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Spinal thoracic tuberculoma in neurofibromatosis type-1

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Keywords
extrudal, hydrocephalus, neurofibromatosis type-1, para-vertebral mass, spinal thoracic pain, spinal tuberculoma, tuberculosis meningoencephalitis

ABSTRACT

The clinical manifestations of neurofibromatosis type-1 have in common the presence of neurofibromas, schwannomas and cafe`-au-lait macules, which can potentially appear within any organ system of the body, involving primarily the skeleton, skin and soft tissues. The spinal thoracic tuberculoma in neurofibromatosis is exceptional.

We report here, the case of a 31-year-old male with neurofibromatosis type-1 who presented a year agoa spinal thoracic pain. The evolution was marked by the appearance of a dorsal para-vertebral mass, progressively increasing in volume for 03 months.

Neurological Examination revealed normal muscle bulk and tone, and power was grade 5/5. Deep tendon reflexes were brisk and the Babinski sign was present. The sensory deficit was present below the D9 level. The remainder of the examination found a voluminous dorsal para vertebral mass ovoid, renitent, and adhering to the superficial and the deep planes. There were also "café-au-lait" spots disseminated on his body. The imagery showed a formation with respect to D11-D12, isointense on T1 and T2, heterogeneously enhanced after contrast with central necrosis, with a foraminal starting point measuring 85x78x66 mm, with external canal extension. Laminectomy D11-D12 was performed with incomplete excision of the lesion. The histological examination showed casein-follicular vertebral tuberculosis, with secondary abdominal changes. Then the patient was put on anti-bacillary drugs. The evolution was marked two weeks later by the reappearance of the back pain and the back mass. On the 42nd day of treatment, the patient had a febrile consciousness disorder with a GCS of 8, right mydriasis and meningeal stiffness. Paraclinic investigations revealed tuberculosis meningoencephalitis, responsible for an active ventricular hydrocephalus associated with a left temporoparietal extrudal hematoma. An external ventricular shunt was performed as well as a left temporo-parietal extrudal hematoma evacuation. The patient died 7 days later in a poly visceral failure chart.

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Introduction

Neurofibromatosis is the most frequent single-gene disorder affecting mankind [1]. It is a disorder of neural crest cells defined as a spectrum of multifaceted diseases, probably hamartomatous in origin, involving neuroectoderm, mesoderm and endoderm [2]. Its clinical manifestations have in common the presence of neurofibromas, schwannomas and cafe-au-lait macules, which can potentially appear within any organ system of the body, involving primarily the skeleton, skin and soft tissues. The spinal thoracic tuberculoma in neurofibromatosis is exceptional. This case appears to be the first reported case in the literatureof this kind of tumor.The aim of this case is to report the clinical outcomes following resection ofspinal thoracic tuberculoma.

Case report

A 31-year-old male of low socioeconomic status, with a history of neurofibromatosis type-1, admitted to the neurosurgery department for a para-vertebral mass of progressive installation evolving for 03 months. The history of the disease dates back to a year before his admission by back painreticent to symptomatic treatment. The development was marked by the appearance of a dorsal para-vertebral mass, progressively increasing in volume for 03 months. The whole evolving in an apyretic context and conservation of the general state. The initial clinical examination was aimed at a conscious, generally good, apyretic and hemodynamically and respiratoryly stable patient. Neurological examination revealed normal muscle bulk and tone, and power was grade 5/5. Deep tendon reflexes were brisk and Babinski sign was present. Sensory deficit was present below D9 level. The locoregional examination found a voluminous dorsal paravertebral mass, ovoid, renitent, adhering to both superficial and deep planes. It measured 23x16 cm with long axis. There were also "café-au-lait macules" scattered around his body (Fig.1).

The medullar MRI revealed a formation with respect to D11-D12, in isotense T1 and T2, heterogeneously enhanced after contrast with central necrosis, with a foraminal starting point measuring 85 x 78 x 66 mm, with external canal extension that measured 140 x 45 mm. We conclude that all of the abnormalities described fall within the scope of an NF1 in its plexiform form with degenerate neurofibroma D11-D12 (Fig.2). Viral serology (HIV, syphilis and hepatic) was negative.

Figure. 1: "café-au-lait macules" scattered around his body and voluminous dorsal paravertebral mass.

We performed median cutaneous incision with respect to D10-D11. Cutaneous dissection of the cyst lesion, showed the sero-viscous contents of which have been removed as a whole. More realization of a laminectomy of D10-D11 and complete excision of an epidural and intracellular lesion of the intra-dural one. This lesion is friable infiltrating with necrosis (Fig.3).

