Afr. J. Med. med. Sci. (1976) 5, 101–103.

Tumour of the sciatic nerve

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Summary

A malignant neurilemmoma of the sciatic nerve is described. The requisites for making this diagnosis are discussed. It is pointed out that such tumours are commonly misdiagnosed as lumbar intervertebral discs and that local examination of the leg along the course of the sciatic nerve should be carried out on every patient complaining of sciatica.

Résumé

Un neurilome malin du nerf sciatique est décrit. Les motifs du diagnostic sont discutés. Il est souligné que ces tumeurs sont souvent confondues avec les disques intervertébraux lombaires et qu'il convient d'effectuer, sur tout malade se plaignant de sciatique, un examen de la jambe le long du nerf sciatique.

A prolapsed intervertebral disc is generally considered to be the commonest cause of sciatica. In these 'back conscious' days, pain in the leg is usually attributed to a lesion of the lumbar spine and one may tend to overlook local examination of the leg itself. Tumours of the sciatic nerve, beginning as small nodules, are therefore likely to be missed, even though causing severe symptoms (Armstrong, 1965). We would like to present a case of malignant tumour of the sciatic nerve, whose presence was not initially suspected.

Case report

A 19-year-old boy presented with a 2 years history of pain and weakness in the right leg. His first symptoms had been a burning pain in the right big toe, and this had spread slowly up the limb over the course of the following year. The pain was continuous, not aggravated by coughing or movement and not relieved by rest. He had no lumbar pain or spincter disturbance. There was no history of trauma or previous ill-health.

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Examination

He was a very tall thin boy who walked with a limp caused by foot-drop. There were no stigmata of neurofibromatosis. Abnormal physical signs were confined to the right lower limb. The right leg was grossly wasted (right calf 28 cm circumference; left calf 33·6 cm) although the thigh was less affected (right thigh 34 cm; left thigh 35 cm). No muscle fasciculation was seen. Wasting of the quadriceps muscle caused mild weakness of hip flexion but severe weakness of knee flexion. The right knee jerk was brisk; the right ankle jerk absent. Neither plantar response was obtainable. Straight leg raising was restricted to 60° on the right side compared to 90° on the left side. There was complete spino-thalamic sensory loss over the L5 and S1 dermatomes of the right leg, and partial loss over the sacral dermatomes of the right buttock.

Spinal movements were normal. There was no spinal tenderness but the region of the right sacro-iliac joint was tender on palpation.

Investigations

Routine haematological and urine examinations were normal. His ESR was 11 mm/h and a Heaf test was negative. Radiological examination of the chest, lumbar spine, sacro-iliac and hip joints was normal. Myelography failed because the dye was injected into the subdural space. The CSF obtained after a blood tap contained 110 mg/100 ml protein and 150 WBC, mainly lymphocytes.

First operation (6 *May* 1970)

Laminectomy was carried out and the L3/4, L4/5 and L5/S1 disc spaces explored. The annulus of the L3/4 disc was rather prominent and this disc was therefore removed. The dura was opened but no intradural pathology was found, and a catheter passed easily along the sub-arachnoid space.

Post-operative progress. His pain was not relieved. Repeated examination showed a point just below the right buttock where pressure caused a shooting pain along the course of the sciatic nerve. It was therefore decided to explore the sciatic nerve.

Second operation (26 *June* 1970)

The approach of Henry (1952), to the sciatic nerve was used. An incision was made from the posterior superior iliac spine along the iliac crest for 6 cm, then downwards to the greater trochanter of the femur, across the lower margin of the buttock fold and then descending in the posterior midline of the thigh. The sciatic nerve at its branch to gluteus maximus was thickened and infiltrated with tumour over a distance of 3 cm. The perineurium separated easily from the nerve fibres and there was no tumour involvement of the perineural tissues. No response was obtained to nerve stimulation so a biopsy of the tumour was taken.

Histological examination. The tumour consisted of round and spindle cells. There was great nuclear pleomorphism and necrosis with a high mitotic rate. It was thought to be a neural crest tumour with appearances midway between neuroblastoma and neurofibrosarcoma.

Post-operative progress. The pain was temporarily relieved. Repeated examination showed that there was no function in the sciatic nerve below the level of the tumour. As the tumour appeared to be entirely confined to the nerve, its malignant histological picture prompted us to attempt tumour removal.

Third operation (3 *August* 1970)

The previous wound was re-opened. The tumour still looked confined to the nerve although there were adhesions to the gluteus medius muscle at the site of the former biopsy. The nerve was divided as high as possible where it passed through the greater sciatic notch and an 18 cm long segment of nerve was excised together with the branches to gluteus maximus, gluteus medius, pyriform, the gemelli and hamstring muscles.

Second histological examination. The tumour was thought to be anaplastic, possibly an histocytic lymphoma or leukaemic. The tumour cells located mainly around the periphery of each neuro-axial bundle. Despite the wide excision, one compartment at the lower extremity of the nerve section was found to be infiltrated with tumour, but the proximal end of the nerve was tumour free.

