Craniovertebral junction cord compression due to neurofibroma

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Accepted 2 June 2018

DESCRIPTION

A 26-year-old man with premorbid cutaneous neurofibromatosis presented with history of insidious onset, slowly progressive, bilaterally asymmetrical quadriparesis along with incontinence of urine since 4 weeks. Simultaneous to the onset of motor symptoms, he also complained of paraesthesias in all four limbs. On examination, multiple cutaneous neurofibromas were noticed all over his trunk, abdomen and all the limbs (figure 1). The upper and lower limbs were spastic. Motor power as per Medical Research Council grading was 3/5 in all four limbs. The deep tendon reflexes were brisk. The plantar response on both sides was extensor. The sensory level was clinically around C3/C4 dermatome.

In view of the premorbid neurofibromata, a possibility of compressive cervical myelopathy was thought of. MRI of cervical spine T2-weighted sagittal view and axial view showed a 'dumbbell' shaped extramedullary, extradural lesion compressing the cervical spinal cord at the craniovertebral junction (figures 2 and 3, respectively). The patient was operated and the lesion was confirmed to be a neurofibroma. The patient did not have other clinical features such as axillary freckles, optic nerve glioma, more than five cafe au lait spots, iris hamartomas and sphenoid dysplasia. However, there was a strong family history. The patient's father had similar lesions on his body.

Hence, the patient fulfilled the criteria of diagnosing neurofibromatosis type 1.

Neurofibromatosis type 1 is an autosomal dominant disorder which may give rise to benign or malignant tumours in affected individuals. Neurofibroma is a benign tumour arising from the endoneurium. They may grow into focal subcutaneous or cutaneous tumours. Seldom, they may manifest as diffuse plexiform neurofibromas that may involve multiple nerve fascicles or plexuses.[1]

The neurofibroma compressing the cervical cord, especially the craniovertebral junction, is rare. There is no age predilection for cervical cord compression due to neurofibroma. C2 nerve roots are most commonly affected. The reason for affinity of neurofibroma for C2 nerve root has been postulated to the repeated low-grade trauma as it

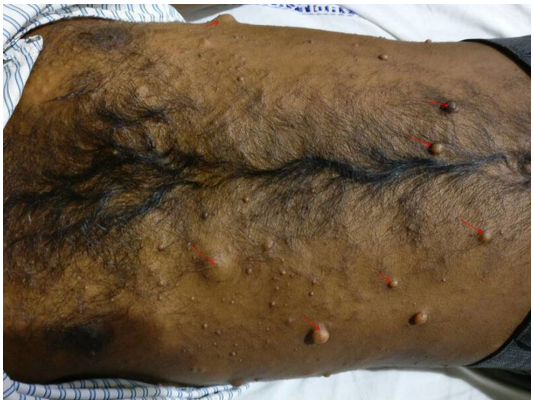

Figure 1 Multiple cutaneous neurofibromas of varying sizes on the lower chest and abdomen (red arrows).

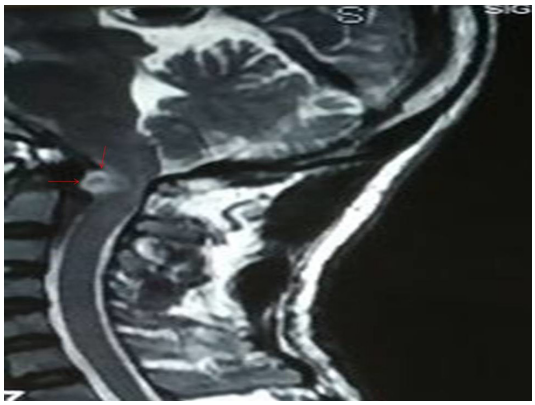

Figure 2 MRI T2-weighted sagittal sequence showing 'dumbbell shaped' lesion (red arrow) compressing the upper cervical cord (yellow arrow) at the craniovertebral junction.

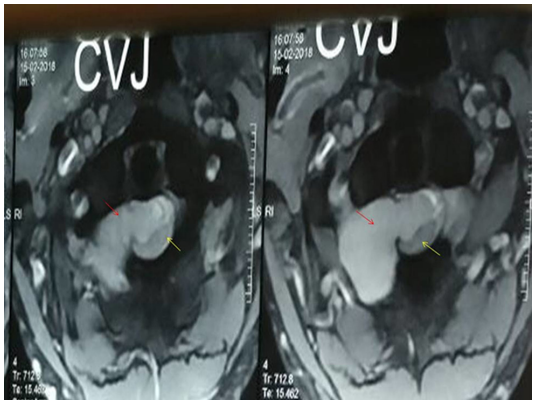

Figure 3 MRI T2-weighted axial view showing 'dumbbell shaped' lesion (red arrow) compressing the upper cervical cord (yellow arrow) at the craniovertebral junction.

To cite: Prasad O, Bompelli N, Reddy R, *et al*. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2018-225286

exits the foramen and winds over the superior aspect of lamina of the C2 vertebra.

Asymptomatic patients with neurofibromatosis type 1 do not require any radiological imaging. Patients developing progressive weakness of all four limbs due to cord compression require urgent neurosurgical intervention. Cord decompression at the level of compression is the preferred neurosurgery procedure.[2]

Learning points

- Neurofibromas are common benign tumours which may manifest as Cranio-vertebral junction anamoly in the form of cord compression rarely.
- C2 region is the most common area involved and is postulated to repeated low grade trauma to C2 nerve root as it winds over the superior aspect of lamina of C2 vertebrae.

Acknowledgements We acknowledge the contribution of our teachers and support of our family members in helping us make this manuscript possible.

Contributors OP and NB made substantial contributions to the conception and design of the work, acquisition, analysis and interpretation of the data. PR and AD were involved in drafting the work and revising it critically for important intellectual content. AD approved the final version to be published.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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