The histological examination showed the casein-follicular vertebral tuberculosis, with secondary abdominal changes (Fig.4). Then the patient was put on anti-bacillary drugs. The evolution was marked two weeks later by the reappearance of the back pain and the back mass. Postoperative spinal MRI showed a slight increase of the tumor formation with respect to D11-D12, in isotense T1, T2, heterogeneously enhanced after contrast with central necrosis, with a foraminal starting point measuring 88x95x88 mm, with external canal extension (Fig. 5).

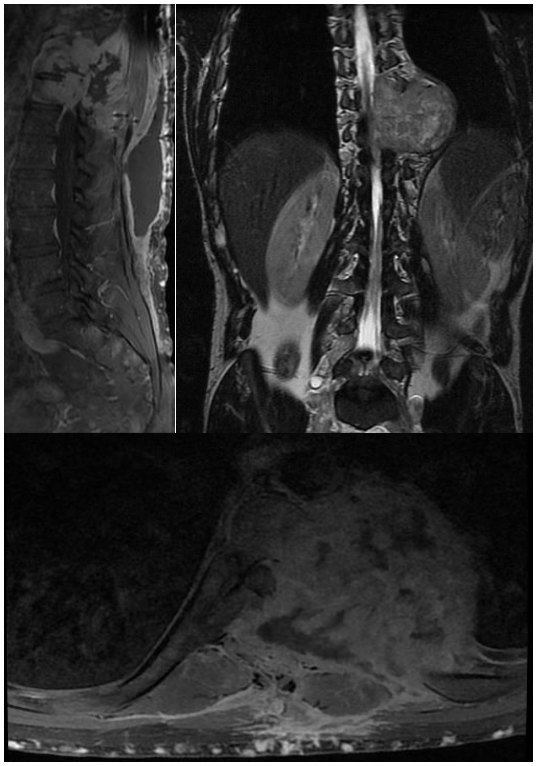

Figure. 2: The medullar MRI revealed a formation with respect to D11-D12, in isotense T1, T2, heterogeneously enhanced after contrast with central necrosis, with a foraminal starting point measuring 85x78x66 mm, with external canal extension that measured 140x45 mm.

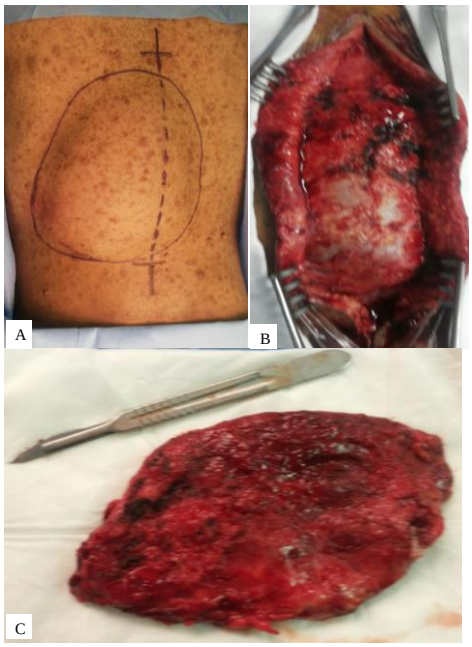

Figure 3: Intraoperative images. A: Anatomical identification with paravertebral swelling, B: the extra-canal and intradural lesion infiltrating the dura mater, C: the operative piece showing caseous necrosis.

