

Surgical Outcomes in Primary Benign Extradural Tumors Of The Cervical Spine: A Case Report

Nurul Musfirah^{1*}, Iskandar², Sigit Ananda³

¹General Practitioners at Harapan Bunda General Hospital

²Department of Neurosurgery, Faculty of Medicine, Syiah Kuala University, Banda Aceh, Indonesia

³Department of Neurosurgery, Faculty of Medicine, Sumatera Utara University, Medan, Indonesia

*Corresponding Author: nurulakla.wahid@gmail.com

ARTICLE INFO

Article history:

Received June, 27th 2023

Revised July, 23rd 2023

Accepted August, 22nd 2023

Available online August, 30th 2023

E-ISSN: 2686-0848

How to cite:

Musfirah N, Iskanda, Ananda S. Selection of surgical interventions in pediatric hydrocephalus: a literature review. Asian Australasian Neuro and Health Science Journal, 2023 August 05(02);69-74



This work is licensed under a Creative Commons Attribution-ShareAlike 4.0 International.

DOI : 10.32734/aanhsj.v5i2.12588

ABSTRACT

Introduction: The prevalence of spinal tumors is rare, only about 15% of all cases of central nervous system tumors and 90% of cases occur at the age of >20 years, which is a productive age for a person.

Case Report: A young 25-year-old woman came to the polyclinic complaining of weakness of both hands and feet (all limbs). The MRI results show a suspected of a space occupying lesion of the extradural spinal cord. The patient then underwent tumor removal laminectomy surgery with a posterior approach. After the operation, his motoric was improving and increased to 4/4. After three months she was able to walk and perform daily activities.

Discussion: In younger patients, the tumor diagnosis may be more catastrophic, and the required treatment can have a severe impact on the patient's health-related quality of life. The patient was also very fast and assisted with early ambulation. This can be related to the progressivity of tumor cells, in this case a benign tumor with a border that is not aggressive in invading the surrounding tissue.

Conclusion: Early diagnostic and complete surgical resection before the occurrence of severe symptoms will show an excellent prognosis in benign primary extradural tumor. Multidisciplinary surgical planning should therefore also include the histological grade, stage, and extent of the tumor to establish the objective biological aggressiveness of the overlying tumor and in turn estimate the feasibility of resection

Keywords: Benign; Primary extradural tumor; Surgical outcome

1. Introduction

Spinal cord tumor is a tumor of the central nervous system, where this condition occurs when the spinal cord experiences abnormal tissue growth that can be benign or malignant.[1] The prevalence of spinal tumors is rare, only about 15% of all cases of central nervous system tumors and 90% of cases occur at the age of >20 years, which is a productive age for a person.[2]

Anatomically, spinal tumors can be classified as extradural tumors, intradural-extramedullary tumors, and intramedullary tumors based on their relationship to the dura and spinal cord. The incidence of primary spinal tumors both benign and malignant is estimated to be about 0.098 cases per 100,000 person-years, and primary spinal tumors account for as much as 4%–8% of all incidences of primary central nervous system (CNS) tumors.[2] Benign tumors account for 60%–70% of all primary spinal tumors. Spinal tumors are often difficult to diagnose because of their nonspecific manifestations, and specific imaging modalities are required.[3]

Treatment of spinal tumors varies depending on the stability of the spine, neurological status and pain level of the patient. The main treatment for spinal tumors is surgery. Indications for surgery may vary

depending on the type of tumor. The aim is to completely remove the tumor while preserving maximal neurological function.[4]

Primary (non-metastatic) spinal tumors can be completely removed by en bloc resection to promote healing. If surgery is being considered, the approach to the tumor is determined by the location of the tumor within the spinal canal. The posterior (back) approach allows identification of the dura and exposure of the nerve roots.[5] This approach is generally used for tumors in the posterior portion of the spine or to expose tumors within the dura. Multiple levels can be decompressed, and graded segmental fixation can be performed if necessary.[6]

2. Case Report

A young 25-year-old woman came to the polyclinic complaining of weakness of both hands and feet (all limbs). This happened slowly and progressively since 6 months ago, initially the patient only complained of numbness, but over time the patient complained of pain to weak limbs. When a neurological examination was carried out, a motor value of 1/1 was obtained in all four extremities.

The patient then performed an MRI examination with sagittal and axial sections to confirm the diagnosis and obtained the results of a suspected primary spinal cord tumor and it was confirmed that there was no metastatic process from other organs. The MRI results show a mix-intensity lesion with a size of 4x3x2 cm as high as the C1-C2 level that is pressing on the spinal canal and is suspected of a space occupying lesion (SOL) of the spinal cord extradural.



