Case Report

Symmetric Dumbbell Ganglioneuroma of Bilateral C1 Roots With Intradural Extension Associated With Von Recklinghausen's Disease: A Case Presentation

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Summary

Introduction: Ganglioneuromas are rare benign tumours arising most commonly from the sympathetic nervous system. They occasionally grow in a dumbbell fashion extending into the spinal canal extradurally. But, ganglioneuromas in association with von Recklinghausen's disease are rare.

Case presentation: A 45 year old male, was admitted to emergency unit with complaints of pain in both of arms and neck, progressive difficulty of swallowing and respiratory distress. A lots of cafe-au-lait and small subcutaneous noduls were seen on whole body and two fregling in axillary region were detected on physical ex amination. A moderate quadriparesis and IX-X cranial nerve dysfunction was observed on neurological examination. Cranio-cervical region magnetic resonance imaging revealed a bilatera lly and symmetric mass lesion located in C1 region, spreaded to intra and extradural space like as known as “dumbbell shape”. The patient was undergone surgery and tumour was totally removed with microsurgical technique via posterior approach. Ganglioneuroma was histopathologically verified. Postoperative period was uneventful and the patient was discharged as neurologically intact. Magnetic resonance imaging performed three months after surgery proved the total resection of tumour.

Discussion: Ganglioneuromas are a subgroup of neuroblastic tumors that have been defined as childhood embrional tumors of migrating neuroectodermal cells derived from the neural crest. Ganglioneuromas originating within spinal canal constitute less than 10 percent of all ganglioneuromas. Furthermore involvement of cervical spine is extremely rare. Total surgical removal of tumour is usually curative and satisfactory. Patient must be follow in neurointensive care unit because the threat of respiratory distress and swallowing difficulty in early postoperative period. Neurological symptoms usually disappear gradually.

Key words: Ganglioneuroma, cervical spine, dumbbell tumor, von Recklinghausen's disease

Von Recklink Housen Hastalığı'na Eşlik Eden İnadural Simetrik Bilateral C1 Köklerinden Kaynaklanan Ganglionöroma; Vaka Sunumu

Özet


Vaka sunumu: 45 yaşında erkek hasta, acil polikliniğe yutma güçlüğü ve solunum zorluğu şikayet ile getirildi. Hastada ciltaltında çok sayıda nörofibrom, koltuk altında 2 adet freckling ve çok sayıda cafe au lait lekesi tespit edildi. Nörolojik muayenede, hastada GAG reflexi
INTRODUCTION
Ganglioneuromas are rare, slow-growing benign tumors arising from tissues of the neural crest, most commonly from the sympathetic nervous system. They occasionally grow in a dumbbell fashion extending into the spinal canal extradurally. However, ganglioneuromas located in the cervical spine originating from the sensory nerve extending intradurally are extremely rare. Association with Von Recklinghausen's Disease=Neurofibromatosis-1 (NF-1) showed rarely(8). We present a case of bilateral and symmetric dumbbell ganglioneuroma of the cervical spine with intradural extention associated with NF-1.

CASE PRESENTATION
A 45 year old male patient was admitted to our hospital with complaints of of 1 year of pain in neck and both of arms, 1 week of progressive difficulty of swallowing and respiratory distress. He was dispneic and there was crepitation on oscillation of lungs. Respiratuar sounds were also diminished on oscillation of both of lungs. Physical examination also revealed a lots of neurofibrom in subcutaneous tissue, lots of “café-au-lait” and two freckling in axillary region. Neurological findings included, moderate quadripareisis with increased muscle tonus and absent of velopharyngeal reflex. Cranio cervical magnetic resonance (MR) imaging disclosed a well demarcated mass bilateral symmetrically spreaded to intradural and extradural space like a “dumbbell shape” at C1 level (Figure-1a,b). Respiratory arrest occurred in a short time after hospitalization. He was entubated and ventilated. He was undergone surgery via posterior approach immediately. Tumour was completely removed by microsurgical technique with bilateral C1 roots which thought as tumor originated (Figure-2a,b). The patient was followed in neurosurgical intensive care unit for 1.5 month. Ventilatory support was ended after 1.5 month and he could able to walk with support after 1 month. The histopathology of the tumour was reported as ganglioneuroma. The patient was diagnosed as ganglioneuroma with accompanying NF-1.
Figure 1a: A hyperintense mass overlying the left transverse process was observed on preoperative, sagittal, contrast enhanced, T1-weighted magnetic resonance images of the spine.

Figure 1b: On axial, T2-weighted magnetic resonance images of the patient performed preoperatively, a hyperintense mass, with intra-and-extradural components, extending toward the bilateral transverse process was observed.