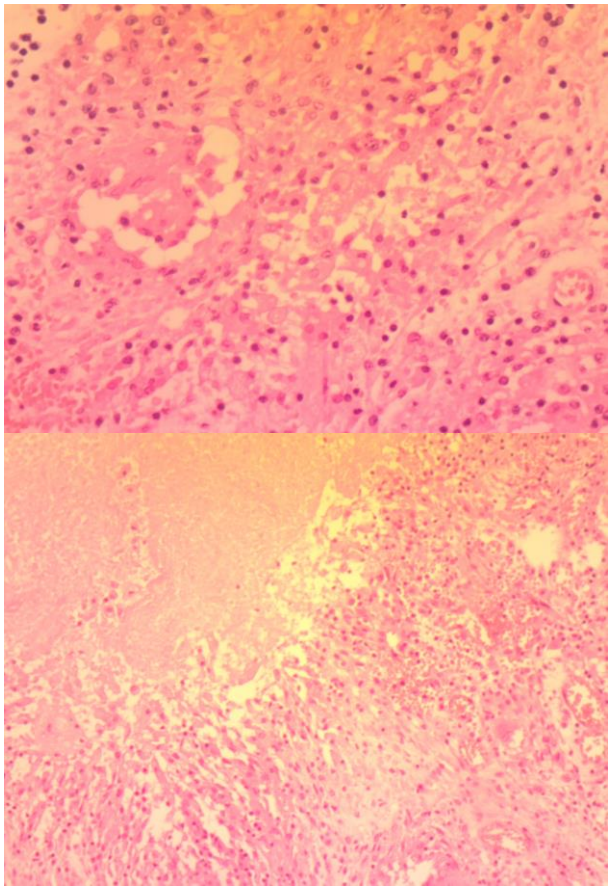

Figure 4: The histological examination showed the caseo-follicular vertebral tuberculosis, with secondary abdominal changes.

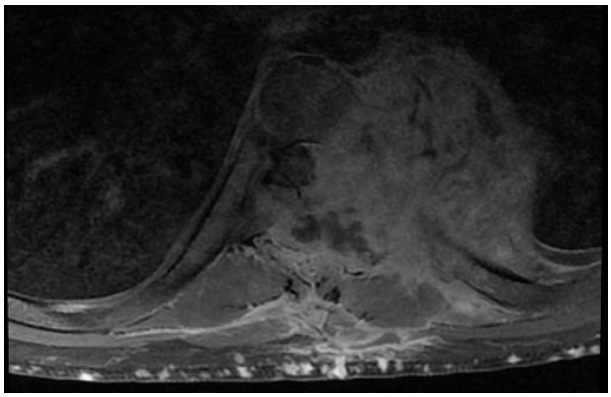

Figure 5: Postoperative spinal MRI showed a slight increase of the tumor formation with respect to D_{11}-D_{12}, in isotense T1, T2, heterogeneously enhanced after contrast with central necrosis, with a foraminal starting point measuring 88 x 95x88 mm, with external canal extension.

At one month of the anti-bacillary treatment, the patient had presented a febrile subicteria with an episode of partial convulsive seizures secondarily generalized. Brain CT-scan showed osteolytic lesions on the right side of the frontal bone (Fig. 6. A).

Biologically, there was an increase in gamma GT at twice the normal. On the 42nd day of treatment, the patient had a febrile consciousness disorder with a GCS of 8, right mydriasis and meningeal stiffness. Brain CT scan had demonstrated ventricular hydrocephalus with a left temporo-parietal extradural hematoma (Fig. 6. B, C). We performed an external ventricular shunt (EVS) in an emergency. The study of ventricular CSF was predominantly lymphocyte-positive at 90%. The brain imagery after EVS showed an increase in the size of the extra-dural hematoma (Fig.6.D) which was evacuated. The patient died 7 days later in a poly visceral failure chart. The autopsy was proposed to find out the exact cause of the death but the family refused because of the usual practices.

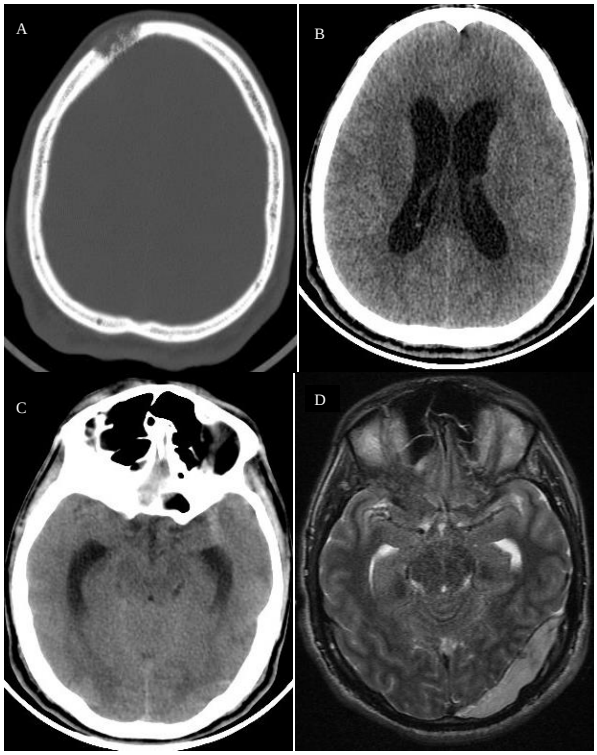

Figure 6: (A) Brain CT scan showed osteolytic lesions on the right side of the frontal bone, (B) Brain imagery had demonstrated ventricular hydrocephalus; (C) left temporo-parietal extradural hematoma; (D) brain imagery after EVS showed an increase in the size of the extra-dural hematoma.

