ejpmr, 2021,8(7), 575-579

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

A CASE OF INTRA-EXTRADURAL CERVICAL SCHWANNOMA IN A CHILD WITH NEUROFIBROMATOSIS - TYPE 1

[1]Dr. A. Susmitha, [2]Dr. V. Anurekha, [3]Dr. S. Amudhadevi, [4*]Dr. K. S. Kumaravel, [5]P. Sampathkumar, [6]Dr. L. Sankar

[1]Junior Resident in Pediatrics,
[2]Assistant Professor of Pediatrics,
[3]Assistant Professor of Pediatrics,
[4]Professor of Pediatrics,
[5]Professor of Pediatrics,
[6]Professor of Neurosurgery,
Govt. Mohan Kumaramangalam Medical College, Salem, Tamilnadu, India.

***Corresponding Author: Dr. K. S. Kumaravel**

Professor in Pediatrics, Govt. Mohan Kumaramangalam Medical College, Salem, Tamilnadu, India.

Article Received on 08/05/2021 **Article Revised on 28/05/2021** **Article Accepted on 18/06/2021**

ABSTRACT

Neurofibromatosis type 1 (NF1) is a rare autosomal dominant inherited disorder caused by mutation in NF-1 gene which is located in chromosome 17q11.2. Loss of the neurofibromin, the inactivator of protooncogen RAS, leads to an increase in incidence of tumors. Thus, patients are predisposed to have any central nervous system tumors, especially gliomas, which are the most common neoplasms in NF1. Spinal tumors are relatively rare. Meningiomas and Schwannomas are the two most common intradural extramedullary tumors associated with neurofibromatosis. Schwannoma is benign slow growing nerve sheath tumour. Schwanomas are mostly intradural extramedullary, and upper cervical in location. About 75% of schwannomas are intradural, 10% intra-extradural, and the rest 15% are extradural. We here report a 10-year-old boy with NF-1 presented with weakness of all four limbs and neck pain and was found to have MRI demonstrated intra-extradural mass, suggestive of schwannoma. Intraoperative findings found to be multi lobulated intradural extramedullary tumor extending with extradural component. The diagnosis of schwannoma was confirmed by Histo Pathological Examination which demonstrated Verocay bodies and with Immuno Histo Chemistry studies which demonstrated the presence of strong S-100 proteins. These are benign tumors and complete functional recovery can be obtained after complete excision.

KEYWORDS: Meningiomas and Schwannomas.

INTRODUCTION

Neurofibromatosis type 1 (NF1) is a rare autosomal dominant inherited disorder caused by mutation in NF-1 gene which is located in chromosome 17q11.2. Loss of the neurofibromin, the inactivator of protooncogen RAS, leads to an increase in incidence of tumors. Thus, patients are predisposed to have any central nervous system tumors, especially gliomas, which are the most common neoplasms in NF1. Spinal tumors are relatively rare. Meningiomas and Schwannomas are the two most common intradural extramedullary tumors associated with neurofibromatosis. The overall incidence of spinal tumor is 1.1 per 100,000 populations.[1] Spinal cord tumors are a relatively rare diagnosis in children and they account for about 1 to 10% of all pediatric central nervous system tumors. A large number of developmental tumors like dermoid, epidermoid, and teratomas are common in the pediatric population. Spinal schwannomas are relatively rare in the pediatric population. Schwannomas that arise from the upper cervical spinal roots are more commonly seen than from any other spinal nerves. It has been reported that about 75% of spinal schwannomas are intradural, 10% intra/extradural, and the rest (15%) are extradural.[2]

Intradural and extradural cervical spinal schwannomas are very rarely reported. There are only very few case reports of intra and extradural cervical spinal schwannomas in the pediatric population reported in the literature. Here in this article, we report the clinical and radiological features, surgical approach and intra operative findings, pathological findings, post- operative follow-up and short-term neurological outcome of a high cervical intra/extradural spinal schwannoma in a 10-year-old child presenting with acute quadriparesis and aching neck pain. This case report is presented for its rarity.

