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Case Report

Bilateral Dumbbell C1-C2 Neurofibromas Leading To Cervical Cord Compression in a Patient with Neurofibromatosis Type 1: A Case Report

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ABSTRACT:

Cervical cord compression by cervical plexiform neurofibroma is one of the most morbid complication in a patient with Neurofibromatosis-1. Urgent spinal cord decompression and exeresis of the tumor should be undertaken in such scenarios. In this communication, we present a case of 30 year old male of NF-1 with quadriparesis and bilateral dumbbell cervical neurofibromas at the level of C1-C2, who underwent a total resection and improved dramatically post surgery. We describe how these tumors are different from tumors at other spinal sites. We briefly discuss various surgical approaches available to excise them and highlight the challenges faced during and after these procedures

Key Words: Postoperative wound infection, Staphylococcus aureus, Escherichia coli

INTRODUCTION

NF-1 is an autosomal dominant disorder, caused by biallelic inactivation of tumor suppressor gene NF1 located on chromose no.17.[1] The neurofibromas associated with NF-1 can be either focal cutaneous / subcutaneous or diffuse plexiform lesions. These plexiform lesions can arise from nerve roots of spine at multiple levels. The incidence of spinal tumors in NF -1 patients is around 40% as detected by MRI but they symptomatic in only 2% of the cases.[2] Cervical cord compression is one of the rarer but dreaded and debilitating presentation of such neurofibromas. Amongst cervical tumors, C1-C2 tumors are even rarer and have atypical features; hence, compel a distinctive management plan.

Case Report

A 30 year old male presented with chief complaints of progressive quadriparesis and increased frequency of falls since the past one month. He was diagnosed as a case of sporadic NF-1 at the age of 16. His general physical examination revealed scoliosis, cafe au lait macules, lisch nodules and multiple neurofibromas all over the body. MRI cervical spine was suggestive of multiple neurofibromas along exiting nerve roots from C1-C7, spinal cord narrowing at the C2 level of cervical spine due to bilateral dumbbell C1-C2 neurofibromas (Fig 1 and 2), neurofibromas in bilateral T1 and T7, right T5, T6, T8 neural foramina, paraspinal area, subcutaneous and cutaneous plane of neck, deep neck and axilla.

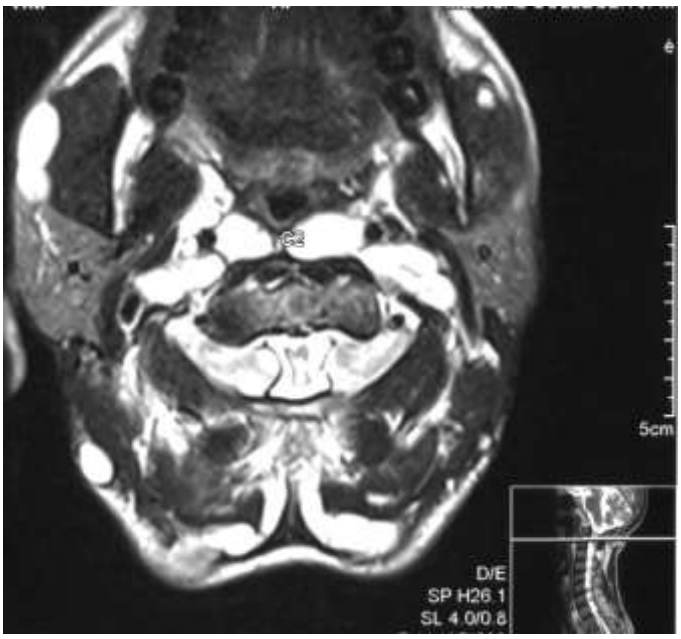

Fig 1: An axial T2W image showing bilateral dumbbell tumor compressing cord at level of C2.

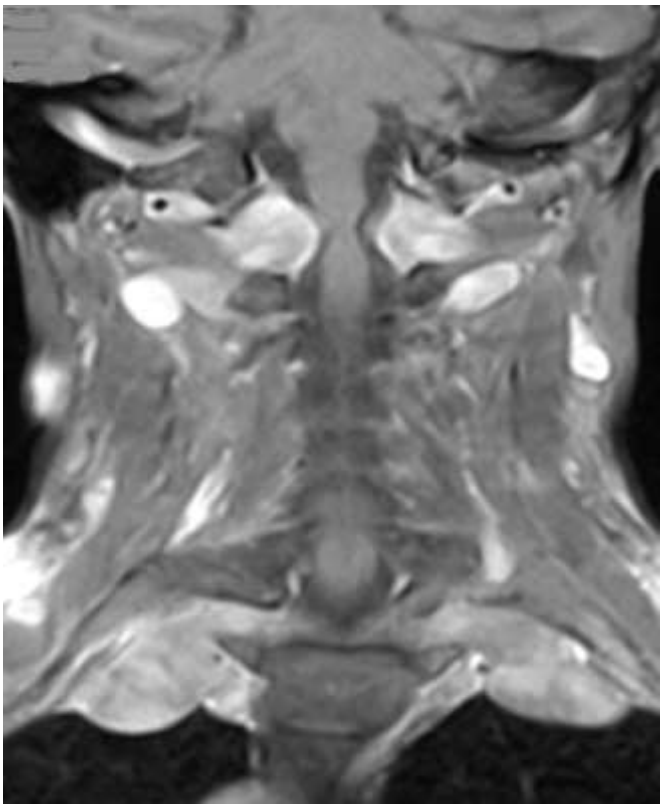

Fig 2: A coronal T1W image showing intra and extra dural neurofibroma compressing spinal cord.