Discussion

Neurofibromatosis type 1 (NF1) is one of the most fascinating and common human mendelian disorders, affecting approximately one in 3000 persons [5]. From the initial artist renderings of patients with NF1 in the 15th century and the earliest medical reports in 18th century, to the complex molecular genetic studies of the late 20th century, physicians and lay persons alike have been fascinated with this disease because of its diverse manifestations and the unusual and bizarre physical appearances associated with the disease. Also known as peripheral neurofibromatosis or von Recklinghausen disease, NF1 is inherited in an autosomal dominant pattern. The disease bears the name of Friedrich von Recklinghausen (1833 - 1910), a German pathologist, who was not the first to report the disease but was the first to recognize that the characteristic peripheral neurofibromas developed from nervous tissue [7]. Patients with NF1 are afflicted with a diverse group of lesions that are predominantly neuroectodermal or mesenchymal in origin. The large and complex NF1 gene, located on chromosome 17, encodes a protein named neurofibromin that works to control cellular proliferation through complex interactions with rasoncogenes [8]. Its clinical manifestations have in common the presence of neurofibromas, schwannomas and cafe-au-lait macules. The spinal tuberculoma is exceptional.

Tuberculoma pathophysiology is complicated to understand.First infected macrophages produce inflammatory mediators which lead to recruitment of peripheral macrophages and monocytes. These infected macrophages migrate to lymph nodes and disseminate to in other part of body. Enzymes in lysosome kill mycobacteria then present their antigens via major histocompatibility complex (MHC) class II to CD4 + T cells; CD4 + T-cells secrete interferon-gamma (IFN-c), which induce macrophage recruitment and enhance lysis of mycobacteria [4]. The patient body react by forming granuloma around the infected and necrosed immune cells to contain it, but viable bacteria has been demonstrated in these granulomas. This granuloma formation is though protective sometime may turn against the patient recovery. This inflammatory process may get exaggerated and produce unwanted effects.

The factors favoring the spinal thoracic tuberculoma being poverty and especially immunosuppressant, would explain its occurrence in NF1. Intradural tuberculous granuloma is the most unusual of all varieties of spinal tuberculous granuloma and can be classified as (a): endural (within two dural layers); (b): subdural; (c):

subarachnoidal or arachnoidal; (d): intramedullary and (e): after transgressing cord and arachnoid sited in subdural spase. According to this classification, our case is of type a.

According to the case of our patient, two kinds of parenchymatous involvement can be described: Type 1: tuberculous meningitis may be present without CNS tuberculoma formation. Rich [6] suggested that tuberculous meningitis is precededby metastatic seeding of Mycobacterium tuberculosis within the parenchyma or meninges, followed by growth andeventual maturation of the caseous tuberculoma into a fluid-filled tuberculous abscess and rupture of this abscess into the subarachnoid space causes tuberculous meningitis. In our case, the contamination of the CNS by the Mycobacterium tuberculosis could be explained by a dissemination of the tuberculoma from the operative focus. Type 2: The development of tuberculous meningitis is a two-step process. Mycobacterium tuberculosis bacilli enter the host by droplet inhalation. The initial point of infection being the alveolar macrophage. Escalating localised infection within the lung with dissemination to the regional lymph nodes produces the primary complex. During this stage there is a short but significant bacteraemia that can seed tubercle bacilli to other organs in the body. In those who develop tuberculous meningitis, bacilli seed to the meninges or brain parenchyma, forming small subpial or subependymal foci. These are called Rich foci. The dissemination to the CNS is more likely, particularly if miliary tuberculosis develops. The second step in the development of tuberculous meningitis is rupture of a Rich focus into the subarachnoid space.

The occurrence of tuberculous meningoencephalitis at the end of the anti-bacillary treatment would come either: The bacillus itself has many ways to protect itself from immune cells and antibiotics. One of the main mechanisms is the inherent resistant nature of mycolic acid present in the cell wall. The paradoxical aggravation under anti-bacillary marked by the reappearance of dorsal pain and back mass two weeks later. The absence of association of corticosteroid therapy with antibacillary therapy could induce this periodontal aggravation.

This complication of tuberculous meningitis may present despite adequate antituberculous treatment [3], as was the case in our patient.

Conclusion

The clinical manifestations of the neurofibromatosis have in common the presence of neurofibromas, schwannomas and “café-au-lait macules”. However, an exceptional form of the spinal thoracic tuberculoma can be seen thus mimicking the already described forms of neurofibromatosis. The management of this kind form would perform by an antibacillary therapy associated with corticosteroid therapy and the surgery for good outcome.

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