



Primary Extradural Spinal Ganglioneuroblastoma: A Case Report

Primer Ekstradural Spinal Gangliyonöroblastom: Bir Olgu Sunumu

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ABSTRACT

We report a case of ganglioneuroblastoma of the spinal cord in a 25-year-old man. Clinical history was short with paraparesis and bladder involvement. The MRI picture was that of an extradural solid tumor with extension to both intervertebral foramina, more suggestive of nerve sheath tumour rather than malignant embryonal tumour. Diagnosis was established by histopathological study. We could not find such a presentation of spinal ganglioneuroblastoma as an extradural, primary tumour in the literature. We discuss the radio-pathological features, surgical management and post-operative outcome.

KEYWORDS: Ganglioneuroblastoma, Spinal, Extradural, Primary

ÖZ

25 yaşında bir erkekte omurilikte gangliyonöroblastom olgusu bildiriyoruz. Klinik öykü kısaydı ve paraparezi ve mesane tutulumu içeriyordu. MRG'de her iki intervertebral foramene uzanan ekstradural bir solid tümör vardı ve malign bir embriyonal tümörden çok sinir kılıfı tümörü düşündürüyordu. Tanı, histopatolojik incelemeyle kondu. Literatürde spinal gangliyonöroblastomun bir ekstradural primer tümör olarak ortaya çıkmasıyla ilgili bir çalışma bulamadık. Radyopatolojik özellikler, cerrahi tedavi ve postoperatif sonucu tartışıyoruz.

ANAHTAR SÖZCÜKLER: Gangliyonöroblastom, Spinal, Ekstradural, Primer

INTRODUCTION

Ganglioneuroblastomas are rare tumours with origin from the neural crest cell. They lie between benign gangliogliomas and frankly malignant neuroblastomas with respect to their malignant potential as they contain both malignant neuroblastomatous and benign ganglioneuromatous elements. They occur mainly in adolescents and young adults. Most commonly they are found in the temporal lobe of the cerebral hemispheres (8). However, they can occur in brain stem, cerebellum, and ventricles. Primary occurrence of the tumour in the spinal cord is rare (9,10,11). Complete surgical excision is the main goal of management, whenever possible. Adjuvant radiotherapy should be given to every case in view of the high-grade nature (Grade IV) of the tumour, even if there is complete removal. In spite of all therapies, the long-term prognosis of these tumours continues to be poor.

CASE REPORT

A 25-year-old man was admitted with a short history of pain in both the lower limbs and urinary retention followed by complete paralysis. Patient had electric shock like sensation and burning pain radiating down both lower limbs, starting 15 days before admission to our hospital. Simultaneously he had retention of urine for which he was catheterized. Within a period of a week there was paraplegia with grade 0 power

around all the joints in both lower limbs. Muscles were grossly hypertonic with exaggerated jerks and clonus. All modalities of sensations were lost below D4 level.

T1-weighted magnetic resonance imaging showed an irregular, hypointense lesion in extradural location behind the body of D2 vertebrae compressing the cord to left side. The major part of it was on right side with both the intervertebral foramina filled with tumour tissue. Left side portion filling the foramen was continuous with its right counterpart anterior to the cord. On T2-weighted images the lesion was isointense relative to normal cord. After contrast administration, there was strong enhancement (Figure 1,2). Hyperintense signal changes were noted in the anterior part of D2 vertebral body. Imaging of the brain for primary lesion was negative.

Intraoperatively, after laminectomy at D-1 & D-2 level, a soft elastic, moderately vascular, extradural tumour compressing the spinal cord on the right side found. Tumour was found to enter both the D2 intervertebral foramina. Right and left portions of the tumour were continuous anterior to the cord. Tumour was removed completely in piecemeal fashion.

