

Letter to Editor

Dumbbell-shaped neurofibroma of the upper thoracic spine: A case report

Dear Editor,

Dumbbell-shaped thoracic neurofibroma represents a distinct type of tumor and can involve both the spinal canal and the posterior thoracic cavity. Dumbbell tumors are those with an intraspinal and paraspinal component, connected through a frequently enlarged and eroded intervertebral foramen. Most dumbbell tumors are located in the thoracic spine. The extraspinal tumor extension is usually larger than the intraspinal tumor part, but the intraspinal tumor component commonly causes the typical symptoms of local pain and symptoms from spinal cord compression in the thoracic spine. The differential diagnosis of dumbbell shaped foraminal masses include neurogenic tumors (schwannoma, neurofibroma) and meningiomas. Asymptomatic cases require no further treatment, but symptomatic cases justify surgical treatment in the form of total removal of neurofibroma. Posterior laminectomy with unilateral facetectomy allows single-stage resection of dumbbell neurofibromas with significant intraspinal and paraspinal components. Here, we report the case of 34-year-old female patient with dumbbell neurofibroma in the upper thoracic region.

A 34-year-old female was admitted to the hospital with complaints of pain over left scapular region and tingling over both lower limbs since 2 years and progressive weakness of both lower limbs for 3 months. She was bedridden for the past 30 days. Clinical examination did not show any neurocutaneous markers. Neurological examination revealed spastic paraparesis, more pronounced distally, hypoesthesia below D3 dermatome and hyperactive deep tendon reflexes.

T1-weighted magnetic resonance imaging (MRI) of the dorsal spine showed isointense signal and hyperintense signal on T2-weighted images, with patchy necrotic area. The intradural component was located in the left lateral thecal sac at D3 level, extending from D2-3 to D3-4 level with large extraforaminal component in the posterior mediastinum seen traversing through widened D2-3 neural foramina. There was scalloping of the left lateral posterior D3 vertebral body, adjacent pedicle and posterior end of left rib. There was compression and displacement of the cord toward the right side at D3 level by intradural component of mass [Figure 1].

The cord showed T2 hyperintense signal at the same level suggestive of cord edema. The extraforaminal component of the mass was extending from D2 vertebral level up to mid D5 vertebral level. The lesion was measuring 7.4 cm × 6.3 cm × 6.1 cm in size [Figure 2].

The patient was operated using a posterior approach. Laminectomy of D2-D3 vertebrae with near total excision

of dumbbell tumor from intraspinal region and posterior mediastinum was done. Capsule of the tumor in the posterior mediastinum was left behind as it was adherent to pleura. Since the case was considered as a neurofibroma, there was a possibility of compression of spinal cord by the intradural component, but no intradural extramedullary tumor extension was noted after opening the dura during surgery. Histopathological examination of resected specimen confirmed the diagnosis of neurofibroma.

Post-operatively physiotherapy and rehabilitation was given. The patient made an uneventful recovery with subsequent improvement in her neurological status. At the 2 month follow-up neurological examination showed 4+/5 power in both lower limbs and pain over shoulder was completely relieved. The patient was able to walk without support and could perform routine activities.

A neurofibroma in the spinal canal, invading the peripheral segment of the nerve by extending out of the intervertebral foramen and presenting itself with a dumbbell tumor is quite uncommon. Of the paraspinal neurofibromas, 72% were with intradural extramedullary localization, whereas 14% were with extradural, 13% were with dumbbell formation and 1% was with intramedullary localizations.^[2,9] The extraspinal part is usually larger than the intraspinal part in dumbbell neurofibroma.^[7] Tumor may attain massive dimensions, can be round to oval, lobulated and show cystic degeneration.

Dumbbell formation is important due to the attachment of the extramedullary part to the surrounding tissues. Local compression of the spinal cord results in the clinical findings. Common presentations include radicular pain and dysesthesia along with progressive motor weakness.^[3,5]

Direct radiographs are sufficient to establish the diagnosis in 50% of the cases. The most frequent findings on direct radiograph are pedicle erosion and vertebral body scalloping. Regular expansion of the interpedicular distance and intervertebral foramen may directly indicate the presence of the dumbbell tumor. MRI is highly sensitive and specific in detecting the disease, determining the accompanying pathologies and following the development of complications. Dumbbell neurofibromas appear as iso- or hyperintense to the spinal cord on T1-weighted images and give hyperintense signal on T2-weighted images. Regular enhancement is seen upon gadolinium administration in dumbbell neurofibroma.^[1,8]

