

Cervical myeloradiculopathy with Hypoglossal Schwannoma mimicking Amyotrophic lateral sclerosis: a Case report

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Case Report

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Abstract

Background: A presentation of upper motor and lower motor clinical signs without sensory impairment may suggest Amyotrophic lateral sclerosis, especially when it involves limbs and bulbar regions. However, co-existence of two or more unrelated pathologies can give rise to a clinical picture similar to Amyotrophic lateral sclerosis.

Case presentation: A 45 year old lady presented with slowly progressive right upper limb weakness for 6 months and mild left upper limb weakness for 2 months associated with muscle twitching. Her family members have noticed a change in character of her speech for few weeks. She couldn't recall a radicular pain at any given time. On examination, her right deltoid was wasted with numerous fasciculations over right proximal muscles. Right shoulder movements were weak. Her left deltoid was slightly wasted and fasciculations were appreciated. Her both biceps and supinator jerks were normal with exaggerated triceps jerks and positive Hoffman sign. Sensory and proprioception examinations were unremarkable. Both lower limbs were hypertonic with exaggerated knee and ankle jerks. Ankle clonus was absent and plantar responses were equivocal. Her tongue was deviated to right side with fasciculations and wasting of right side. Palatal movements and Jaw jerk were normal. She was clinically diagnosed as possible amyotrophic lateral sclerosis. Electromyography showed denervation changes in deltoids and biceps with right predominance and right genioglossus muscle too showed denervation changes. Magnetic resonant imaging of brain stem was undertaken especially because her tongue wasting and fasciculations were unilateral and it showed a right sided hypoglossal schwannoma. Magnetic resonant imaging of cervical spine revealed degenerative disk disease with mild cord compression and exit foramina narrowing at multiple levels.

Conclusions: Pure motor symptoms with a mixture of upper motor and lower motor signs may suggest a clinical diagnosis of Amyotrophic lateral sclerosis. However, when there are rare manifestations of a relatively common disease, such as unilateral tongue involvement as in the above scenario, a higher degree of clinical suspicion is needed to think of a dual pathology.

Keywords: Hypoglossal schwannoma, cervical myeloradiculopathy, Amyotrophic lateral sclerosis, dual pathology.

Background

A presentation of upper motor and lower motor clinical signs without sensory impairment may suggest Amyotrophic lateral sclerosis (ALS), especially when it involves limbs and bulbar regions [1]. Gradual progression of symptoms and signs involving several regions is generally observed in ALS [2]. Clinically heterogenous initial presentation of pure upper motor, pure lower motor and a mixture of upper motor and lower motor signs involving bulbar, cervical, thoracic, or lumbosacral body segments are seen in anterior horn cell disease and later may progress to ALS or other spectrum of motor neuron disease. [3]

ALS is a differential diagnosis for cervical myelopathy. [4] Degenerative disc disease of cervical spine should be considered in a patient who carries heavy loads on shoulders, leading to prolonged extension, flexion and vigorous bending of neck. [5] However, if the patient has bulbar involvement, it cannot be explained from degenerative disc disease of cervical spine alone, unless it has involved the upper cervical region. [6] Even though cervical myeloradiculopathy due to Degenerative disc disease is not an uncommon encounter in a neurology clinic, isolated hypoglossal nerve palsy is rare. [7]

We report a case of a rare isolated hypoglossal nerve palsy due to a hypoglossal schwannoma co-existing with cervical myeloradiculopathy due to occupation related degenerative disc disease, clinically mimicking ALS.

Case Presentation

A 45 year old previously healthy Sinhalese lady, a mother of 4 children from a southern rural village of Sri Lanka, presented to Teaching Hospital Karapitiya, Sri Lanka, with slowly progressive right upper limb weakness for 6 months associated with muscle twitching and mild left upper limb weakness for 2 months.

She is a tea plucker who carries tea leaves on her shoulders and works long shifts. Six months back she has noticed a weakness in her right arm needing more effort to continue her job which required repeated shoulder abductions. Despite the weakness she continued to work. Few weeks later she noticed a wasting of the right shoulder and experienced occasional muscle twitching.

Four months later, she noticed a weakness of her left arm together with difficulty in walking due to stiffness of the lower limbs. She couldn't recall a radicular pain involving cervical or lumbosacral regions at any given time. She never complained of muscle twitching in her lower limbs.

Her family members have noticed a change in character of her speech for a few weeks, even though she did not notice any abnormality on her own. She did not have dysphagia, nasal regurgitation or emotional lability. She attained menopause 3 years back and was not on hormone replacement therapy. There was no history to suggest a present or a past malignancy.

