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Spinal Anaplastic Ependymoma In Neurofibromatosis Type 2 : Case Report

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Abstract

Neurofibromatosis type 2 (NF2) is a rare genetic autosomal dominant disorder, Its sign is the presence of bilateral vestibular schwannomas with multiple spine tumors. Ependymomas account for more than 75% of intramedullary spinal cord are usually low-grade and especially located in the lower cervical and upper thoracic spine, however, anaplastic ependymomas of spine appear to be very rarely. We report a case of a young female patient with spinal multiple tumors associated with bilateral vestibular schwannomas attribute features of NF2, underwent surgical treatment, whose histologic exam showed anaplastic ependymoma. Our objectif through this case is to demonstrate clinic and radiologic findings, and discuss about histologically features.

Keywords : *Spine, Anaplastic, Ependymomas, Neurofibromatosis*

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I. Introduction

Neurofibromatosis type 2 (NF2) is a rare autosomal dominant disorder, it was first described in the 1822 by Wishart, characterized by presence of bilateral vestibular schwannomas and current in about 90% to 95% of sufferers [1]. Ependymomas account for more than 75% of intramedullary spinal cord tumors related with neurofibromatosis type 2 [2] [3] [4]. On the other hand, Spinal anaplastic ependymomas appear to be tremendously rare, histologically the malignant subtype corresponds to WHO Grade III tumors. We report a case of a young female patient with spinal multiple tumors associated with bilateral vestibular schwannomas attribute features of NF2 and we describe clinic, radiologic and histologic findings.

II. Case Raport :

A 12 years old girl was admitted to our departement of neurosurgery due to the fact of headache and then after weakness in her lower extremities. She did not have any relative identified of NF2 in first degree, and no other family history responded to neurofibromatosis.

2.1 Neurologic exam :

Neurological examination showed spastic paraparesis with 2/5 muscle power in the lower limbs,bilateral hyperreflexia with ankle clonus, auditory symptoms such as hearing loss on the right side,Tinnitus and dizziness.

2.2 Systemic exam

In systemic exam, no café-au-lait spot or neurofibroma was seen on trunk and sphincter function was normal.

2.3 Imagery

Magnetic ressonance imaging (MRI of spine) : showed well-defined multiple spinal segments and displaying an exophytic growth pattern as well as frequent dissemination with masse compressing in cervicothoracic jonction, isointense T1 and hyperintense on T2, with contrast enhancement ‘string of pearls’ appearance along the spinal cord with intact bony structures.
Magnetic ressonance imaging (MRI of brain) : showed bilateral vestibular schwannomas more marked in right side

2.4 Hearing test : (audiometry and audiogram)

Right sensorineural hearing loss has been objectified

Genetic test was not done.
We planned staged operations depending on age, overall clinical neurogical status, and Imaging findings

2.5 Surgery procedure

Resection is realised, in prone position using a posterior midline approach a tow level posterior laminotomy D1-D2 is developed and Ligamentum flavum is removed all the way to the next bony segment cranially and caudally then opening the dura mater, Exploration shows a yellowish exophytic tumor reaches cord surface and infiltrating the spinal cord without a visible cleavage plane in depth , Internal prudent debulking are performed ,evoked potentials is not available, hemostasis has been obtained, The dura is closed with running sutures in a watertight manner and return of the bone to its place than the closure of the superficial planes.

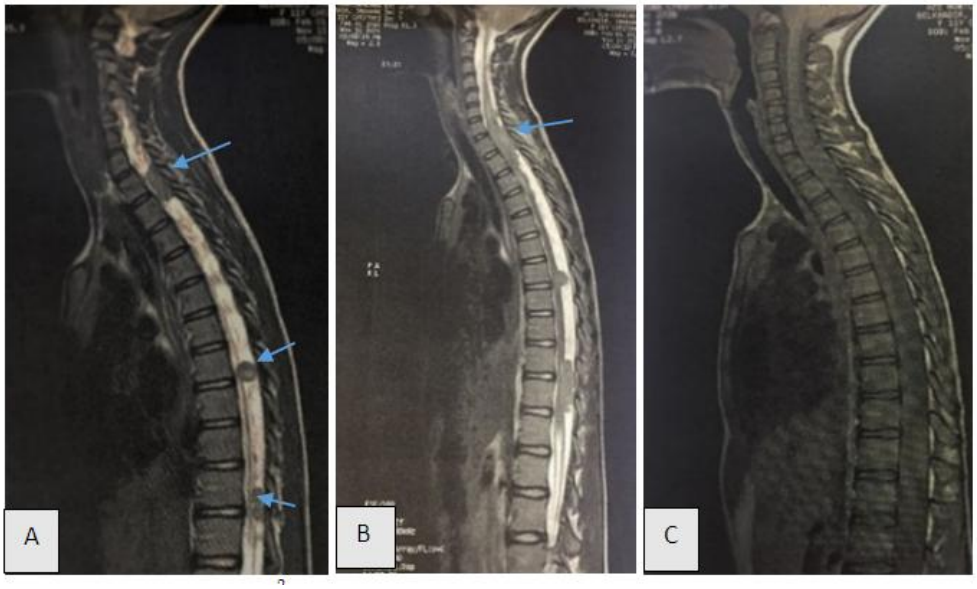


Fig. 1 : MRI Of spinal cord : (T2, STIR), T1 : showing multiple level lesions (A) with CMJ compression(B).