Hematoxylin-eosin-stained sections showed tumor cells in all stages of neuronal differentiation, from ganglion cells to immature neuroblasts. The tumour cells were found to be arranged in well-defined nests composed of a mixture of

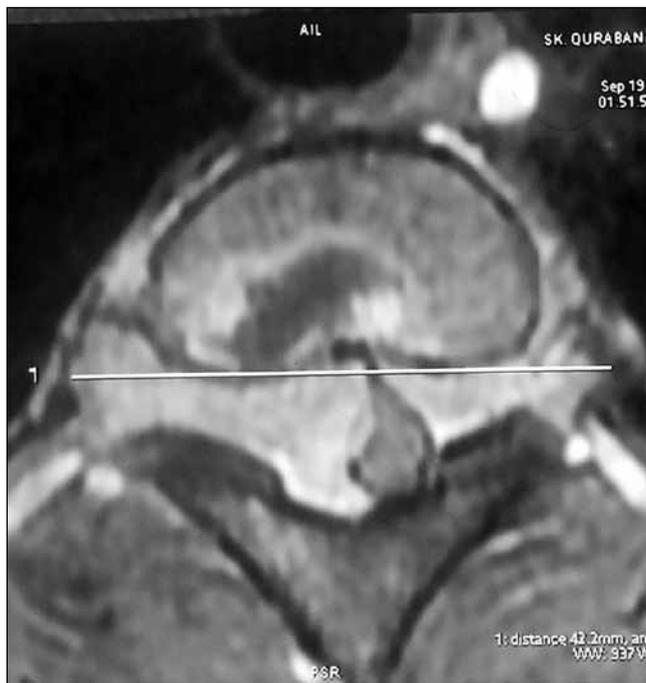


Figure 1: Axial contrast MRI showing extradural location of the tumour.



Figure 2: Sagittal contrast MRI.

neuroblastic cells and maturing ganglions in a neurofibrillary background (Figure 3). Immunohistochemistry revealed a negative staining for glial fibrillary acidic protein, whereas S-100 immunoreactivity was positive in most of the cells (Figure 4). The final diagnosis was ganglioneuroblastoma of the spinal cord.

Post-operatively the patient's spasticity in both lower limbs decreased and there were small flickering movements in right lower limb. In view of the high-grade nature of the ganglioneuroblastoma, adjuvant radiotherapy was advised, but the patient declined. After 4 months of post-op follow-up, the patients' power in both lower limbs was grade 2.

DISCUSSION

In its recent classification of central nervous system tumours in 2007, the World Health Organisation (WHO) divided the embryonal tumours into 3 categories, namely, the medulloblastomas, the primitive neuroectodermal tumours (PNETs) and the atypical rhabdoid/teratoid tumours (4). All of them belong to malignant grade IV tumours histologically. PNETs include CNS ganglioneuroblastoma, CNS

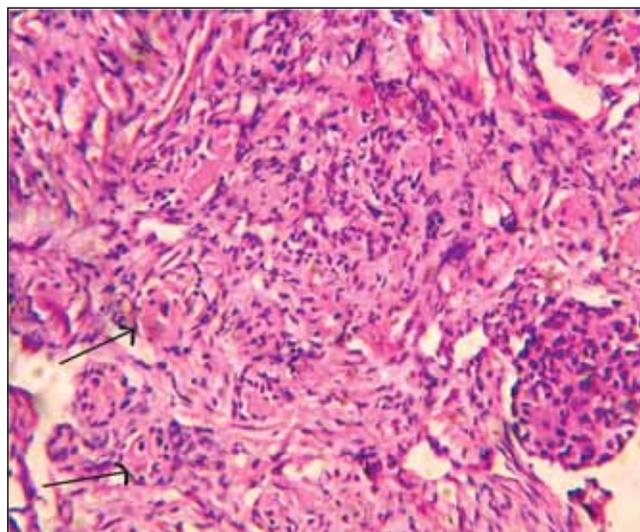


Figure 3: Light microscopy showing small round tumour cells arranged in well defined nests with fine, fibrillary network between groups of tumour cells (arrow marks).