On general examination, her body mass index was 25.5 Kg/m^2 and oriented in time, place and person. She was afebrile. On cranial nerve examination, her speech was slurred and tongue was deviated to right side with mild wasting of right side of the tongue with fasciculations on the same side (figure 1). Her palatal movements and gag reflexes were normal with normal jaw jerk. Her cerebellar functions and other cranial nerves examination were normal.

On upper limb examination, her right deltoid was wasted with numerous fasciculations over proximal muscles. Her left deltoid was slightly wasted and fasciculations were appreciated over the deltoid. Tone was normal in both upper limbs. Right shoulder movements, elbow flexion and extension were weak. Left shoulder abduction was weak. Small muscles power and function was normal. Both biceps and

supinator jerks were normal with exaggerated triceps jerks and positive Hoffman sign bilaterally. Sensory and proprioception examinations were unremarkable. Her neck movements were full and painless.

Both lower limbs were hypertonic with exaggerated knee and ankle jerks. Ankle clonus was absent and plantar responses were equivocal. Wasting and fasciculations were not seen in lower limbs.

Pure motor involvement in multiple body segments and preserved reflexes in wasted muscles with a combination of upper motor and lower motor signs without sensory involvement pointed us to a clinical diagnosis of anterior horn cell disease.

Electromyography showed denervation changes on deltoids, biceps and triceps with right predominance. Right genioglossus showed denervation changes as well while the left genioglossus was normal.

Magnetic resonant imaging (MRI) of brain stem was undertaken, because her tongue wasting and fasciculations were unilateral. MRI revealed an avidly enhancing mass lesion at right hypoglossal canal measuring 1.1 cm × 1.5 cm × 1.6 cm with mild compression of right proximal jugular vein. The lesion showed T1 hypointensitiy and T2 and Fluid-attenuated inversion recovery (FLAIR) hyperintensity. A right hypoglossal schwannoma was diagnosed. (Figure 2, Figure 3, Figure 4)

X-ray of cervical spine revealed straight cervical spine and degenerative changes from C3 to C7 vertebrae. (Figure 5) MRI of cervical spine revealed modic type 2 changes at C4-C5 and C6-C7 end plates, disc bulging with indentation of thecal sac without cord compression at C3-C4 level, disc osteophyte complex causing indentation of thecal sac and mild cord compression at C4-C5 level, bilateral exit foramen narrowing due to osteophytes causing compression of both C5 roots and diffuse disc bulging with indentation of thecal sac without cord compression at C5-C6 and C6-C7 level with bilateral exit foramina narrowing. Cervical myeloradiculopathy possibly due to occupation related degenerative disc disease was the overall impression. (Figure 6)

Excision of hypoglossal schwannoma was planned and advised to avoid carrying heavy loads in the future. Medical recommendation was given to change her occupation to light work from heavy work and currently her degenerative disc disease of cervical spine is being followed up.

Discussion And Conclusions

Pure motor involvement of limbs are seen myopathies, neuromuscular junction disorders, Motor neuronopathies/neuropathies including Guillain-Barre variants and anterior horn cell pathologies and motor strokes. [8, 9, 10, 11] Pure motor cranial nerves include Trochlear nerve, Abducens nerve, Accessory nerve and Hypoglossal nerve. [12] When pure motor cranial nerves involve in isolation or in combination, the clinical picture appears as a pure motor pathology, even though it may not be the actual scenario. When there is involvement of both cranial nerves and peripheral nerves without sensory signs or symptoms, the treating physician subconsciously try to fit the clinical findings to a unifying diagnosis via pattern recognition. [13]

In this case scenario, patient has mixed upper and lower motor signs with multiple body segment involvement suggesting ALS. However unilateral hypoglossal nerve involvement with only lower motor signs made our clinical diagnosis of ALS doubtful.

Mixed upper motor and lower motor signs in upper limbs with pure upper motor signs in lower limbs together with the fact that she is a manual laborer supported the possibility of cervical myeloradiculopathy due to occupation related degenerative disc disease of cervical spine. Hypoglossal nerve palsy due to upper cervical degenerative disease has been reported, which further supports the above diagnosis. [14] However as hypoglossal nerve palsy secondary to degenerative cervical spine is rare, we sought another explanation.

