

Intradural Extramedullary Neurilemmoma of Spinal Cord

Khurram Ayub and Naim-ur-Rehman

Department of Neurosurgery, Shaikh Zayed Hospital, Lahore

SUMMARY

A case of spinal cord tumour presenting with paraplegia is reported. Tumour was completely removed and Out-patient review a month later showed her to be asymptomatic. Literature review also includes discussion on diagnosis and treatment of spinal cord tumours. Lastly, suggestions for diagnosis and management of these tumours have been given.

INTRODUCTION

Paraplegia is usually a depressing situation, but when it is caused by Benign Intradural tumours, the message is one of hope rather than despair. Spinal cord tumours have been written about for many centuries, but removal of an intradural extramedullary tumour was first reported almost hundred years from now in 1888, by Sir William Gowers and Sir Victor Horsley. "At the National Hospital, Queen Square, London, Sir Victor Horsley performed a Laminectomy on an army officer with extensor spastic paraplegia. The patient had been treated by Sir William Gowers, who had diagnosed a spinal tumour and advised surgery. In the event no tumour was found and the operation was about to be concluded when Mr. Charles Ballance who was assisting urged Horsley to extend the Laminectomy to a higher level. On removal of another lamina a clearly circumscribed tumour was found and removed. For the first time in the history of medicine a patient operated on for a spinal tumour recovered from the operation completely and gradually resumed his previous occupation".

CASE REPORT

A 45 years old female presented in Casualty with complaints of sudden weakness of both legs and excruciating bilateral sciatica following Myelography. She was previously ambulatory and could walk with some help prior to Myelography. She had presented in orthopaedics out-patient with

pain in the back for past three years shooting from hip to back of thigh on left side and in front of thigh on right side. On examination she was a middle aged female shrieking with pain. Her both hips and knees were in flexion due to muscular spasm. She was unable to stand or walk. Weakness of all the muscle groups in both legs was noticed. Both knee and ankle jerks were absent bilaterally. Patchy anaesthesia was found below the groin. Myelographic findings were classical of intradural extramedullary tumour. Myelogram done via Lumbar route showed an almost complete block at the level of 2nd Lumbar vertebra. Crescentic margin of the contrast outlined the lower and upper poles of the tumour. Cauda equina was pushed to one side (Fig. 1 a and b). Patient was advised urgent surgery. Complete laminectomy of L1-L3 was carried out. Dural tube was expanded. On incision of the dura a well defined tumour that was compressing the Cauda Equina was found and completely removed (Fig. 2). On Histopathology it proved to be Neurilemmoma (Fig. 3). Post-operatively patient was completely pain free and by 10th Post-operative day patient started walking about and was discharged two weeks after surgery. Outpatient review a month later showed her to be completely asymptomatic.

LITERATURE REVIEW

In 1976 Nittner's review of large number of series suggested that approximately one fifth of C.N.S. tumours occur in spinal cord. Extradural Neoplasms of spinal cord are about 20% whereas 80% are Intradural. 20% of the intradural variety are

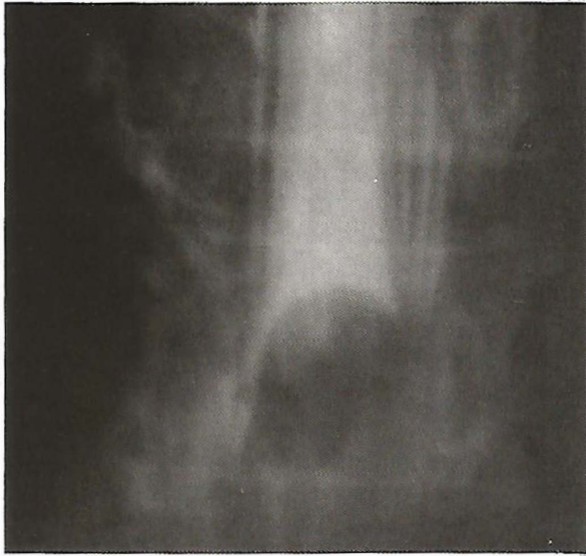


Fig. 1a: AP view Myelogram done via lumbar route showing an almost complete block at the level of second lumbar vertebrae. Crescentic margin of the contrast outlines the lower pole. Cauda equina pushed to one side are outlined on the left side.

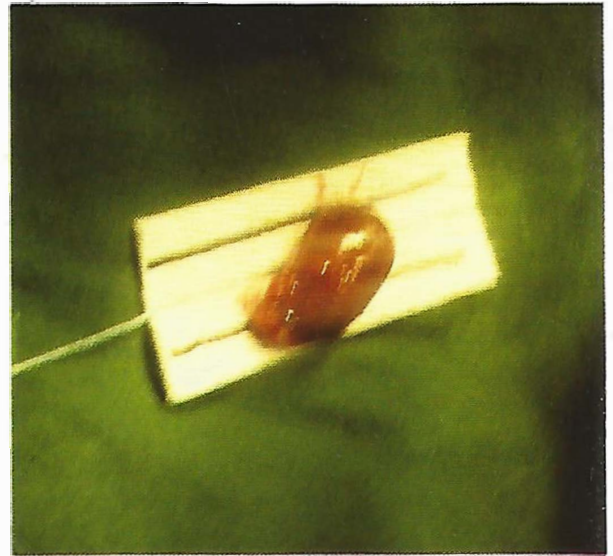


Fig. 2: Sausage shaped tumour removed completely shows against a 2.5 cm X 5 cm Neurosurgical patties.