Figure 1. MRI of spinal cord with sagittal and axial sections.

The patient then underwent tumor removal laminectomy surgery with indications of primary extradural spinal cord tumors with a posterior approach. Intraoperatively, lamina C2 was destroyed, duramater intake and total tumor removal was performed.



Figure 2. Intraoperative and postoperative total removal laminectomy with posterior approach

After total tumor resection then the sample was examined at Anatomical Pathology and histopathological results showed tissue consists of large spindle cells with collagen fibers with small spindle cells arranged to form a palisade and no signs of malignancy, conclusion is a neurofibroma.

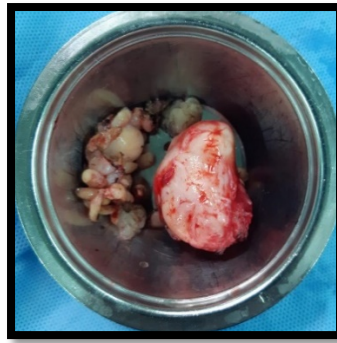


Figure 3. Macroscopic morphology of the tumor

After the operation, three days postoperative the patient had done early ambulation and his motoric was improving and increased to 4/4. After three months postoperative, the patient returned to polyclinic for control and was able to walk and perform daily activities.



Figure 4. Condition of the patient 3 months postoperative

3. Discussions

Primary extradural tumors of the spine have a small percentage of 4% compared to all spinal tumor classifications. Extradural tumor cases are usually malignant and a metastatic process, thus increasing the challenges for management, especially radioresistance which will be relatively increased in these cases, invasive nature and surgery can increase morbidity and mortality quite high.[7] In addition, if the tumor is malignant it will be very difficult to achieve complete resection due to irregular tumor margins and invasion into surrounding tissue with its aggressive nature making it difficult to achieve a clean tumor margin, this may be greatly complicated by critical surrounding anatomy (e.g. nerve roots/spinal cord).[8]

Extradural benign tumors refer to benign tumors arising from the spine and curvilinear nerves of the body. Benign tumors are rare in the spine, are slow-growing bone and soft tissue tumors that are often found as incidental findings. It rarely grows from the bony cortex and causes spinal pain.[9]

The most common symptom of both benign and malignant spine tumors is back pain. The neurological symptoms that occur are a result of pressure on the spinal cord and radices. The degree of neurological impairment can vary from mild weakness, increased reflexes or paraplegia. About 70% of symptomatic lesions are found in the thoracic region, 20% in the lumbar region and 10% in the cervical region. In intradural or extramedullary tumors paraparesis or plegy occurs more quickly, when compared to extradural tumors.[10] Depending on the extent of the lesion the secondary neurologic deficits varies as sensory abnormalities, mild to total weakness. Pain due to spinal cord tumors can occur if the tumor compresses the radices, often in extradural tumors with manifestations of radicular pain. The association of radicular pain with asymmetric reflexes and insidious onset supports the suspicion of a spinal cord tumor.[2]

Comprehensive bone diagnostic procedures are necessary in cases where metastasis is suspected. Each patient should undergo a thorough clinical examination and a thoracic photograph, and then a photograph of the entire spine. Plain radiographs may reveal erosion of the pedicle and vertebral corpus. This can be clarified by MRI with or without contrast in screening for soft tissue involvement.[11]

MRI is the diagnostic procedure of choice in establishing the diagnosis of spinal cord tumors. The detailed images of the spinal canal and spinal cord in sagittal, axial or coronal sections have displaced other diagnostic procedures as the primary choice.[12] In this case, the T2-weighted MRI results show that a hyperintense rim and central area of low signal (“target sign”) may be seen; this is thought to be due to a dense central area of collagenous stroma. This sign is highly suggestive of neurofibroma, it is occasionally also seen in schwannomas and malignant peripheral nerve sheath tumors.[11]

Extradural benign spinal tumors are infrequent and, due to the limited literature available, diagnostic and treatment algorithms do not yet exist. Despite the heterogeneous appearance of the tumor, the diagnosis of a primary tumor is a particularly dramatic event for each patient.[10] In younger patients, the tumor diagnosis may be more catastrophic, and the required treatment can have a severe impact on the patient's health-related quality of life. In some entity, the pathogenetic activity of the tumor relies on the growth process of the host and is thus conflated with adolescence and younger age.[13]