Hypoglossal nerve has long course. Segmental approach of hypoglossal nerve lesion helps to identify the pathology. Hypoglossal nerve palsy can be supranuclear, nuclear or infranuclear.[15] Infranuclear hypoglossal nerve has five segments, namely medullary segment, cisternal segment, skull base segment, carotid space segment, sublingual segment. [16] Our patient had an infranuclear, skull base segment lesion. Nerve sheath tumors such as schwannomas are a cause for hypoglossal nerve palsies involving skull base segment.[15] Schwannomas are slow growing and benign neoplasms of schwann cells. Ninety percent of schwannomas are acoustic neuromas.[17] Schwannomas are usually solitary, unless it is associated with neurofibromatosis or carney complex.[18,19] However our patient did not have features to support such syndromes.

Hypoglossal schwannomas are classified to three types. Type A schwannomas are primarily intracranial with small extension into the bone. When the tumor mass is located mainly within the bone, irrespective of intracranial extension, it is considered Type B. Type C is primarily extracranial and may show minor extension into bone.[20] Our patient's schwannoma was detected at hypoglossal canal.

Surgery is the mode of treatment for hypoglossal schwannoma. Generally, postsurgical recovery is good. Residual deficits and adjacent structural damage has been documented post surgically according to the tumor size, location and complexity of surgery. [21]

This case report is an eye opener. Pattern recognition helps to come to a unifying diagnosis, but when there are rare manifestations of a relatively common disease, such as unilateral tongue involvement in suspected amyotrophic lateral sclerosis as in our patient, the clinician should look for double pathology.

Abbreviations

ALS: Amyotrophic Lateral Sclerosis

MRI: Magnetic Resonant Imaging

FLAIR: Fluid-attenuated inversion recovery

Declarations

Ethics approval and consent to participate

Ethics approval was not taken as not applicable.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

SBA, DNW, AD and SS were involved with investigating this case. SBA was the main author responsible for the collection of data and writing up of the manuscript. DNW, AD and SS read and corrected the manuscript. All four authors read and approved the final manuscript.

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Not applicable.

Competing interests

The authors declare that they have no competing interests.

Consent for publication

Consent for publication was obtained from the patient and from the Director of Teaching Hospital Karapitiya, Sri Lanka

References

1. Brooks B. El escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*. 1994;124:96-107.
2. Brown R, Al-Chalabi A. Amyotrophic Lateral Sclerosis. *New England Journal of Medicine*. 2017;377(2):162-172.
3. Elman L, McCluskey L. UpToDate [Internet]. Uptodate.com. 2018. Available from: <https://www.uptodate.com/contents/clinical-features-ofamyotrophic-lateral-sclerosis-and-other-forms-of-motor-neuron-disease>.
4. Dvorak J, Sutter M, Herdmann J. Cervical myelopathy: clinical and neurophysiological evaluation. *European Spine Journal*. 2003;12(0):S181-S187.
5. Jäger H, Gordon-Harris L, Mehring U, Goetz G, Mathias K. Degenerative change in the cervical spine and load-carrying on the head. *Skeletal Radiology*. 1997;26(8):475-481.
6. Blankenship L, Basford J, Strommen J, Andersen R. Hypoglossal nerve palsy from cervical spine involvement in rheumatoid arthritis: 3 case reports. *Archives of Physical Medicine and Rehabilitation*. 2002;83(2):269-272.
7. Patro S, Torres C, Riascos R. An Unusual Case of Isolated Hypoglossal Nerve Palsy Secondary to Osteophytic Projection from the Atlanto-Occipital Joint. *The Neuroradiology Journal*. 2014;27(3):361-364.
8. Garg N, Park S, Vucic S, Yiannikas C, Spies J, Howells J et al. Differentiating lower motor neuron syndromes. *Journal of Neurology, Neurosurgery & Psychiatry*. 2016;88(6):474-483.
9. Liepert J, Restemeyer C, Kucinski T, Zittel S, Weiller C. Motor Strokes. *Stroke*. 2005;36(12):2648-2648.
10. Hill M. THE NEUROMUSCULAR JUNCTION DISORDERS. *Journal of Neurology, Neurosurgery & Psychiatry*. 2003;74(90002):32ii-37.
11. Russell J. General Approach to Peripheral Nerve Disorders. *CONTINUUM: Lifelong Learning in Neurology*. 2017;23(5):1241-1262.
12. Clarke C, Howard R, Rossor M, Shorvon S. *Neurology A Queen Square Textbook*. 2nd ed. John Wiley & Sons, Ltd; 2016.
13. Rutkove S. Pattern recognition. *Neurology*. 2003;61(4):585-586.
14. Weindling S, Goff R, Wood C, DeLone D, Hoxworth J. Is Hypoglossal Nerve Palsy Caused by Craniocervical Junction Degenerative Disease an Underrecognized Entity?. *American Journal of Neuroradiology*. 2016;37(11):2138-2143.
15. Chen Y, Dai M, Ho C, Huang T. Isolated Hypoglossal Nerve Paralysis. *The American Journal of Medicine*. 2014;127(10):926-927.
16. Thompson E, Smoker W. Hypoglossal nerve palsy: a segmental approach. *RadioGraphics*. 1994;14(5):939-958.
17. Rachinger J, Fellner F, Trenkler J. Dumbbell-shaped hypoglossal schwannoma. A case report. *Magnetic Resonance Imaging*. 2003;21(2):155-158.