CASE HISTORY

A10-year-old male child, born of non-consanguineous marriage and who was developmentally normal was brought to our hospital with complaints of neck pain and weakness of all 4 limbs for the past 10 days. Weakness was symmetrical, insidious in onset, progressive and was associated with wasting of hand muscles. Child also had generalized tonic clonic seizures on admission. In the past history, the child had seizures since 5years of his age and was on anti-epileptic drugs since then. The mother also gave a history of multiple hyper pigmented macules present since birth, which is now increasing in size, pigmentation and in numbers. There was no history of similar illness in their family.

EXAMINATION

On general examination, multiple hyper pigmented macules were seen over the face, trunk and extremities, more than 6 in number, with the largest one measuring 3 X 2cm which were suggestive of café-au-lait macules. There was a large brown hyper pigmented patch over the back extending from nape of the neck extending towards the external border of scapula bilaterally and measuring about 12 cms horizontally (Fig 1). Hair follicles were present over the macule. Multiple firm rubbery swellings in trunk, back, and extremities, suggestive of neurofibromas were seen along the peripheral nerves. Ophthalmic evaluation showed multiple Lisch nodules in both the eyes without any visual impairment. Diagnosis of NF-1 was made based on NIH Consensus conference diagnostic criteria for NF-1 (2).

In the neurological examination, weakness of both upper limbs with a power of 4/5, hypotonia and hyporeflexia were observed. The distal muscles of the upper limbs were more involved with prominent thenar and hypothenar muscle wasting. The lower limbs examination revealed hypertonia, hyperreflexia, positive Babinski sign, clonus, and a power of 3/5 in both lower limbs. The examination of sensory system, cranial nerves, cerebellar functions, autonomic functions, and higher mental functions were normal. In the examination of spine and cranium, cervical spine tenderness was noted from C2 to C6 spinal process. There were no gibbus or neck rigidity.

The hematological and metabolic parameters were normal. The Magnetic Resonance Imaging (MRI) of brain and spine with contrast revealed a well-defined mass measuring 5.2 X 1.8 cms which was T1/T2 heterogeneously hyper intense, associated with spinal canal widening, which was intra-extradural in location, extending from C4 to D1 levels and extending across right neural foramina at these levels (Fig: 2). Lesion shows moderate heterogenous enhancement in contrast study (Fig: 3). Few cystic areas were noted in the mass and the cervical cord was compressed and displaced left laterally. Brain parenchymal imaging was normal.

The child was taken for surgical resection of the tumor electively. With child in prone position, vertical midline incision from C1-D1 was made. Paraspinal muscle was separated from C4-C7 and laminectomy was done. Dura was opened. A multilobulated intradural extramedullary tumor extending from C4-C7 was identified and separated (Fig: 4). The tumor was found to be cystic inferiorly with a grayish yellow solid portion superiorly. The tumor was excised in piecemeal. The tumor involved the C5-C6 nerve roots which were incised and tumor was delivered out. The extradural portion extended into C5 to C6 level. The vertebral portion was excised leaving below a small portion attached to small capsule adjacent to vertebral artery. Hemostasis was attained and the dura closed. The wound closed in layers. The resected tumor was sent for Histo-Pathological Examination and Immuno Histo Chemistry (IHC).

The Histo-pathological Examination of the tumor revealed both hypocellular and hypercellular areas. The hypocellular areas showed sheets of short spindle cells with wavy nuclei and scant indistinct cytoplasm and delicate blood vessels. The hypercellular areas show fascicles and sheets of spindle cells with elongated nucleus, scant indistinct cytoplasm interspersed by Verocay bodies (Fig: 5). Occasional mild nuclear atypia was seen in few areas. There was no mitosis or necrosis. In the IHC, S-100 was strongly positive (Fig: 6).

Post operatively the child was on mechanical ventilator for 5 days after which he was gradually weaned from the ventilator. He was found to have gradual improvement in the weakness of lower limbs and upper limbs. He was advised for regular follow up and discharged after 3 weeks.