Surgical resection is an essential part of treatment for most of the lesions and follows the main principles outlined by Enneking's classification. Outcomes of surgical treatment are described by the term Enneking appropriate (EA) or Enneking in-appropriate (EI) instead.[14] For benign primary spinal tumors, intradiscal resection of S2 tumors can be EA, whereas for graded tumors S3 is not considered sufficient. Multidisciplinary surgical planning should therefore also include the histological grade, stage, and extent of the tumor to establish the objective biological aggressiveness of the overlying tumor and in turn estimate the feasibility of resection.[13]

In this case, the patient then underwent tumor removal laminectomy surgery with indications of primary extradural spinal cord tumors with a posterior approach. There has long been controversy regarding the best approach to the cervical spinal cord, but there is no doubt that the clinical indication and the surgical procedure required will determine the best surgical approach for the majority of cases. From a purely anatomical point of view, it seems that the anterior approach is best for partial corpectomy and discectomy and the posterior approach is best for laminectomy, laminoplasty, foraminotomy and the reason is obvious, the anatomical location of the body and the intervertebral disc in the front and the lamina in the back. The choice of approach in the cervical spine should be determined by the location of the primary pathology.[15]

After tumor resection, the sample was examined histopathological, and the results showed tissue consists of large spindle cells with collagen fibers with small spindle cells arranged to form a palisade and no signs of malignancy, conclusion is a neurofibroma.

Spinal neurofibromas most commonly show a location in the thoracic region, followed by a predilection for the cervical and lumbar regions. Neurofibromas located in the sacral region are quite rarely observed and show an asymptomatic course until they grow to a large size.[16] Of this spinal neurofibroma, 72% occur in the intradural extramedullary, 14% in the extradural, and 13% in the intradural and extradural “dumbbell formation”. Only 1% of spinal neurofibromas are intramedullary.[17]

Macroscopically, neurofibroma is gray-white, gelatinous, and soft in shape. It is difficult to macroscopically dissect nerves from neurofibromas due to the close relationship between nerves and neurofibromas.[15] Solitary and plexiform neurofibromas that exhibit similar microscopic characteristics consist of long, thin fusiform cells dispersed among collagen fibers within a matrix rich in mucopolysaccharides.[16] In addition, these cells have a uniform shape and contain hyperchromatic nuclei. Occasionally, some difficulty can be found in separating neurofibromas from schwannomas due to their similar cell contents. Nuclear atypia and hyalinized vascular components observed in neurofibromas are significantly less frequently expressed compared to schwannomas.[15]

In this patient, complete surgical resection was chosen in this case. Yüzbas et al. said early diagnostic and complete surgical resection before the occurrence of severe symptoms will show an excellent prognosis.[18] In this case, after surgery with total removal of the tumor there was a significant improvement

in symptoms. the time taken by the patient in this case was also very fast and assisted with early ambulation. this can also be related to the progressivity of tumor cells, in this case a benign tumor with a border that is not aggressive in invading the surrounding tissue.

4. Conclusion

Early diagnostic and complete surgical resection before the occurrence of severe symptoms will show an excellent prognosis in benign primary extradural tumor. Multidisciplinary surgical planning should therefore also include the histological grade, stage, and extent of the tumor to establish the objective biological aggressiveness of the overlying tumor and in turn estimate the feasibility of resection.

Acknowledgements

None.

Conflict of Interest

The authors declare no conflicts of interest in preparing this article.