18. Shrikrishna B, Jyothi A, Kulkarni N, Mazhar M. Extracranial Head and Neck Schwannomas: Our Experience. Indian Journal of Otolaryngology and Head & Neck Surgery. 2015;68(2):241-247.
19. Borkar S, Kamath S, Kashyap N, Sagar S, Rao L, Warrier R et al. Carney Complex: case report and review. Journal of Cardiothoracic Surgery. 2011;6(1).
20. Kaye A, Hahn J, Kinney S, Hardy R, Bay J. Jugular foramen schwannomas. Journal of Neurosurgery. 1984;:1045-1053.
21. Fornaro R, Salerno A, Filip D, Caratto E, Caratto M, Casaccia M. Schwannoma of the hypoglossal nerve: Review of the literature based on an illustrative case. Molecular and Clinical Oncology. 2017.

Figures



Figure 1

The tongue is deviated to right side. Atrophy of right genioglossus is demonstrated.

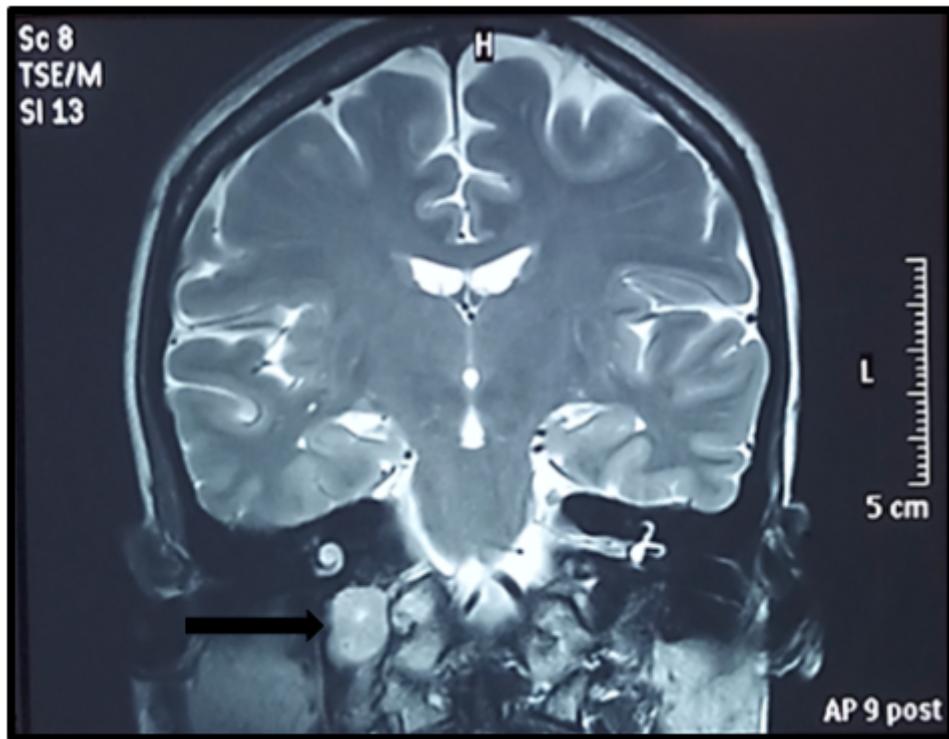


Figure 2

Coronal section of T 2 weighted MRI of brain shows right sided hypoglossal schwannoma. (Arrow)