Figure 2a: Sagittal T2-weighted MR images of the cervical spine of the patient taken 1 month after the operation clearly show that the intradural component of the tumor at the level of C1-C2 was excised and there was no spinal cord compression.

Figure 2b: Axial T2-weighted MR images of the cervical spine of the patient taken 1 month after the operation clearly show that the intradural component of the tumor at the level of C1-C2 was excised and there was no spinal cord compression.
DISCUSSION

Ganglioneuromas are a subgroup of a class of neuronal tumour which include cells express a mature neuronal phenotype. Ganglioneuromas can be originated in central nervous system, peripheral nervous system or in spinal column. Peripheral nervous system involvement is much more common than the others. Peripheral neuroblastic tumors are classified into three subgroups depending on the degree and type of neuroblastic differentiation and the degree of schwannian stroma development; neuroblastomas, ganglioneuroblastomas and ganglioneuromas\(^\text{(2,13,15)}\). Spinal ganglioneuromas encompass the features of both the central and peripheral types\(^\text{(6,7)}\). Ganglioneuroma is the fully differentiated benign counterpart of neuroblastoma. The majority of ganglioneuromas occur in children older than 10 years of age and are chiefly located in the posterior mediastinum and retro peritoneum.

According to report from the Armed Forces Institute of Pathology, among 88 cases of ganglioneuromas 34 were encountered in the posterior mediastinum, 27 in the retroperitoneum, 19 in the adrenal, 5 in the pelvis, 2 in the cervical area and 1 in the parapharyngeal space\(^\text{(4)}\). Paraspinal ganglioneuromas may extend rarely by the neuronal foramen into the spinal canal\(^\text{(1,10)}\). Multiple occurrences and an association with NF-1 are another unusual features\(^\text{(12,14,16)}\). To date Approximately 20 cases of ganglioneuromas associated with NF-1 have been reported but the genetic association between NF-1 and ganglioneuromas is unclear\(^\text{(5,9)}\). Jiang et al reported, surgical approach and development of an anatomic classification system in surgical treatment of cervical dumbbell tumors. According to the study the classification covers all tumor types and is easier to remember. It is practical and useful for determining the surgical approach. The recurrence rate decreases significantly after radial tumor resection. Revision surgeries are associated with more complications\(^\text{(17)}\). Multiple ganglioneuromas are often found outside the spinal column. The term “dumbbell” is used to designate the growth pattern of the tumour such as in relation to the intervertebral foramen; intraspinal-extravertebral, intraspinal-foraminal or

*Figure 3a:* Contains a thin fibrous capsule of the tumor area.

*Figure 3b:* Under the capsule formed by mature ganglion cells and proliferating schwann cell bundles and irregular and scattered ganglion cells.
foraminal-extravertebral, and also in relation to the dura in the spinal canal, intra-extradural[3,9]. In many cases of dumbbell ganglioneuroma the intraspinal portion of the tumor extends extradurally but intradural extension is extremely rare[14,16]. Ganglioneuromas of the cervical spine are also extremely rare, with only 5 pathologically confirmed and previously reported cases[18,19]. All these cases presented a dumbbell pattern. Only two these have located symmetrical and extradural and NF-1 have associated in only one case. The intraspinal component can cause spinal cord compression. Spinal tumour may present as incidental or spinal cord compression symptoms[11,16]. In the present study, the tumour was originated from C1 root and spread to intradural and extradural space symetric and bilaterally at C1 level. A ganglioneuroma accompanying to NF-1. Ganglioneuromas as are rare, slow growing, benign tumours. Total surgical removal should be preferred treatment in symptomatic patients. Total excision can be achieved in most patients. En bloc resection with source of root can be done some spinal level. Preoperative evaluation of radiologic findings will be benefit to decide whether scarify the affected root. The majority of reported cases in literature showed significantly improvement of symptoms following surgical removal of tumors. Postoperative care is crucial for critical location for vital parameter like in the present case.

CONCLUSION
This report concerns an extremely rare case of ganglioneuromas in a 45 year old man with associated NF-1 who presented bilateral, extradural and symmetric dumbbell ganglioneuromas with intradural extension. This tumor involved the bilateral C1 nerve roots. A case similar to our patient has been reported as unique one by Kyoshima et al. Although these tumors are begin and grows slowly upper cervical localization may cause progressive neurologic deterioration and death due to respiratory distress in early period. Total surgical removal should be preferred therapeutical management. Gradually recover of neurological symptoms may respect with follow in neurointensive care unit after easily postsurgical period in patients with critical location like in present case. Patients with ganglioneuroma accompanying NF-1 disease should be taken long term follow-up because of possible systemic problem as other tumors.

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