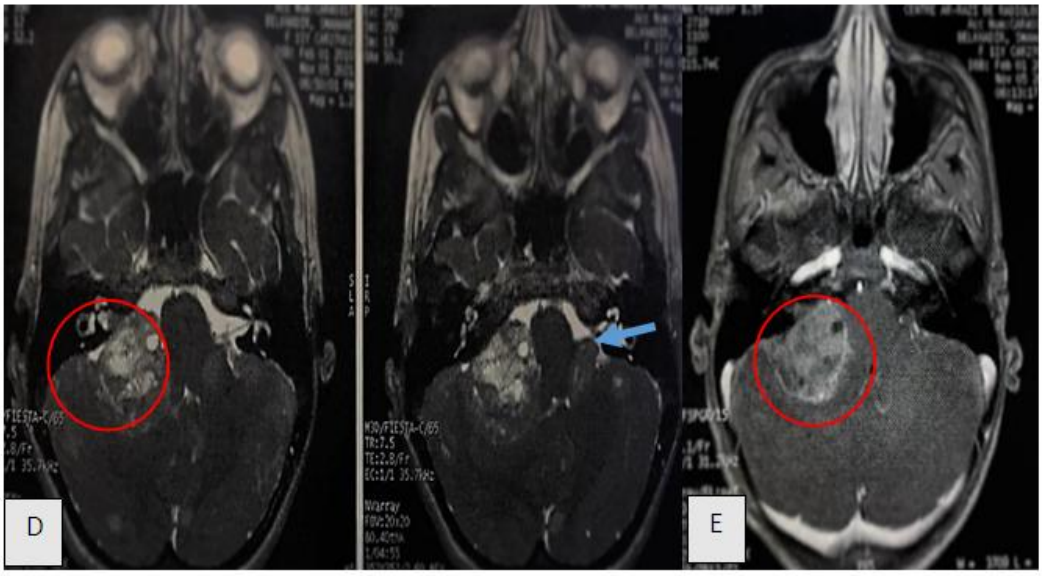


Fig. 2 : Brain MRI T2 (D) and T1(E) with Gadolinium enhancement vesticular schwannomas more marked on the right side

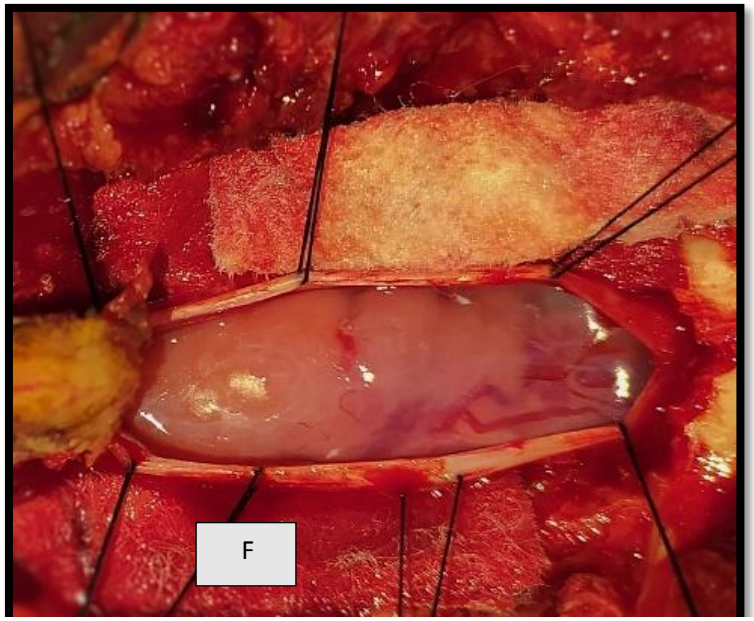
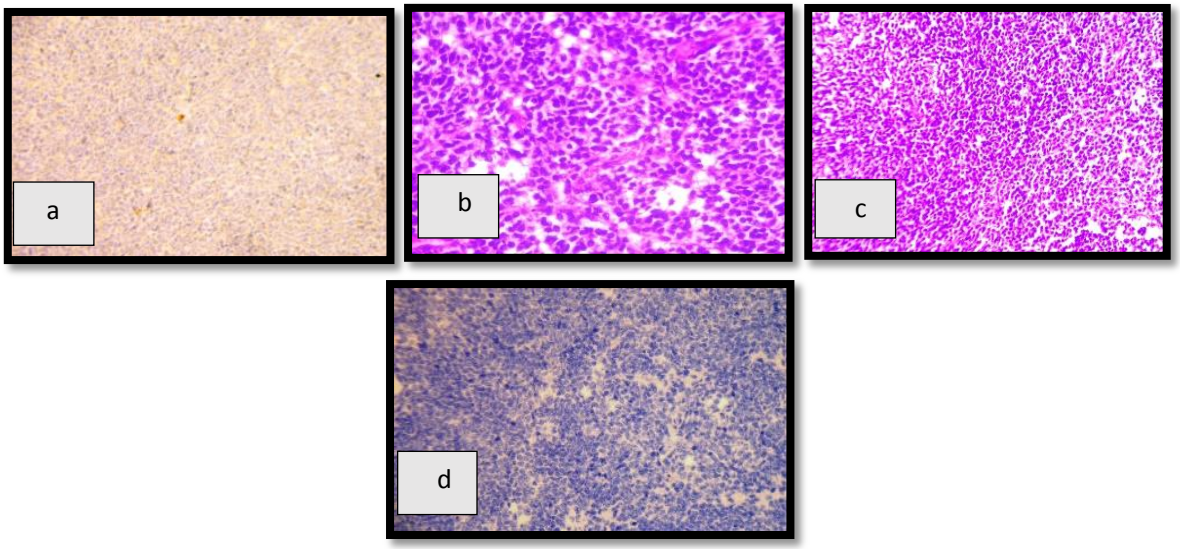