DISCUSSION

Intra/extradural cervical spinal schwannoma is a rare disease. This case report describes the case history and review of literature of cervical spinal schwannoma. Based on anatomic location, the cervical tumors can be divided into two - Extradural tumors are present between the bony structures and the dura and Intradural tumors, which are again subdivided into extra- medullary and intramedullary.[1] Extramedullary tumors are present inside the dura but they are not part of the spinal cord whereas intramedullary tumors are within the spinal cord parenchyma. Multilobulated schwannomas have both intraspinal and paraspinal components which are connected through eroded intervertebral foramen. Though the extraspinal component of the schwannomas is larger than the intraspinal tumor, the intraspinal tumor is responsible for the typical symptoms of spinal cord compression in the cervical region.[3]

The differential diagnosis of spinal tumors in children includes schwannoma, neurofibroma and meningiomas. About 30% of all the primary spinal cord tumors are spinal schwannomas.[4] Around 70% of the spinal schwannomas arise from the sensory roots, 20% arise

from the motor roots and another 10% are known to arise from both the motor and sensory roots.[2] Among the cervical spinal schwannomas, the most common tumor arises from C2 spinal nerve root. It constitutes to 15% of all spinal schwannomas. Multiple schwannomas are usually seen in type 2 neuro- fibromatosis. Multiple schwannomas are seen in about 4% of patients in the spinal cord.

Symptoms and signs of this condition include radicular pain, swelling in the neck, and motor weakness.[2] These are known to be slow growing tumors and will usually attain a large size before becoming more symptomatic. Also, sometimes they can present with unusual symptoms like, headaches, syncopal attacks and unrelated motor and sensory symptoms.

Most schwannomas are moderately vascular and their consistency is usually firm. On histopathologic examination, the schwannomas characteristically have 2 patterns- Antoni A and Antoni B. Type A tissue is usually cellular and demonstrates nuclear palisading and the characteristic Verocay bodies. The Verocay body is characterized by prominent extracellular matrix and secretion of the laminin. The Antoni type B pattern is a loosely organized tissue with cystic and myxomatous changes which may also represent the degenerated Antoni A tissue.[5] Schwannomas are relatively simple to resect.[6,7] For dissection, they have a well-defined arachnoid plane intra- durally and well-defined capsule extradurally. For rapid and complete neurological recovery, the total tumor is to be resected.

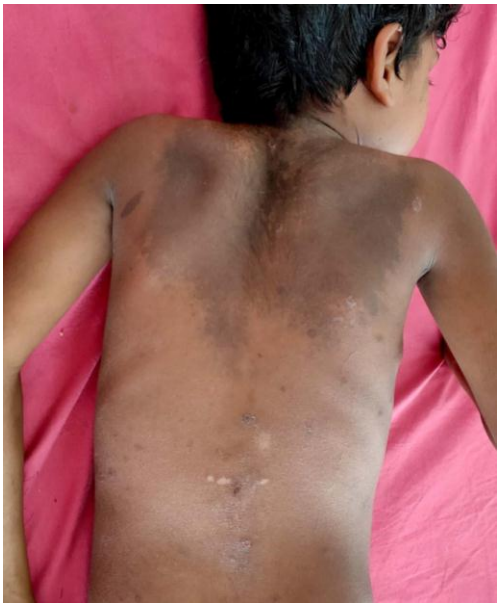

Fig: 1 Large Café-au-lait spot on the back.

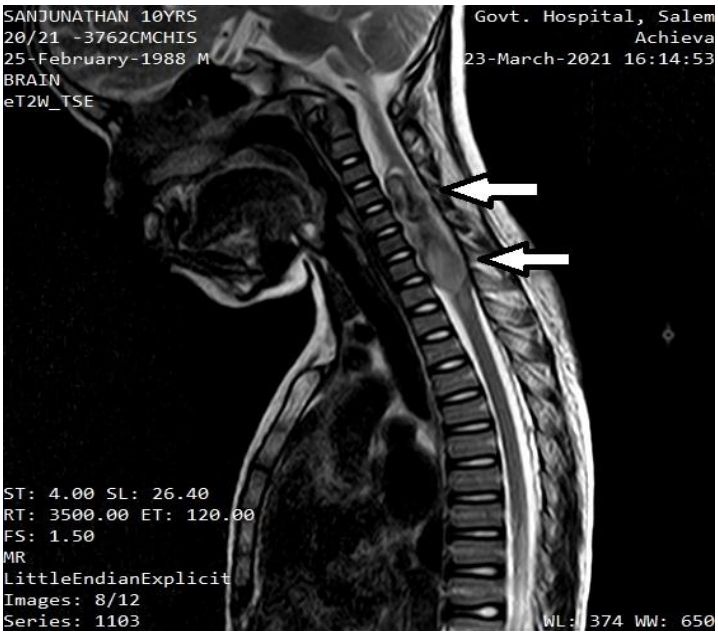

Fig: 2 T2W MRI image showing the lesion extending from C4 to D1 vertebral level.

