CERVICAL TRIDENT SHAPED NEUROFIBROMA A RARE VARIANT

ABSTRACT

BACKGROUND

Spinal nerve root tumors can arise throughout the spine and at multiple levels, likely representing plexiform neurofibromas that grow from the nerve root into the intraspinal space either intradurally or epidurally and exit through the neural foramen, producing a dumbbell appearance. Although cases of extramedullary neurofibromas of dumbbell shape in cervical spine have been reported, to the best of our knowledge, there are no reports of extramedullary neurofibromas of trident shape.

METHODS

A 26-year-old woman was presented with swelling over right side of the neck. Diagnostic work-up included magnetic resonance imaging (MRI), contrast enhanced computed tomography (CECT) Neck that revealed an intradural, extramedullary tumor mass at the right C2– C6 level with extraspinal extension.

RESULTS

Spinal cord compression or canal compromise is the most reliable indication for surgery. The solitary cervical neurofibroma was treated surgically in a single stage through laminoplasty and excision of the intradural tumor along with that of the neck component and done without any complications.

CONCLUSION

A single stage double approach was adopted in this case and after total excision the shape found was not that of a typical dumbbell but rather appeared as that of a trident. Hence, here we would like to suggest a new nomenclature for this neurofibroma as Trident neurofibroma.

KEY WORDS

Spinal tumors, extramedullary, neurofibroma, dumbbell, laminoplasty

Introduction

Spinal nerve root neurofibromas can arise throughout the spine and at multiple levels, and likely represent plexiform tumours based on the involvement of multiple nerve fascicles.[1] Cervical spinal neurofibromas are benign, well-defined, encapsulated tumor, usually arising from the nerve roots. Neurofibromas may present either as solitary lesions or as part of a syndrome, i.e. neurofibromatosis or von Recklinghausen's disease of the skin (NF-1).[2,3]The aetiology behind solitary neurofibroma is still unknown. Marocchio et al.,[3] considered solitary neurofibroma to be hyperplastic hamartomatous malformations rather than neoplastic. Anatomically, neck neurofibromas are relatively rare, with slow painless growth. Neurofibromas are usually treated by excision, especially when they are symptomatic or disfiguring.[2] Based on their location, these tumors can take the form of a dumbbell-shaped mass. The dumbbell neurofibromas (NF) are classified into nine types on the basis of extraforaminal extension.[4,5]. The Type I (within spinal canal) and Type IIa (extension up to intervertebral foramen) NF are usually approached through conventional hemi-laminectomy or open-door laminoplasty.[5] The Type IIb-c possesses a surgical challenge because majority of tumor lies in the extraforaminal region. Type III tumors are those which have both dural and foraminal constriction. The anatomical knowledge of posterolateral neck triangle and surgical corridors is needed for maximal safe excision.[4] The variability in terms of location, extent, and size of a dumbbell cervical spinal neurofibroma (NF) possesses unique technical challenges for the surgeon.[4] The attachment of the extramedullary part to the surrounding tissues makes dumbbell tumors much important. Surgical expertise is offered over adjuvant radiotherapy as it’ s a type of benign disease. In this article, we present a study of type IIIb tumor, an intradural and extradural-paravertebral type and that with a different shape.

Case Presentation

A 26-year-old woman was presented at our Department of Neurosurgery, Guwahati Neurological Research Centre (GNRC), Dispur, Guwahati, Assam with a history of swelling over the right side of the neck (Fig 1A), gradually progressing in size but with no pain, tingling and numbness or weakness of limbs nor any bladder and bowel disturbance. On presentation patient had no deficit. The CT scan of her neck showed a bilobed soft tissue density lesion. It was noted in the right paravertebral region of the neck at C3-C4 disc level causing widening of the right C3 neural foraminal involving C4 exiting nerve root. The lesion was seen abutting and displacing the spinal cord laterally at C3-C4 level. The lesion showed mild central enhancement on post contrast study feature suggesting nerve sheath tumor likely Neurofibroma involving right C4 nerve root. Magnetic resonance imaging (MRI) revealed clearly widening of the right C3-C4 neural foramen (Fig1B). A T2 mildly hyperintense and T1 isointense well defined lesion was seen in the middle third of the neck on the right lateral aspect deep to the sternocleidomastoid muscle extending through the widened neural foramen (Fig 1C). There was an intraspinal component of the lesion measuring about 10.5 cm x1.7cm (Fig 1F) effacing the right lateral thecal sac. There was compression and displacement of the cord to the left. The component of the lesion in neck measured approx 4.1 cm x2.5 cm. Post contrast scan revealed moderate enhancement of the lesion. There was compression of the right internal jugular vein (IJV). Features were suggestive of neurofibroma arising from the right C4 existing nerve root with both intra and extra spinal components.

TREATMENT

She was explained in details regarding the procedure which consisted of twin approach to the tumor and was operated under general anaesthesia. The operation procedure adopted was C2-C6 laminoplasty and excision of intradural trident tumour along with excision of neck component. The neck component was excised first followed by the intradural part. The specimen after total excision did not appear as a dumbbell shape as the intradural component was significantly long extending from C2 to C6 level, rather it looked like a trident (Fig.1F). Since such a shape of neurofibroma is not reported and described in literature to the best of our knowledge, we would like to address such neurofibromas as TRIDENT Neurofibroma. The resected specimen was sent for histopathological examinations. The histopathology examination of the tumor on hematoxylin and eosin (H&E) staining demonstrated spindle cells with a loose myxoid background (Fig.1G). The tumor cells had spindle-shaped nuclei, and some of them were

curved. Deposition of collagen bundles in the stroma between the spindle tumor cells were also seen. The results thus confirmed the diagnosis of neurofibroma. Post-operatively patient was mobilized the next day and did not have any motor or sensory deficit with normal power in all the four limbs. The patient made an uneventful recovery and was discharged on fourth postoperative day. At the follow-up neurological examinations she showed normal motor power and sensations. She showed no deficit and was afebrile, continent, ambulatory.