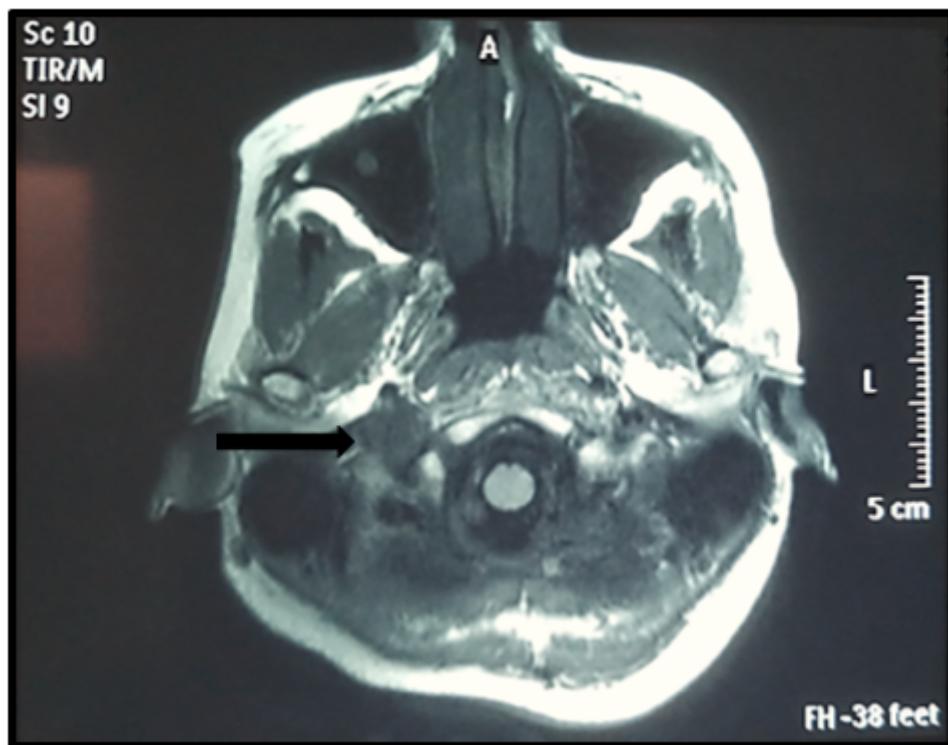


Figure 3

Transverse section of T 1 weighted MRI of brain shows hypointense lesion. (Arrow)

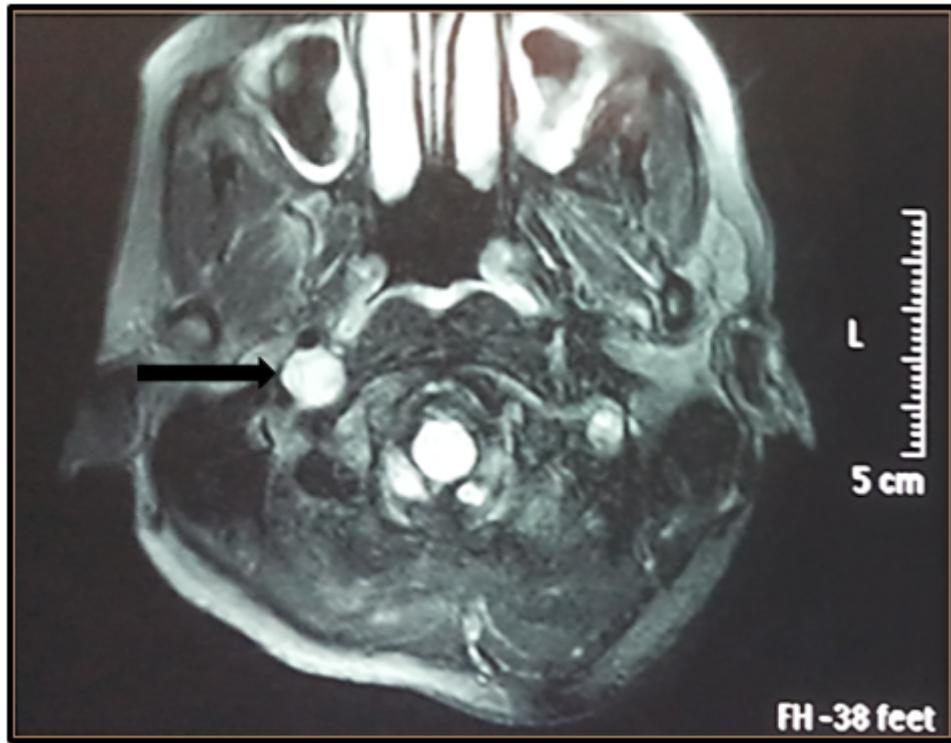


Figure 4

Transverse section of Fluid-attenuated inversion recovery (FLAIR) MRI brain shows well demarcated hyperintense lesion of right Hypoglossal nerve. (Arrow)