A diagnosis of bilateral dumbbell C1-C2 neurofibroma was made and patient was taken for surgery. Posterior arch of C1 was removed for adequate spinal cord exposure. Two extradural large dumbbell shaped neurofibromas were found located laterally to the cord, arising from C1 nerve root and compressing cord in midline. Glistening white, encapsulated, moderately vascular tumors were removed piecemeal with CUSA and bipolar and sent for histopathology, which revealed benign neurofibromas (Fig 3). In post-operative period, patient improved substantially and was discharged one week later in ambulatory condition.

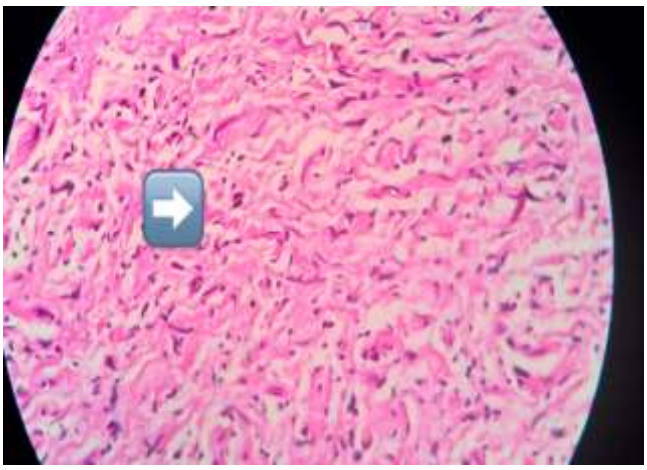

Fig 3: Microscopic section showing 'shredded carrot like appearance' of collagen in between neural elements. Neural cells are spindly with moderately eosinophillic cytoplasm, elongated nuclei with tapered ends and many buckled nuclei.

Discussion

Freidrich Von Recklinghausen described NF -1 first in 1882. It has an incidence of one in 2500-3000 births. NF1 gene encodes for a protein, neurofibromine which is a tumor suppressor, inactivation of this gene leads to tumorigenesis.[1] Pathognomonic features of NF-1 are cafe au lait spots and neurofibromas; but it has a wide clinical spectrum which includes optic gliomas, malignant peripheral nerve sheath tumors, brainstem gliomas, lisch nodules, scoliosis and pseudoarthrosis to name a few.[3]

Symptomatic cervical cord compression due to plexiform neurofibromas is an under-reported complication. It is characterised by progressive quadriplegia, hemiparesis, paraparesis, cervical neck pain, suboccipital headache, urinary disturbances and rarely Lhermitte's sign. Leonard et al conducted a study on 1500 patients with NF-1 and found cervical cord compression in only 13 patients mainly at the level of C2-C3.[4] Creange in 1999 conducted a study on 158 patients with NF-1 and found two patients to have cervical cord compression.[5] Sarica et al too dicussed a case where neurofibromas were present along the entire spinal cord and decompression was done at C3, C4, C5 and C6 levels.[6] In our study, the culprit tumor was present at C1-C2 level which is the least common site.

C1-C2 neurofibromas are unique as they have a tendency to expand in hourglass fashion. They acquire a dumbbell shape on encountering anatomical barriers like duramater, nerve root foramen and bony elements.[7] C1-C2 tumors have great anatomic peculiarity as well. The diameter of cervical cord at the level of C1 is 23 mm, 20 mm at C2 and 15 mm below C2. Roomy spinal canal at this level ensures that the tumor has achieved massive dimensions before it is symptomatic.[8] Surgical excision of these tumors raises several challenges like adequate spinal exposure, preservation of involved spinal nerve roots, relationship with vertebral artery, post-operative kyphosis, cervical instability, risk of general anesthesia and high recurrence rates.[9]

Surgical approaches to C1-C2 tumor include the posterior, postero-lateral, anterior, antero-lateral and extreme lateral approaches. Posterior approach includes standard bilateral laminectomy with or without suboccipital craniectomy. It is an easy and straightforward approach with least risk of instability but inadequate ingress to anteriorly placed tumors. Postero-lateral approach is a standard posterior approach followed by medial ipsilateral partial facetectomy. Anterior approach is a transoral approach with good access to tumor but high risk of infection. Antero-lateral approach partially removes the anterior portion of vertebral body and transverse foramen but preserves the facet. Lastly the extreme lateral transcondylar approach provides benefit of removal of both intra and extradural part of the tumor, but obviously bilateral tumors cannot be operated in the same sitting.[9,10] We went ahead with posterior arch removal to decompress foramen magnum effectively, as just the laminectomy was not adequate.

Another important consideration in cervical tumors is their relationship with vertebral artery. This artery is usually displaced anteromedially by the tumor and is always separated from the tumor by a thin layer of periosteum and perivertebral veins. Therefore if dissection is carried out in this plane, there are limited chances of ischemic injury.[11]

In a patient of neurofibromatosis the risk of post-laminectomy kyphosis is higher due to pathological defects in bone growth and development.[12] Hence fixation should be carried out wherever stability seems compromised. Due to these challenges sometimes complete resection of tumor is not possible. This is a major risk factor for high reccurence rates seen with these cases, which are generally to the tune of 10.7% at 5 years and 28.2% at 10-15 years.[13].

Conclusion

Tumors arising from C1 and C2 nerve roots are different from tumors found at other spinal sites. Separate and streamlined protocols should be developed for the management of these unique entities. We believe screening MRIs in cases of NF-1 might be helpful in detecting these time bombs early, although it has been repeatedly emphasised in literature[4,5] that screening MRI for spinal tumors is not necessary in the patients of NF-1 due to low incidence of tumors that are symptomatic and require surgical excision.

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