

## Case Report:

# Extension of Retroperitoneal Neuroblastoma to Spinal Canal: A Case Report



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## ABSTRACT

Neuroblastoma is a nervous system malignancy. The extension of intra-abdominal neuroblastoma to the spinal canal is a rare condition. In this case report, we present a patient with monoplegia and a huge intra-abdominal mass with intra spinal extension and cord compression.

## Highlights

- Neuroblastoma is a malignancy of nervous system
- Intra- abdominal tumor can extended to spinal canal due to the motor and sensory deficits

## Plain Language Summary

Neuroblastoma is a malignancy of nervous system. The extension of this tumor to spinal canal and spinal cord compression is very rare and can have permanent neurological deficit like paraplegia or sensory loss. This tumor should be examined in infant patient with monoplegia or paraplegia.

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## 1. Introduction

**N**euroblastoma is a nervous system malignancy that accounts for 8-10 % of all pediatric tumors [1] and can involve the spinal canal. The patient is admitted to the neurosurgery ward with a sign of spinal cord compression and it is estimated that 10% of all patients with neuroblastoma have spinal epidural compression and half of them are infants. The extension of the intra-abdominal tumor to the spinal canal is very rare and many articles emphasize this [1-3]. Thus, we introduce an 8 month old patient admitted with a monoplegia and epidural mass.

## 2. Case Presentation

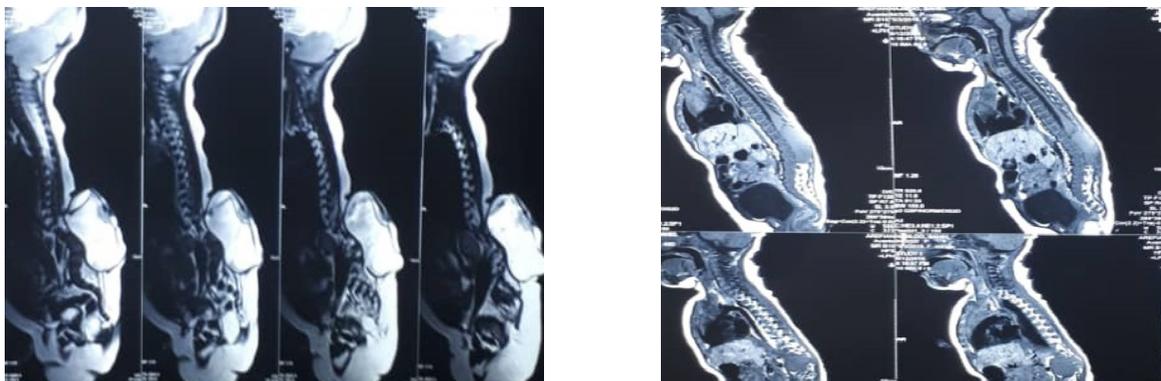
A female 8 month old is presented with no motility of the left leg as of 1 month ago, revealed by the clinical evaluation to be spastic and completely plegic. Other examinations were normal and her general appearance was normal. Spinal Magnetic Resonance Imaging (MRI) was performed and showed a solid retroperitoneal mass, 5×3.5 cm in diameter, entering the spinal canal through the foramina from T- 12- L1 until L4-L5 (Figure 1). Because of cord compression, the patient was taken into the operation room. In the prone position, standard laminectomy was done. The dura matter was opened from the midline and all the tumoral parts were resected and the roots of L1- L5 were released. The part that extended to the abdominal and retroperitoneum were intact after irrigation and hemostasis. After the patient's dura matter, fascia, and skin were repaired, she was admitted to the ICU. Twenty four hours after surgery, the patient was able to move her leg, and her spastic tone improved significantly. At the time of writing this article, her left foot muscles force (M4).

## 3. Discussion

Neuroblastoma is a tumor of the sympathetic nervous system most commonly seen in infancy. In the United States, this is seen in 1 case per 100,000, and each year, 700 cases are added to this number [4]. The embryonal studies show this malignancy arising from neural crest cells. Based on the International Neuroblastoma Pathology Classification (INPC), four subtypes can be identified: neuroblastoma (Schwannian stroma-poor), ganglioneuroblastoma- intermixed (Schwannian stroma-rich), ganglioneuroblastoma-nodular (composite, Schwannian stroma-rich/ stroma-dominant and stroma-poor), and ganglioneuroma (Schwannian stroma-dominant) [5].

In this patient, we found soft tissue nearly 3x4 cm in diameter involved in at least 4 levels of the spinal column. In microscopic evaluation, this extended into the retroperitoneum where we found round blue cell tumors small to medium in size, salt and pepper colored, with an elongated shape, possibly distinct nucleoli, indiscernible amounts of cytoplasm, vague cytoplasm borders compatible with neuroblastoma. A genetic analysis showed no chromosomal abnormality. The patient who presented us with extra cranial neuroblastoma had an excellent outcome due to low affinity of metastasis [6]. Also, children at this age can completely be cured if they have favorable genetic profiles, particularly consisting of a single copy of MYCN oncogene and no abnormal 1p and 11q chromosomes. [7, 8].

