Abstract: Spinal neurofibromas are the most prevalent group of spinal tumors. They occur sporadically or in association with Neurofibromatosis type-1. A neurofibromas developing a dumbbell tumor with CVJ extension is a situation which is quite often seen. Surgical intervention is indicated when myelopathy and motor deficits develop in the case of paraspinal neurofibromas. The goal of surgery is total removal of the tumor. However, in selected cases partial removal of the tumor with adequate spinal cord decompression can be preferred to prevent severe complications such as vertebral artery injury. We present a case of Dumbbell C2 Neurofibroma with CVJ extension that was in close relation with the vertebral artery.

Keyword: Dumbbell formation, spinal neurofibroma, surgical treatment, cranio vertebral junction

Introduction
Nodular neurofibromas may occur anywhere on the peripheral nerves. They frequently originate from the dorsal roots and invade the sensory branches. They are capsulated and have a round and flexible structure. No treatment is needed for asymptomatic neurofibroma cases. Symptomatic cases justify surgical treatment. Surgical treatment of neurofibromas is total removal. Posterior laminectomy with unilateral facetectomy allows single-stage resection of dumbbell neurofibromas with significant intraspinal and paraspinal components. Best results are obtained from patients with minimal neurological deficits in the preoperative period.

Case Report
A 30 yr female patient admitted with c/o Occipital Head Ache x 1 year, Radiating to nape of neck and left lower jaw. H/o Tightness of all 4 limbs, H/o weakness in all for limbs, H/o gait disturbances, H/o multiple nodules over the face trunk and extremities since 10 years, gradually increase in size.

Fig 1. Patient
O/E NF1 Neurocutaneous markers (+), Multiple peripheral neurofibromas, Café AU lait spots >10 in No, Largest measuring 6 cm.

Fig 2. Café AU lait spots
Neurological examination revealed spastic Quadriaparesis and hyperactive deep tendon reflexes and bilateral plantar extensor.

Fig 3. MRI T1 contrast enhancing Dumbbell Mass with CVJ extension

Fig 4. MRI T2 Dumbbell Mass
In these dumbbell neurofibromas, the extraspinal part is usually larger than the intraspinal part. Tumor may reach massive dimensions, be lobulated and exhibit cystic degeneration. Dumbbell formation is important due to the attachment of, especially, the extramedullary part to the surrounding tissues. Its vicinity to the VA is critical. Clinical findings develop as a result of local compression of the ventral or motor nerve roots. While root symptoms develop during the early period, long-tract findings develop later. Cervical and lumbar regions are more frequently invaded. Radicular pain and dysesthesia were present in 80% cases. Motor weakness that we detected in our case is seen in some 10% of the cases. Direct radiographs are sufficient to establish diagnosis in 50% of the cases. Pedicle erosion and vertebral body scalloping are the most frequent findings on direct radiograph. Regular expansion of the interpedicular distance and intervertebral foramen may directly indicate the presence of the dumbbell tumor. Thanks to its sensitivity and specificity, MRI is quite important in detecting the disease, determining the accompanying pathologies and following the development of the complications in NF1 cases. Neurofibromas appear iso- or hyperintense to the spinal cord on T1-weighted images while they give hyperintense signal on T2-weighted images. Dumbbell neurofibromas enhance regularly upon gadolinium administration. No treatment is needed for asymptomatic neurofibroma cases. Symptomatic cases justify surgical treatment. Majority of the nerve fibers are entrapped within tumoral tissue in dumbbell neurofibroma cases, as in our case. It is impossible to remove the tumor without sacrificing the nerve root and aggressive surgery may result in severe neurological deficits. Thus, partial resection should be preferred in dumbbell neurofibroma cases that cause compression of the spinal cord. As the aim of partial resection is to resolve the symptoms, the extent of surgical treatment is shaped according to the clinical picture of the patient. 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. For cervical tumors, the VA is another issue to be considered. In most instances, meningiomas and nerve sheath tumors receive little blood from the spinal cord and are attached by few adhesions to the spinal cord. Most cervical dumbbell tumors can be adequately accessed through a standard laminectomy and complete unilateral facetectomy. As in our case, this allows paraspinal access up to 3 cm from the lateral dural margin. A second-stage anterior procedure may be required if further tumor extension is present. The VA is consistently displaced anteromedially by dumbbell neurofibromas of the cervical spine. The artery is neither encased nor invaded by these tumors but is separated from the tumor capsule by a thin layer of periosteum and perivertebral veins. These tissues serve as an effective and easily developed plane of dissection that is rarely associated with VA injury. Thus, because of the low risk of either VA injury or its potential ischemic consequences, preoperative angiography and/or test occlusion or early intraoperative control and mobilization do not seem warranted.
Prognosis is excellent after the surgical resection. While pain is diminished in 80% of the cases, total remission occurs in 60% of the cases. 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. However, upper cervical neurofibroma cases characterized by dumbbell formation, as in our case, are treated by partial resection, thus they have the risk of recurrence. It is crucial to screen cervical spine in these patients by advanced imaging modalities such as MRI to detect recurrence.

CONCLUSION

The most significant feature of dumbbell neurofibromas is the adhesion of the tumor to the environment by enlarging the foramen and projecting outward from the spinal canal. The goal of surgery is total removal of the tumor. Although a variety of surgical approaches for these lesions is available, most cervical spine dumbbell tumors can be effectively managed with a single-stage posterior exposure with partial laminectomy and unilateral facetectomy. However, in selected cases partial removal of the tumor with adequate spinal cord decompression can be preferred to prevent vertebral artery injury.

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