Fig. 1b: Lateral Myelogram. Small amount of contrast has leaked alongside the tumour and is visible above the upper level of the block.

intramedullary and 80% extramedullary. Spinal tumours are cross-classified according to their relationship to the dura and the cord and by Histological types. In Nittner's review of 4885 spinal cord tumours 1129 were found to be Neurilemmomas i.e. about one quarter of all spinal tumours. A Mayo clinic series of 1322 spinal

tumours showed 29% incidence of Neurilemmomas. 72% Neurilemmomas are located in intradural extramedullary space, 14% are extradural, 13% are dumbbell and 1% are intramedullary. Most frequently they occur in thoracic, less frequently in cervical and least often in the lumbar region. They have no particular sex predilection and occur primarily in fourth and fifth decade^{1,2}.

Diagnosis is based on clinical features, investigations and histopathology. Clinical feature are due to spinal compression causing a transverse spinal lesion. In 1923 Oppenheim classified the three stages of spinal cord compression. First stage is characterized by root pain and segmental sensory or motor disturbances. Second stage is of incomplete transection or Brown-Sequard syndrome and third stage is of complete transection. Slow growing tumours such as Neurilemmomas and Meningiomas usually produce spastic paresis as compared to flaccid paresis in case of rapidly progressive tumours. Compression of the dorsal surface of the cord is more common in Neurilemmomas as compared to Meningiomas. This involves all modalities of superficial sensations including position & vibration. Tumours of the conus medullaris and cauda equina are distinguished by early loss of bladder and bowel control with symmetrical saddle anaesthesia. Pain occurs late in conus lesions whereas cauda equina lesions produce pain early and sphincter involvement late^{2,3}.



Fig. 3a: Compact Mass of Antoni-A tissue showing nuclear palisades, characteristic of Neurolemmoma.

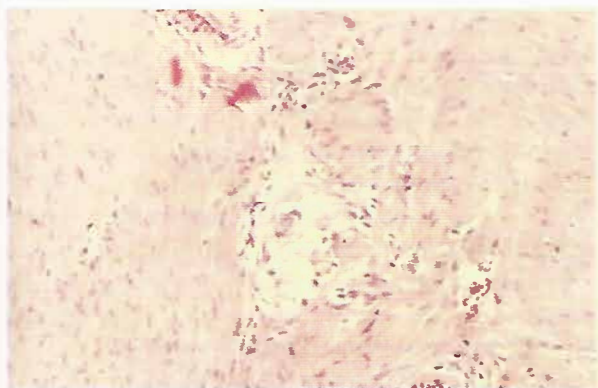


Fig. 3b: Partially thrombosed blood vessels surrounded by loose myxoid Antoni-B and cellular Antoni-A areas.

The first special diagnostic test performed for suspected spinal tumour should be plain spine X-Rays. Most common finding is widening of interpedicular distance. Thinning & loss of pedicle may be seen. A spinal tap should not be performed before myelography as it may precipitate a cord shift with tumour incarceration and consequent rapid deterioration requiring an emergency operative

procedure before the lesion has been adequately localized. It occurred in three patients in series reported by B. Guidetti & is well illustrated by our case^{1,2}. Therefore it is a standard practice that Lumbar puncture & Myelogram are to be done on Inpatient basis. A high protein level upto 5 gm (5000mg) per 100 ml with Xanthochromia, but no cells is the result of Mechanical Block. It is known as FROIN's syndrome. A dry tap may occur if the needle enters a tumour filling the lumbar sac. Myelogram alone demonstrates a spinal block, showing exactly its level and its degree of completion. It is possible to deduce whether the mass is inside or outside the dura. Upper & Lower poles of the tumour shows a crescent of contrast at the level of the block. Triangle of contrast capping the tumour poles is pathognomonic of intradural extramedullary tumours. These findings are beautifully illustrated in the Myelogram of our patient (Fig. 1 a & b). Spinal angiography, radioisotope scanning and Electromyography are of less value. Spinal computerized tomography following metrizamide injection is helpful in precise localization of the tumour in relation to the cord^{1,2,3}.

Treatment is total excision. Best results are with the least Neurological deficit before operation. But recovery has been noted in some cases even at complete transection stage, provided urgent decompression is carried out. Operative mortality is from 0% to 7% at various centres. Radiation therapy is of no value, neither is chemotherapy^{1,2}.

CONCLUSION

In last 100 years great advances have been made in spinal surgery. Dandy introduced Air Myelography in 1919. Sicard and Forestier introduced positive contrast Myelography in 1921. Greenwood introduced Bipolar coagulation in 1940. Kurze introduced operative Microscope in 1964². The latest refinement is "CAVITRON" — the ultrasonic aspirator. All these modalities have considerably reduced the morbidity & mortality and made it possible to safely excise both intramedullary & extramedullary tumours. It should be emphasized that all cases of suspected spinal cord compression should have myelogram only after admission with facilities for immediate Neurosurgical intervention, should a myelographic block be found. Initial recovery of functions after removal of benign

extramedullary tumour is usually dramatic as was seen in this case. But the final stages of improvement are slow. In case of cauda equina lesion it may take as long as two years. This fact should be impressed upon the patient and is to be kept in the mind of Physiotherapist. Recovery of Neurological deficit is usually rapid & complete if surgery is carried out early but can be delayed and incomplete if treatment is delayed.

"Case discussed in Clinicopathological Conference dated August, 25, 1988."

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The Authors:

Khurram Ayub,
Medical Officer,
Department of Neurosurgery,
Shaikh Zayed Hospital,
Lahore.

Naim-ur-Rehman
Professor & Head of Department of Neurosurgery,
Shaikh Zayed Hospital,
Lahore.

Address for Correspondence:

Khurram Ayub,
Medical Officer,
Department of Neurosurgery,
Shaikh Zayed Hospital,
Lahore.