Asymptomatic cases require no treatment while symptomatic cases need surgical intervention. Majority of the nerve fibers are entrapped by tumoral tissue in dumbbell neurofibroma cases as was the case in our patient as well. It is impossible to remove the tumor without sacrificing the nerve root and aggressive surgery may result in severe neurological deficits.^[3,4] Thus, partial resection should be preferred in dumbbell neurofibroma cases that cause compression of the spinal cord. As the aim of partial

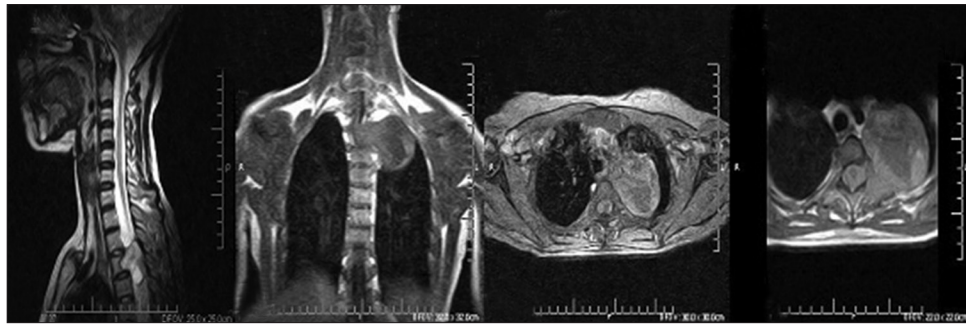


Figure 1: T1-weighted and T2-weighted magnetic resonance imaging of the thoracic dumbbell neurofibroma with extension to posterior mediastinum, the lesion being isointense on T1 sequence and hyperintense on T2 sequence

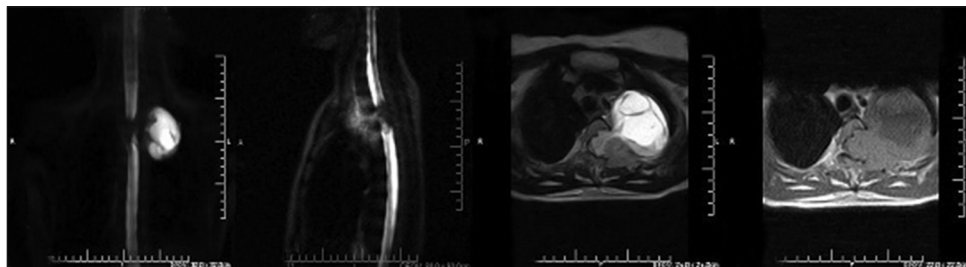


Figure 2: T1-weighted and T2-weighted magnetic resonance imaging of the thoracic dumbbell neurofibroma with extension to posterior mediastinum, the lesion being isointense on T1 sequence and hyperintense on T2 sequence

resection is to relieve the symptoms, the extent of surgical treatment is based upon the clinical picture of the patient. Decompressive surgery is a partial resection that carries the risk of recurrence and repeat surgery may be needed.^[3,5]

Dumbbell tumors with significant dissemination into the paraspinal region may require complex spinal exposure. Although two-stage operations may be performed to manage the intraspinal and paraspinal components separately, a single-stage procedure is preferable.^[6,10-14] Our patient presented with myelopathic findings and therefore decompressive excision of the tumor was planned. In our patient, near total excision of foraminal and extradural component including extension into the posterior mediastinum carried out using a single stage posterior approach with posterolateral extension. Tumor capsule was left intact in the mediastinum because it was adherent to pleura. Intraoperatively, dural exploration was performed for the intradural extramedullary component, but no tumor extension was found. Prognosis is excellent after the surgical resection. Recurrence is very rare subsequent to total excision. Recurrence after 3 years was noted in one of 66 paraspinal neurofibroma patients who were treated by Levy *et al.*^[3]

The most significant feature of dumbbell neurofibromas is the adhesion of the tumor to the surrounding environment by enlarging the foramen and projecting outward from the spinal canal.^[3,5] Dumbbell neurofibroma may present clinically as paraparesis or paraplegia (compressive myelopathy) even without intradural extramedullary extension as noted in our case. The goal of surgery is total removal of the tumor. Although a variety of surgical approaches for these lesions is available, most thoracic

spine dumbbell tumors can be effectively managed with a single-stage posterior approach with posterolateral extension (single posterior midline approach with laminectomy and costotransversectomy), which is easy to perform, secure and less invasive and was well-tolerated by the patient.^[3,7,13-14]

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