In this patient, the diagnosis was made with microscopic histopathology where we evaluated the urinary vimentin after the pathologist reported that the clinical value of urine vimentin is high. Based on the guidelines of the International Society of Pediatric Oncology Eu-



**Figure 1.** sagittal and parasagittal spinal MRI in T1 & T2 sequence : Huge mass with intra abdominal and extra abdominal compartment which was Hyper signal in T2 and iso- hyper signal in T1 and compress the spinal cord

rope Neuroblastoma (SIOPEN) for symptomatic neuroblastoma [9], they emphasized the physician to strongly recommend surgical approach once they reach a definitive diagnosis [10-12]. In this patient, the complete resection of the spinal part of the tumor was done. Meanwhile chemotherapy is the second step of treatment, at the time of writing this article, the patient improved significantly, and no mass effect was seen in the post operation MRI. Guidelines and many authors advise rapid diagnosis, prompt chemotherapy, and early decompression in order to avoid a subsequent huge mass in the spinal canal.

#### 4. Conclusion

Neuroblastoma can be managed with chemotherapy, but surgical evaluation should be considered when the physician needs tissue samples for a definitive diagnosis or detection of a neurological deficit. Extension and manifestation of the neuroblastoma to the spinal canal with a sign of cord compression and neurological deficit is a rare condition, though early decompression and chemotherapy can resolve the tumor and prevent permanent neurological deficits.

#### Ethical Considerations

##### Compliance with ethical guidelines

All phases of this research have been confirmed by the ethical committee of Urumia University of Medical Sciences (Code Number: 43256/4354).

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##### Authors contributions

All authors contributed in designing, running and writing all parts of the research.

##### Conflict of interest

The authors declared no conflict of interest.

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#### References:

- [1] Wylie LA, Hardwick LJA, Papkovskaia TD, Thiele CJ, Philpott A. Ascl1 phospho-status regulates neuronal differentiation in a Xenopus developmental model of neuroblastoma. *Disease Models & Mechanisms*. 2015; 8(5):429-41. [DOI:10.1242/dmm.018630] [PMID] [PMCID]
- [2] Nakagawara A, Azar CG, Scavarda NJ, Brodeur GM. Expression and function of TRK-B and BDNF in human neuroblastomas. *Molecular and Cellular Biology*. 1994; 14(1):759-67. [DOI:10.1128/MCB.14.1.759] [PMID] [PMCID]
- [3] Shimada H, Ambros IM, Dehner LP, Hata JI, Joshi VV, Roald B, et al. The international neuroblastoma pathology classification (the Shimada system). *Cancer: Interdisciplinary International Journal of the American Cancer Society*. 1999; 86(2):364-72. [DOI:10.1002/(SICI)1097-0142(19990715)86:2<364::AID-CNCR21>3.0.CO;2-7]
- [4] Brodeur GM, Pritchard J, Berthold F, Carlsen NL, Castel V, Castelberry RP, et al. Revision of the international criteria for neuroblastoma diagnosis, staging and response to treatment. *Journal of Clinical Oncology*. 1993; 11(8):1466-77. [DOI:10.1200/JCO.1993.11.8.1466] [PMID]
- [5] Schleiermacher G, Janoueix-Lerosey I, Ribeiro A, Klijanienko J, Couturier J, Pierron G, et al. Accumulation of segmental alterations determines progression in neuroblastoma. *Journal of Clinical Oncology*. 2010; 28(19):3122-30. [DOI:10.1200/JCO.2009.26.7955] [PMID]
- [6] Bourdeaut F, Ferrand S, Brugières L, Hilbert M, Ribeiro A, Lacroix L, et al. ALK germline mutations in patients with neuroblastoma: A rare and weakly penetrant syndrome. *European Journal of Human Genetics*. 2012; 20(3):291-7. [DOI:10.1038/ejhg.2011.195] [PMID] [PMCID]
- [7] Devoto M, Specchia C, Laudenslager M, Longo L, Hakonarson H, Maris J, et al. Genome-wide linkage analysis to identify genetic modifiers of ALK mutation penetrance in familial neuroblastoma. *Human Heredity*. 2011; 71(2):135-9. [DOI:10.1159/000324843] [PMID] [PMCID]
- [8] De Pontual L, Kettaneh D, Gordon CT, Oufadem M, Boddaert N, Lees M, et al. Germline gain-of-function mutations of ALK disrupt central nervous system development. *Human Mutation*. 2011; 32(3):272-6. [DOI:10.1002/humu.21442] [PMID]
- [9] Stallings RL, Nair P, Maris JM, Catchpoole D, McDermott M, O'Meara A, et al. High-resolution analysis of chromosomal breakpoints and genomic instability identifies PTPRD as a candidate tumor suppressor gene in neuroblastoma. *Cancer Research*. 2006; 66(7):3673-80. [DOI:10.1158/0008-5472.CAN-05-4154] [PMID]
- [10] Janoueix-Lerosey I, Novikov E, Monteiro M, Gruel N, Schleiermacher G, Llorion B, et al. Gene expression profiling of 1p35-36 genes in neuroblastoma. *Oncogene*. 2004; 23(35):5912-22. [DOI:10.1038/sj.onc.1207784] [PMID]
- [11] Stigliani S, Coco S, Moretti S, Oberthuer A, Fischer M, Theissen J, et al. High genomic instability predicts survival in metastatic high-risk neuroblastoma. *Neoplasia*. 2012; 14(9):823-32. [DOI:10.1593/neo.121114] [PMID] [PMCID]
- [12] Kryh H, Carén H, Erichsen J, Sjöberg RM, Abrahamsson J, Kogner P, et al. Comprehensive SNP array study of frequently used neuroblastoma cell lines; Copy neutral loss of heterozygosity is common in the cell lines but uncommon in primary tumors. *BMC Genomics*. 2011; 12(1):443. [DOI:10.1186/1471-2164-12-443] [PMID] [PMCID]

