disabling neuralgia. The clinical examination found two masses of painless consistency with poorly defined borders and no inflammatory signs. The lymph nodes were free and the skin examination revealed multiple café au lait spots on the back and limbs (more than 5). A CT scan of the lumbosacral and cerebral spine with cervical sections was performed as part of the trauma assessment, which showed:

CT of the Lumbosacral Spine

A large subcutaneous hematoma of the dorsal wall of the lumbosacral region, lateralized to the right, spontaneously hyperdense, surrounded by significant soft tissue infiltration, measuring 55x160x140 mm (APxTxCC).

Cerebral CT

Individualization of a subcutaneous retro-mastoid and left occipital soft tissue mass with a tissue and fluid component crossed by vessels, associated with serpentine vascular formations creating a vascular lace with an afferent artery from the homolateral internal

carotid artery which is small downstream and the presence of a bone defect opposite which may be related to an AVM. Faced with these inconclusive results, a spinal cord and brain MRI was requested which showed:

At the Brain Level

Individualization of a subcutaneous lesion process centered on the high posterior cervical region and extending opposite the occipital bone lateralized to the left, in heterogeneous T1 iso signal, heterogeneous T2 hypersignal, intensely and heterogeneously enhancing after gadolinium injection, with individualization of a richly vascularized region in the lower left occipital area with a fibrillary appearance, scalloping on the left occipital with bone lysis and arriving in intimate contact with the left lateral sinus which is compressed but remains permeable, with no signs of intra parenchymal invasion, measuring approximately 120 x 24 x 76 mm (T x AP x CC).

At the Medullary Stage

Individualization of a subcutaneous mass in the lumbosacral region developing medially and laterally on the right in heterogeneous T1 hyposignal with a component in hyper signal not erased on the FS sequence, heterogeneous T2 hypersignal, It enhances intensely and heterogeneously after injection of gadolinium with enhancement of the surrounding subcutaneous fat, and comes into contact with the posterior lumbosacral arches with a dyraphic region of the S2 without endo-canal extension, measuring 182 x 251 x 70 mm (CC x T x AP).

It is associated with a dorsa-lumbar scoliosis with left convexity the diagnosis of occipital and lumbosacral plexiform neurofibroma was made.

Discussion

Plexiform neurofibroma is a rare benign tumor of the peripheral nerves the expense of the connective cells of the perineura.

Its non-capsulated nature explains the diffuse infiltration of adjacent nerve trunks, cellulo fatty tissue and muscle. It is often considered pathognomonic of neurofibromatosis type 1 (NF1 or Von Recklinghausen disease) and is seen much more frequently than Schwannoma in NF1 [1, 2]. NFP belongs to the four types of neurofibromas encountered in NF1 according to the 198 National Institute of Health Development Consensus Conference classification. The diagnosis of the disease is based on the presence of at least two of the following criteria: At least six café au lait spots larger than 5mm before puberty and larger than 15mm after puberty; Two or more neurofibromas, one or more plexiform neuromas; Lentiginous spots of the axillary or inguinal region; Two or more hamartomas of the iris (Lisch nodules), one optic tract glioma; A characteristic bony lesion (pseudarthrosis of a long bone, spheno orbital dysplasia, cervical kyphosis). Isolated plexiform neurofibromas may be observed outside the context of NF1, the diagnosis of which must remain a diagnosis of elimination. It would appear that some of these cases are part of a segmental neurofibromatosis (NF5) [3-5]. Rapid growth may occur during puberty or pregnancy without spontaneous regression they may be uni or bilateral and sit at different levels so their symptomatology is variable depending on their topography. NFP develops from the nerve branches of the fifth, seventh or ninth cranial pair during craniofacial localization. At the cervical level, the neurofibroma or plexiform neuroma may cause spinal cord compression or may extend to the sympathetic or brachial plexus resulting in Claude Bernard Horner syndrome or peripheral nerve palsy [6]. On the thoracic and abdominal levels, they pose a problem of differential diagnosis with tuberculosis, lymphoma or sarcoidosis [1, 7]. In the pelvic region they simulate adenopathies or abscesses of the psoas. Imaging is of great help in the diagnosis, allowing the lesions to be characterized with a view to a positive diagnosis, to search for possible associated lesions, to assess the prognosis and to carry out a follow-up. On ultrasound, NPs are tortuous lobulated masses with a clustered, hypoechoic appearance, with well-defined contours oriented along the axis of the nerve trunk on longitudinal sections and characterized by the Target sign on transverse sections with an echogenic centre and a hypoechoic periphery. Cystic formations can be seen within the mass as well as posterior enhancement found in 70% of cases. Color Doppler reveals different types of 8; moderate, central or predominantly peripheral vascularity. Some may be poorly vascularized [8]. CT study reveals nodular, fusiform or clustered lesions, less dense than muscle (20-30HU). This low density is explained by the presence of lipid inclusions in Schwann cells, adipocytes, cystic degeneration and a mixed stroma. The behaviour after injection is variable: homogeneous or heterogeneous contrast [1]. The NP appears on magnetic resonance imaging sequences, which is the reference radiological examination, by a relative T1 hypo signal in relation to the muscle, T2 hyper signal and when it is voluminous, it may contain a central hypo signal producing a characteristic cocoon aspect. The enhancement is variable: central, diffuse, peripheral, target [2, 9]. The diagnosis of NFP is essentially anatomopathological, particularly outside the context suggestive of neurofibromatosis type I. Formerly called plexiform neuroma or royal tumor, NFP differs from other types of neurofibromas by the importance of its schwannian component [10]. The risk of sarcomatous degeneration justifies, whenever technically possible, the removal of the lesion as completely as possible. Finally, these patients should be monitored clinically and radiologically by a multidisciplinary team, at least once a year until the age of ten, and then regularly, in order to assess any recurrence or malignant transformation [5].

Figure 1: Cervical MRI, T1 weighted axial section without injection
Gadolinium: Right latero-cervical process in T1 hypersignal

Figure 2: Cervical MRI, T2-weighted axial slice: of a subcutaneous lesion process centered on the high posterior cervical region extended opposite the occipital bone lateralized to the left, heterogeneous T2 hypersignal

Figure 3: Cervical MRI, sagittal section in weighted sequence T1 after gadolinium injection: the mass is strongly enhanced after gadolinium injection

Figure 4: Spinal cord MRI, sagittal section in T1-weighted sequence: of a subcutaneous mass in the lumbosacral region developing medial and lateral right in T1 heterogeneous hyposignal with unblurred hyper signal component on the FS sequence, T2 heterogeneous hypersignal

Figure 5 : Spinal cord MRI, axial section in T1-weighted sequence after injection of gadolinium: intensely and heterogeneously enhancing after injection of gadolinium with enhancement of the surrounding subcutaneous fat

Conclusion

Plexiform neurofibromas are rare benign tumors of the peripheral nerves. They are pathognomonic tumors of NF 1 which is a multisystemic pathology with clinical and radiological polymorphism. MRI is the examination of choice in the exploration of this pathology. It provides decisive arguments for the positive diagnosis, the evaluation of the prognosis and the follow-up of the lesions.

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