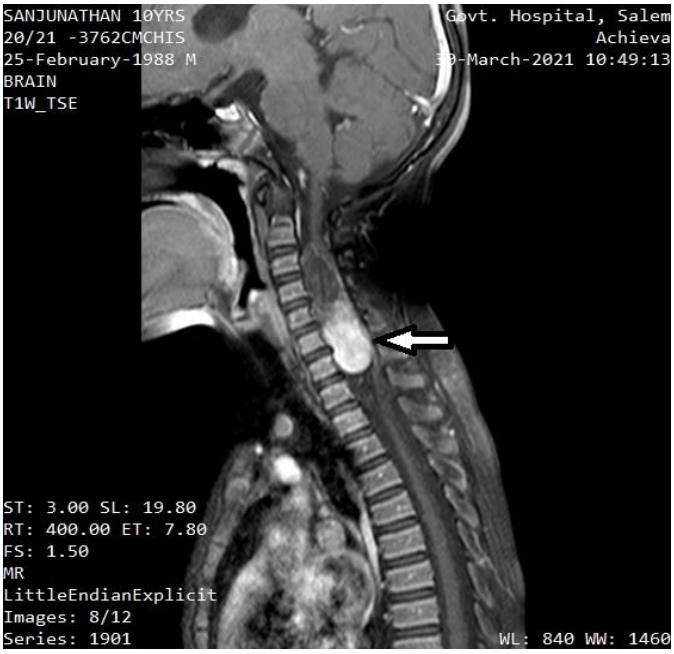

Fig: 3 Contrast MRI image showing the enhancement of the lesion with contrast.

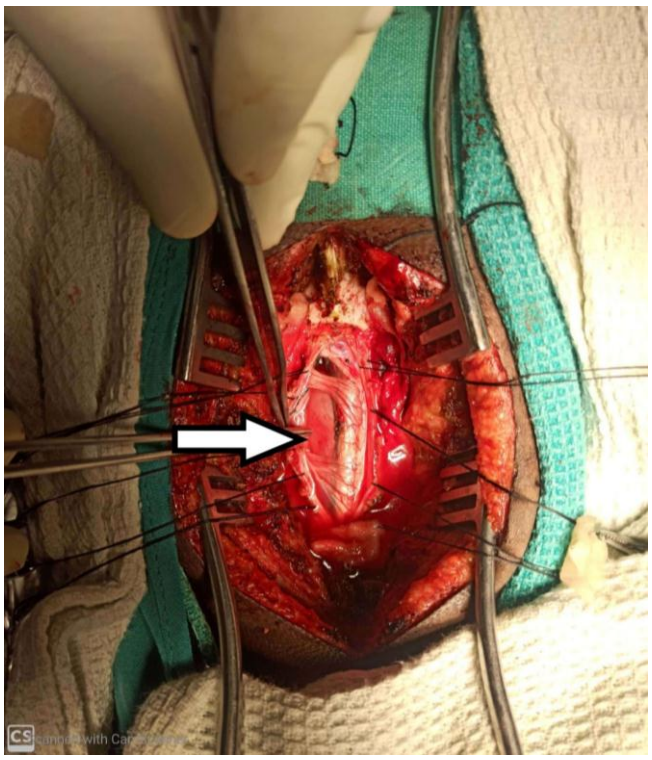

Fig: 4 Intra operative picture showing the multilobulated schwannoma.