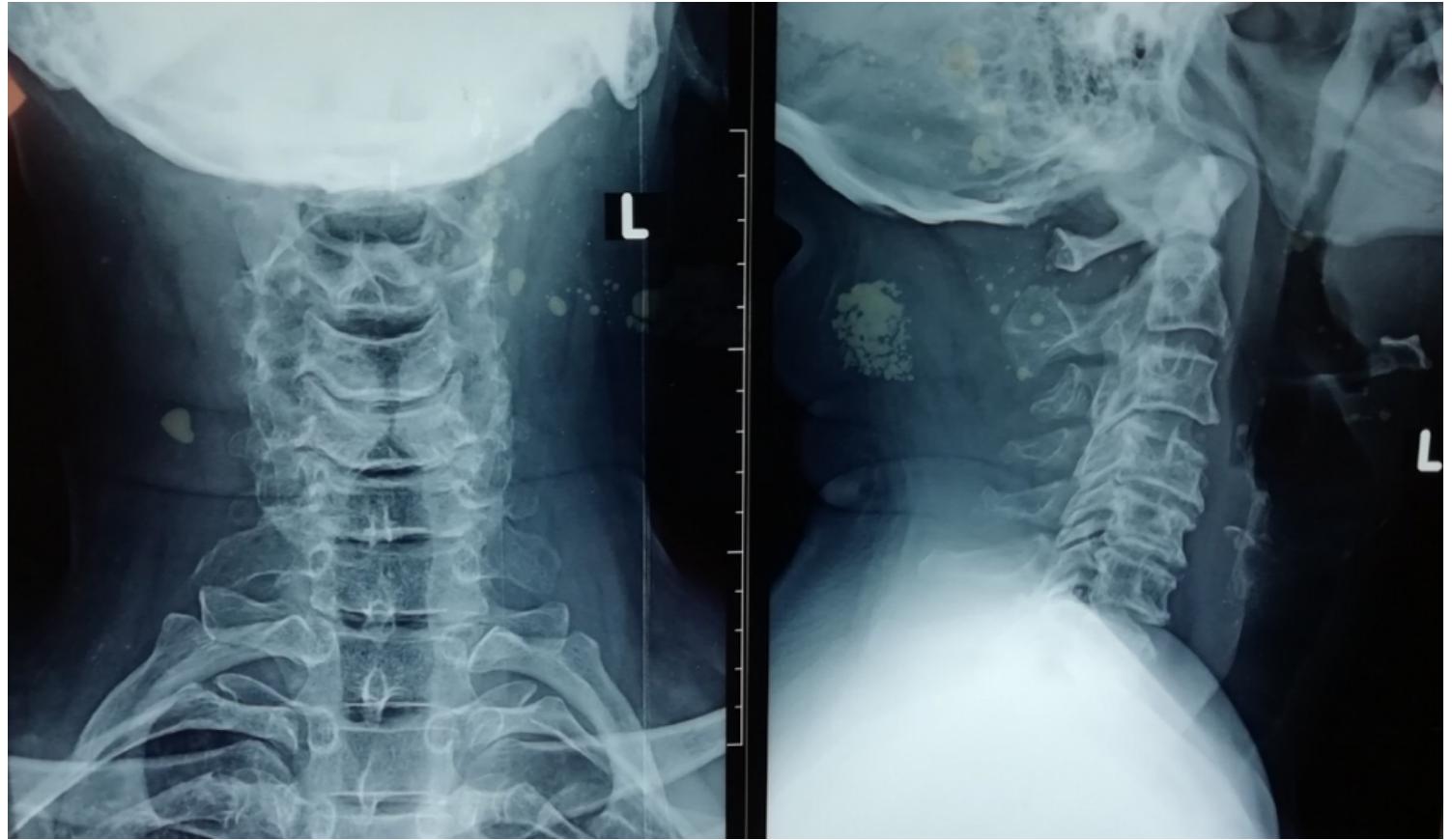


Figure 5

Anteroposterior and lateral views of X-ray cervical spine demonstrating straight cervical spine with degenerative disc disease.



Figure 6

MRI of cervical spine demonstrating thecal indentation at C3-C4, C4-C5, C5-C6 and C7-C8 levels with disc osteophyte complex causing mild cord compression at C4-C5 level due to degenerative disc disease of cervical spine.

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