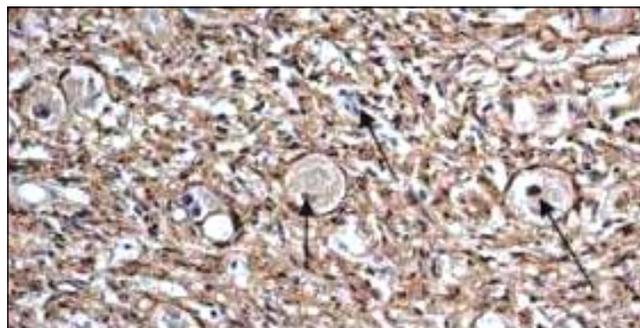


Figure 4: Immunohistochemistry: S100 positivity.

neuroblastoma, medulloblastoma and ependymoblastoma. Histologically, ganglioneuroblastomas show small, round, immature tumour cells in all stages of neuronal differentiation with immature ganglion cells interspersed in between them. It is most commonly seen in older children and adolescents. Adrenal medulla is the commonest site for these types of malignant tumours. The most common extra-adrenal site is posterior mediastinum and they are rarely found in the central nervous system. The most common site in the brain is the temporal lobe followed by the cerebral hemisphere, the floor of the third ventricle, the brain stem, the cerebellum and the spinal cord. Ganglion cell tumours of the spinal cord are extremely rare. Most of the reported cases of ganglioneuroblastomas are located at cervical spinal level. Being highly malignant these tumours could have spread from their more common primary location, i.e. the brainstem. Therefore, before labelling these tumors as primary spinal, it is important to rule out clinico-radiologically any primary site in brain particularly brainstem and posterior fossa which could have metastasized to cervical region.

Disturbance and difficulty in walking due to motor weakness in lower limbs is the most common presentation of ganglioneuroblastomas. Sometimes there may be bladder, bowel involvement, particularly in intra-medullary variety. Scoliosis and asymmetric lower limb length can also be found. The standard treatment of ganglioneuroblastoma is complete surgical excision. Durotomy with myelotomy is required for intramedullary tumours. However, complete surgical excision is not always feasible due to infiltrative nature of the tumour as well as its frequent intramedullary location. Simple laminectomy with complete removal of the tumour is possible in pure extradural tumours, as in the present case. The amount of residual tumour is the most important predictor of recurrence. However, the effect of degree of resection on overall survival is not clearly known due to rarity of such tumours. All patients with confirmed diagnosis of ganglioneuroblastoma should receive radio-chemotherapy. Radiotherapy should be given as adjuvant treatment even if there is complete excision in patients over 12 years of age, in view of the high grade (Grade IV tumours) nature of the ganglioneuroblastoma. Rare cases of complete response to chemotherapy alone without any surgery, where excision was not possible has also been reported in the literature (7). In spite of good surgical excision and with adjuvant therapy, prognosis of patients with these tumours is unfavourable due to the aggressive, malignant nature of the tumour with frequent recurrences as well as distant metastases. The 5-year survival even after complete removal is only around 30%; a high recurrence rate and meningeal involvement has also been reported (1,2). Deaths are mostly due to distant metastases such as to the lung and liver, and intercurrent infections.

The most common location of primary spinal ganglioneuroblastoma is the thoracic, intramedullary site. Our illustrative case is significant because it was located in extradural region and filling the intervertebral foramina on both sides. Although

there has been a report of an extradural spinal ganglioneuroblastoma, it was associated with an abdominal component and the primary nature of the spinal component was not well established (3). Most recently Miele et al reported a extensive holocord primary ganglioneuroblastoma from the cervical to the conus region at the intramedullary location (5). Although Ozdemir et al reported a case of primary primitive neuroectodermal tumour in the lumbar extradural space, the histology of ganglioneuroblastoma was not specified (6). We could not find any other report of a primary ganglioneuroblastoma in an extradural location in the literature. There was no primary lesion either in brain, mediastinum or retroperitoneum. Our patient had a history of very short duration indicating the aggressive and malignant nature of the tumour. The tumour was radiologically similar to more common benign tumours of the thoracic region with intervertebral foramina extension such as neurofibroma and except for short clinical history there was no other specific difference. Neuroblastic embryonal tumours should therefore be considered in the differential diagnosis of extradural thoracic tumours in a young patient with no primary source found elsewhere.

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