Fig. 3 : Intraoperative image schowing the exophytic lesion aspect(F)

2.6 Histologic results :

Histology confirmed the diagnosis of ependymoma classified by the WHO system as anaplastic grade III.



follow-up was Unfortunately marked by death due to respiratory dysfunction 3 days after the operation.

III. Discussion :

Neurofibromatosis type 2 (NF2) is a rare genetic autosomal dominant disorder with an incidence is about 1 in 33,000 to 40,000 individuals [5] Other nervous system tumors might also occur,including cranial nerve and spinal nerve root schwannomas, intracranial and spinal meningiomas, and intrinsic spinal cord tumor[6]Multiple spinal tumors have to be viewed as a hallmark of the disease. Cumran et al [7] documented a excessive frequency of spinal intramedullary tumors in a kindred of 23 family membre affected by using NF2. In a population-based find out about involving solely partial examination of the spine in symptomatic patients, spinal tumors were found in 26% of the patients [8]. In different study through Parry et aI [9] MR Imaging of the whole spine used to be carried out in forty of 49 patients with NF2 and published spinal tumors in 30 patients (75%).

it is necessary to distinguish neurofibromatosis 2 from neurofibromatosis 1, also called Von Recklinghausen's neurofibromatosis. it is also essential to differentiate between neurofibromatosis 2 and the sporadic unilateral acoustic neuroma. In neurofibromatosis 2, symptoms usually begin in the teens or early twenties [10]. MRI is useful for visualizing multiple intracranial and intraspinal tumors,as well as peripheral nerve tumors.[11].

Intramedullary tumors are more concerned by the surgycal management than extramedullary tumors. Brotchi et al demonstrated that the preoperative neurological status and the size of tumor are the main prognostic factors in patients with intramedullary tumor [12] . Aboukais et al, reported that in their three symptomatic patients operated on for intramedullary tumors, neurological recovery was only partial in two patients and absent in one [13].

In patients with NF2, intramedullary tumors are usually low-grade ependymomas and especially located in the lower cervical and upper thoracic spine [14]. Indeed, a rare subtype of spinal cord anaplastic ependymoma predominantly affecting young adults, and tumors are often large at diagnosis, involving multiple spinal segments and exhibit an exophytic growth pattern as well as frequent dissemination [15][16][17] .

Chemotherapy according to different protocols was often administered in children with incompletely resected or anaplastic (WHO grade III) tumors in addition to radiotherapy [18] [19] [20]. {Citation}

IV. Conclusion :

Spinal tumors at a hight variety and frequency, are a characteristic feature of NF2, Intramedullary tumors are usually low-grade ependymomas and especially those tumors located in the lower cervical and upper thoracic spine, However, spinal cord, anaplastic ependymomas appear to be tremendously rare. histologically corresponds to WHO Grade III, displaying an exophytic growth pattern tumors. Indeed, close follow up clinical and radiological investigation is more logical option for NF2 patients with asymptomatic multiple intraspinal tumors and to discuss treatement before the occurrence of neurogical aggravation but for patients with symptomatic tumors, with a compressive lesion on imagin resection may be warranted depending on age, overall clinical status, and lesion accessibility .

Conflict of interest statement
The authors declare no conflict of interest.

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