References

- [1] Ciftdemir M, Kaya M, Selcuk E, Yalniz E. Tumors of the spine. World J Orthop. 2016 Feb 18;7(2):109-16. <https://doi.org/10.5312/wjo.v7.i2.109>.
- [2] Priyanto, Bambang & Rosyidi, Rohadi & Siradz, Bayu. (2019). Tumor Spinal Intradural Ekstramedula. Unram Medical Journal. 8. 25. 10.29303/jku.v8i1.331.
- [3] Chamberlain, Marc & Tredway, Trent. (2011). Adult Primary Intradural Spinal Cord Tumors: A Review. Current neurology and neuroscience reports. 11. 320-8. 10.1007/s11910-011-0190-2.
- [4] Raj VS, Lofton L. Rehabilitation and treatment of spinal cord tumors. J Spinal Cord Med. 2013 Jan;36(1):4-11. doi:10.1179/2045772312Y.0000000015. PMID: 23433329; PMCID: PMC3555105.
- [5] Arvind V, Nevzati E, Ghaly M, Nasim M, Farshad M, Guggenberger R, Sciubba D, Spiessberger A. Primary extradural tumors of the spinal column: A comprehensive treatment guide for the spine surgeon based on the 5th Edition of the World Health Organization bone and soft-tissue tumor classification. J Craniovertebr Junction Spine. 2021 Oct-Dec;12(4):336-360. doi: 10.4103/jcvjs.jcvjs_115_21. Epub 2021 Dec 11. PMID: 35068816; PMCID: PMC8740815
- [6] American Association of Neurosurgical Surgeons (AANS). Spinal Tumor. <https://www.aans.org/en/Patients/Neurosurgical-Conditions-and-Treatments/Spinal-Tumors>
- [7] Lam FC, Arle JE, Glazer PA, Kasper EM. Primary Extradural Tumors of the Spine - Case Review with Evidence-guided Management. Surg Neurol Int. 2014 Aug 28;5(Suppl 7):S373-5. doi: 10.4103/2152-7806.139673. PMID: 25289164; PMCID: PMC4173213.
- [8] Islam, M.A.; Afreen, M.S.; Montemurro, N.; Chaurasia, B. Surgical Approach for Spinal Tumors: Our Experience in Combined Military Hospital Dhaka. *Surgeries* 2021, 2, 303-307. <https://doi.org/10.3390/surgeries2030030>
- [9] Kumar N, Tan WLB, Wei W, Vellayappan BA. An overview of the tumors affecting the spine-inside to out. Neurooncol Pract. 2020 Nov 18;7(Suppl 1):i10-i17. doi: 10.1093/nop/npaa049. PMID: 33299569; PMCID: PMC7705529.
- [10] Wein S, Bickle I, Todorovic D, et al. Spinal neurofibroma. Reference article, Radiopaedia.org (Accessed on 24 Jun 2023) <https://doi.org/10.53347/rID-19576>
- [11] Lee CS, Jung CH. Metastatic spinal tumor. Asian Spine J. 2012 Mar;6(1):71-87. doi: 10.4184/asj.2012.6.1.71. Epub 2012 Mar 9. PMID: 22439092; PMCID: PMC3302920.
- [12] Chung JY, Lee JJ, Kim HJ, Seo HY. Characterization of magnetic resonance images for spinal cord tumors. Asian Spine J. 2008 Jun;2(1):15-21. doi: 10.4184/asj.2008.2.1.15. Epub 2008 Jun 30. PMID: 20411137; PMCID: PMC2857486.
- [13] Disch AC, Boriani S, Luzzati A, Rhines LD, Fisher CG, Lazary A, Gokaslan ZL, Chou D, Clarke MJ, Fehlings MG, Schaser KD, Gernscheid NM, Reynolds JJ, The Ao Spine Knowledge Forum Tumor. Extradural Primary Malignant Spinal Tumors in a Population Younger than 25 Years: An Ambispective International Multicenter Study on Onco-Surgical Outcomes. Cancers (Basel). 2023 Jan 30;15(3):845. doi: 10.3390/cancers15030845. PMID: 36765803; PMCID: PMC9913243.

- [14] Fisher, Charles & Saravanja, Davor & Dvorak, Marcel & Rampersaud, Raja & Clarkson, Paul & Hurlbert, John & Fox, Richard & Zhang, Hongbin & Lewis, Stephen & Riaz, Salman & Ferguson, Peter & Boyd, Michael. (2011). Surgical Management of Primary Bone Tumors of the Spine: Validation of an Approach to Enhance Cure and Reduce Local Recurrence. *Spine*. 36. 830-6. 10.1097/BRS.0b013e3181e502e5.
- [15] Wael Fouad, Ehab Elzawawy, An anatomical study of the different neurosurgical approaches of the cervical spinal cord, *Alexandria Journal of Medicine*, Volume 47, Issue 1, 2011, Pages 43-51, ISSN 2090-5068, <https://doi.org/10.1016/j.ajme.2011.03.001>.
- [16] Sarica FB, Cekinmez M, Tufan K, Erdogan B, Sen O, Altinors MN. A rare case of massive NF1 with invasion of entire spinal axis by neurofibromas: Case report. *Turkish Neurosurgery*. 2008;18(1):99-106
- [17] Birol Sarica, Feyzi. 'Surgical Principles for Spinal and Paraspinal Neurofibromas'. *Brain and Spinal Tumors - Primary and Secondary*, IntechOpen, 29 Jan. 2020. Crossref, doi:10.5772/intechopen.85760.
- [18] Yüzbaşı M.F., Muçuoğlu A.O., özyörük Ş., Karabekir S., Kaptan H. Case report: cervical extradural schwannoma. *Austin J. Clin. Pathol*. 2018;5(1):1054.