

EXTRA SPINAL EPENDYMOMA

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Most of the spinal ependymomas are confirmed within the dura and become clinically evident by compression of the spinal cord and nerve roots of the cauda equine¹. Rarely an ependymoma may arise in the bone or soft tissues of the pelvis or sacral subcutaneum²⁻⁴. We report a case of extraspinal ependymoma involving the lumbosacral region of the spine.

CASE REPORT

A 27-year old man admitted with a history of low grade back pain of six years duration. The pain gradually got worse. A year later, the pain started radiating down both legs to the outer aspect of the ankles, and it was constant and throbbing in nature. There were periods when the pain got slightly better, but he was never free of pain. There were no sensory changes. He had been taking non-steroidal anti-inflammatory analgesics for pain. Examination of the back showed no visible deformity. On flexion, he could reach just above the knee by the tip of his fingers. Straight leg raising was 60° bilaterally. Knee and ankle reflexes were present and normal. Plantar reflexes were normal. His right extensor hallucis longus was weak (Grade IV), and he had a wasted right quadriceps by 1 cm difference. Tomogram (Figure 1)

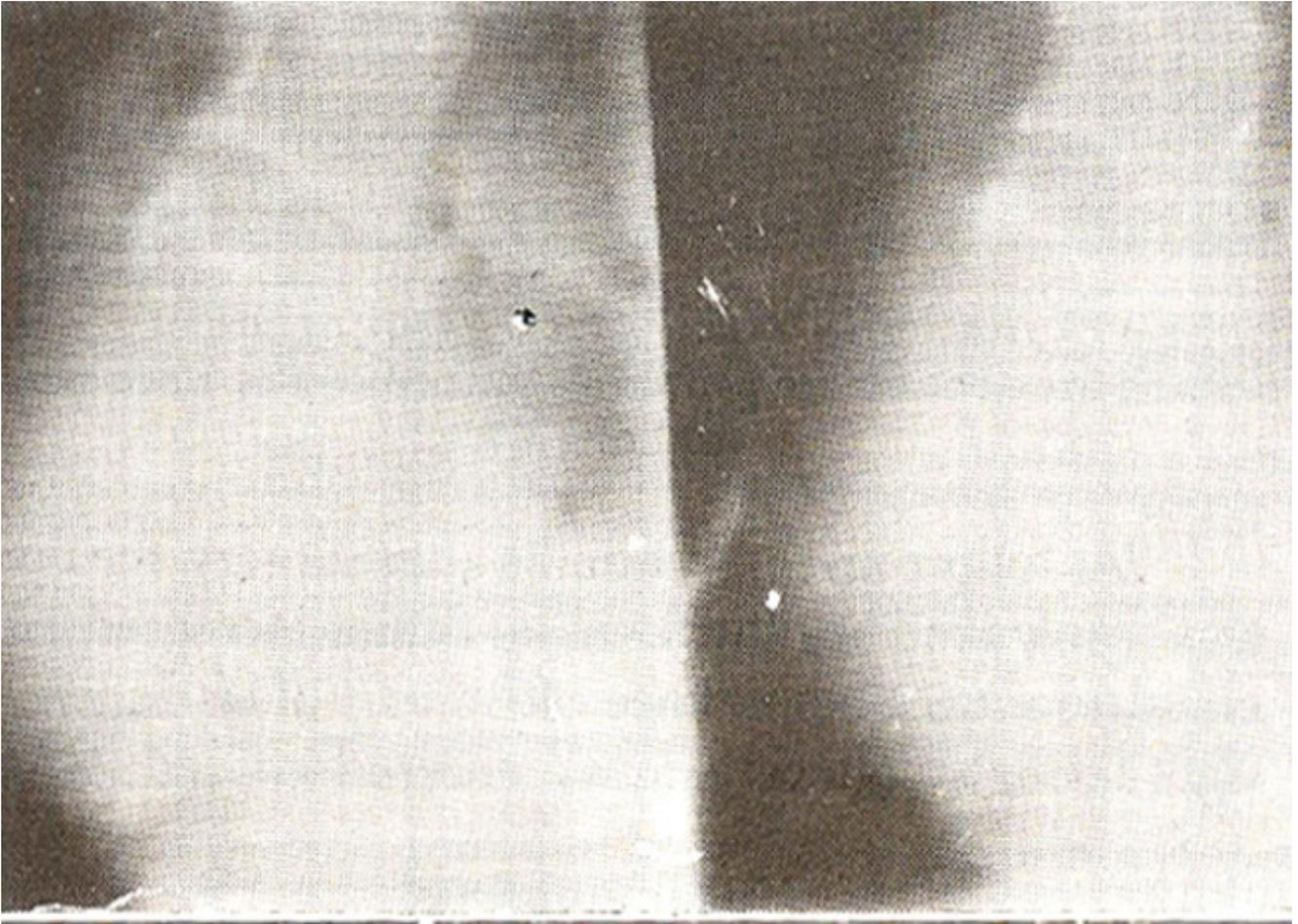


Figure 1. Tomogram-lateral view showing complete destruction of sacrum and distal two thirds of L5 by tumour.