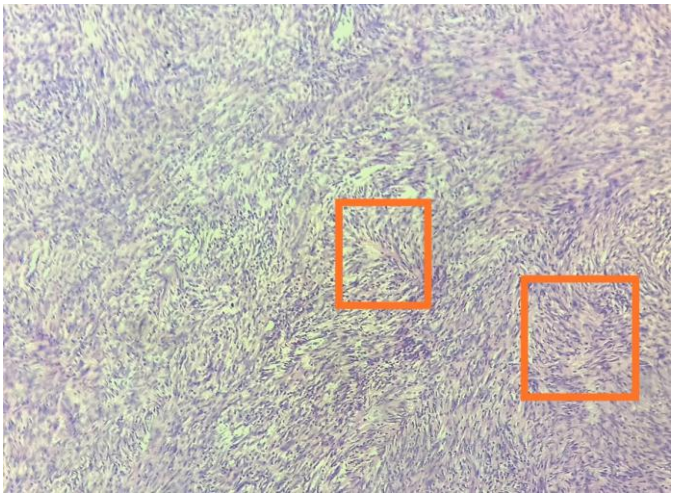

Fig: 5 Histopathological examination showing the fascicles and sheets of spindle cells with elongated nucleus, scant indistinct cytoplasm interspersed by Verocay bodies.

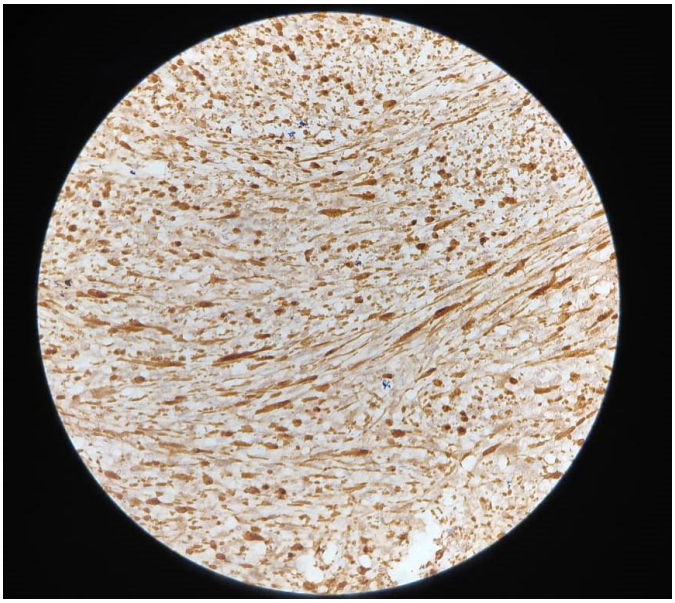

Fig: 6 Immunohistochemistry demonstrating a strongly positive S-100.

CONCLUSION

This case report describes a rare case Intra-Extradural cervical spinal schwannoma in a child with NF-1 in the pediatric population. These are benign tumors and complete functional recovery can be obtained after complete excision.

ACKNOWLEDGEMENT

We sincerely thank the Departments of Pathology and Radio-diagnosis for their immense help and support in the management of this child.

REFERENCES

1. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, JouvetA, etal. The 2007 WHO Classification of Tumours of the Central Nervous System. Acta Neuropathol, 2007; 114(2): 97–109.
2. Neurofibromatosis. NIH Consens Statement Online 1987 Jul 13-15 [cited 25/05/2021]; 6(12): 1-19.
3. Levy WJ, Latchaw J, Hahn JF, Sawhny B, Bay J, Dohn DF. Spinal neurofibromas: a report of 66 cases and a comparison with meningiomas. Neurosurgery, 1986; 18(3): 331–4.
4. Stiller CA, Nectoux J. International incidence of childhood brain and spinaltumours. Int J Epidemiol, 1994; 23(3): 458–64.
5. Donner TR, Voorhies RM, Kline DG. Neural sheath tumors of major nerves. J Neurosurg, 1994; 81(3): 362–73.
6. Hardison HH, Packer RJ, Rorke LB, Schut L, Sutton LN, Bruce DA. Outcome of children with primary intramedullary spinal cord tumors. Childs Nerv Syst., 1987; 3(2): 89–92.
7. Nadkarni TD, Rekate HL. Pediatric intramedullary spinal cord tumors. Critical review of the literature. Childs Nerv Syst., 1999; 15(1): 17–28.