Post-operative progress. Bone marrow examination excluded a diagnosis of leukaemia but the CSF examination now revealed the presence of 7 malignant cells/mm^3. The protein level was 20 mg/100 ml. It was our intention to give him a course of Cyclophosphamide but this was postponed when he developed a pyrexia and wound infection. The wound broke down and was re-sutured. He developed persistent severe pain in the right leg and scrotal swelling due to epididymitis. This responded to antibiotics and the swelling of the scrotum resolved. On 18 November 1970 he was given 5 ml of intrathecal Phenol in Myodil in an effort to control the pain in the right leg. He had temporary retention of urine and required catheterization for 9 days. It was then discovered that he had bilateral sacral anaesthesia, but we were not sure whether this was the result of the Phenol in Myodil injection or progression of his original tumour. From 24 November 1970 onwards he began to have a high swinging fever. Investigations showed a urinary infection due to proteus, but despite appropriate antibiotic therapy he continued to have high pyrexia and his general condition slowly deteriorated. The right leg became swollen and oedaematous and deep venous thrombosis was suspected. This abated spontaneously. X-rays of the right leg and chest did not show any abnormality. His downhill progress continued and he died on 17 February 1971. The relatives unfortunately refused us permission to carry out an autopsy.

Discussion

Tumours of the peripheral nerves have always caused a lot of confusion when it comes to classifying them. The classification of Willis (1967) into four

groups is widely accepted. These are: (a), neurilemmoma (Schwannoma); (b), solitary neurofibroma; (c), multiple neurofibromatosis (von Recklinghausen); (d), malignant neurofibroma.

The terminology of malignant nerve tumours is even more confusing and no fewer than fifteen synonyms are mentioned by Harkin & Reed (1969).

The malignant nature of the tumour in our case was not doubted. It was difficult to be very specific about its nature. It was thought to be a neural crest tumour and this suggested that it was a malignant neurilemmoma (Schwannoma) as it is believed that the sheath of Schwann is a derivative of neuroectodermal cells of the neural crest (Harrison, 1924).

It is often difficult for a histo-pathologist to distinguish a malignant nerve tumour from other fibrosarcomas which may surround and even invade a peripheral nerve (Russell & Rubenstein, 1959). It must be proved that the tumour has arisen from nerve tissue (Vieta & Pack, 1951). D'Agostino, Soule & Miller (1963) therefore laid down certain criteria to be met before a diagnosis of malignant neurilemmoma was established. These are: (a), the tumour should be a circumscribed swelling of the nerve; (b), the nerve fascicles should be splayed out or intertwined within the fusiform mass: (c), there should be minimal extension beyond the epineurium; (d), there should be histological evidence of malignancy. These requisites appear to be fulfilled in our case.

On review of the literature, one finds that benign and malignant neurilemmomas tend to favour different sites. Thus malignant neurilemmomas are commonly found arising from the sciatic nerve, a site not considered common for a benign tumour. D'Agostino *et al.* (1963) in a review of twenty-four solitary malignant neurilemmomas found ten cases arising from the sciatic nerve.

Malignant neurilemmoma may arise with or without generalized neurofibromatosis, and does not necessarily mean that malignant change has occurred in a benign neurilemmoma (Stout, 1948; Vieta & Pack, 1951).

Misdiagnosis of sciatic nerve tumours is commonly reported and patients may be investigated by X-rays of the spine, pelvis and hips, by myelography and even by spinal exploration before the true diagnosis is made. Five cases of D'Agostino *et al.* (1963) were subjected, like our patient, to laminectomy with a presumptive diagnosis of prolapsed intervertebral disc. Palpation along the course of the sciatic nerve is mandatory, especially if sciatica occurs without low back pain.

A high rate of local recurrence of the tumour is reported even after radical local excision (D'Agostino *et al.*, 1963). Vieta & Pack (1951) say that only amputation or radical local excision give any hope of cure. They report a 30% 5 year survival rate, but note that in seven of their twenty cases treated by amputation metastatic spread to the lungs occurred. The finding of malignant cells in the lumbar CSF and infiltration beyond the distal level of nerve excision in our case shows that this tumour can spread over long distances within the nerve sheath even though macroscopically it appeared to be a localized tumour.

Acknowledgments

Our thanks are due to Mr Donald Gibson, FRCS, and to the Chief Medical Officer, Uganda, Ministry of Health for permission to publish this paper.

References

ARMSTRONG, J.R. (1965) *Lumbar Disc Lesions, Pathogenesis and Treatment of Low Back Pain and Sciatica*, p. 201. Livingstone, Edinburgh and London.

D'AGOSTINO, A.N., SOULE, E.H. & MILLER, R.H. (1963) Primary malignant neoplasms of nerves (malignant neurilemmomas) in patients without manifestations of multiple neurofibromatosis (von Recklinghausen's disease). *Cancer*, **16**, 1003–1014.

HARKIN, J.C. & REED, R.J. (1969). *Tumours of the Peripheral Nervous System*, p. 71. Armed Forces Institute of Pathology, Washington.

HARRISON, R.G. (1924) Neuroblast versus sheath cell in the development of peripheral nerves. *J. Comp. Neurol.* **37**, 123–194.

HENRY, A.K. (1952) *Extensile Exposure Applied to Limb Surgery*. Livingstone, Edinburgh and London.

RUSSELL, D.S. & RUBENSTEIN, L.J. (1959) *Pathology of Tumours of the Nervous System*. Edward Arnold, London.

STOUT, A.P. (1948) Fibro-sarcoma: malignant tumour of fibroblasts. *Cancer*, **1**, 30–63.

VIETA, J.O. & PACK, G.T. (1951) Malignant neurilemmomas of peripheral nerves. *Amer. J. Surg.* **82**, 416–431.

WILLIS, R.A. (1967) *Pathology of Tumours*, 4th edn. Butterworths, London.

(*Received* 21 *April* 1972)