showed extensive destructive lesion of the sacrum and L5 with involvement of soft tissue as well. CT scan (Figure 2)

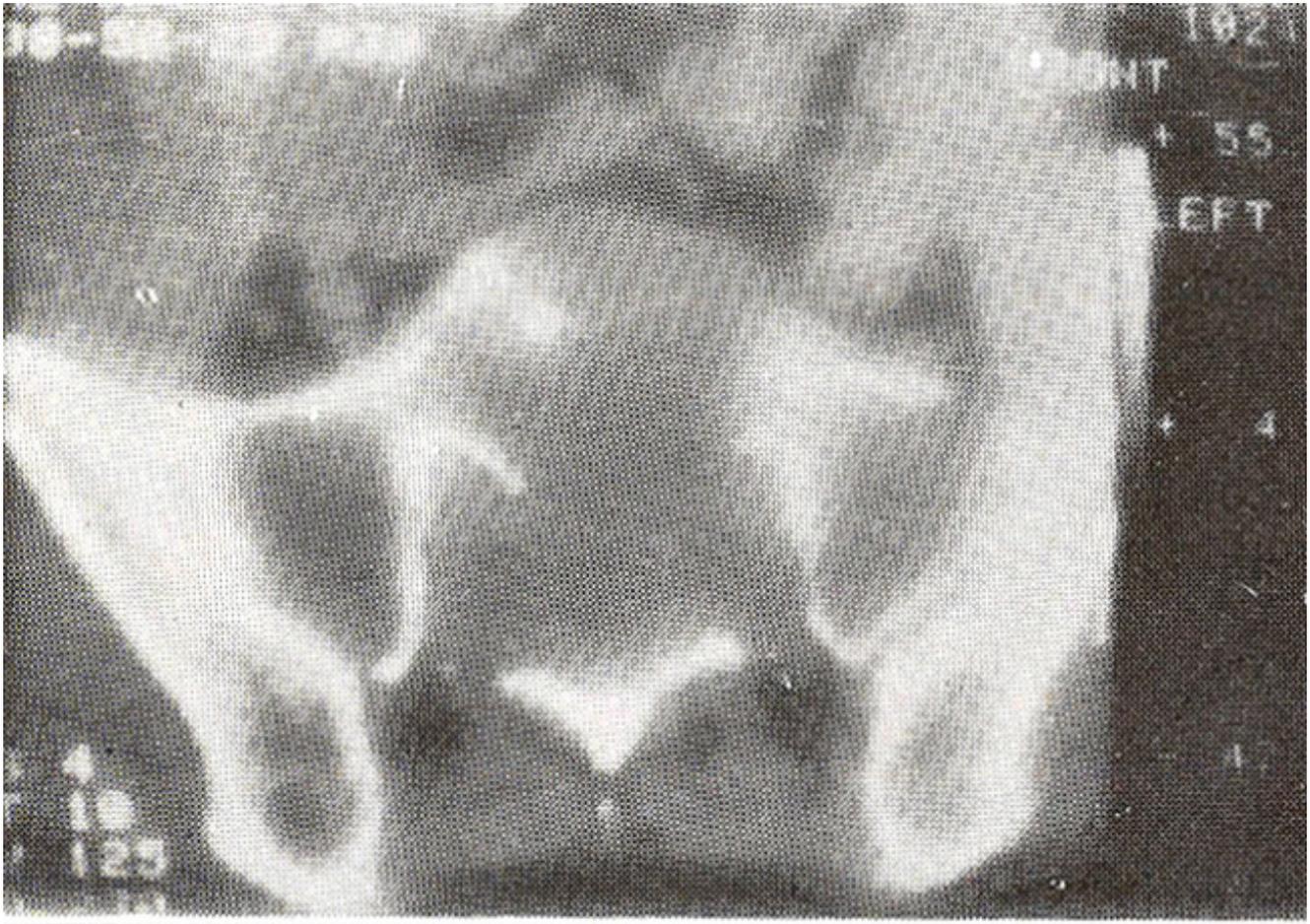


Figure 2. CT Scan showing tumour mass in sacral region invading sacrum which is largely destroyed.

confirmed the above findings. Blood investigations revealed an ESR of 40mm and a negative brucella titre. At surgery, the lumbosacral region was approached and a fungating mass was seen at the posterior edge of L1. Deroofing of the sacral spinal canal was made and laminectomy of L5 and part of L1 was done. The mass was dark brown in colour and very friable, but not vascular, it extended into the neural canal above L1 and down into the sacrum. The mass was extradural, pushing the nerve roots laterally and anteriorly. All the accessible mass was scooped out. The histological examination of the tissue revealed a typical cellular ependymoma. Characteristic zones of high nuclear density and perivascular pseudorosettes were seen. However, true rosettes or papillary structures were not found. The tumour showed a positive staining with glial fibrillary acidic protein by the immunoperoxidase technique (Figure 3).



Figure 3. Histological section of tumour showing tumour cells with round to oval nuclei and perivascular pseudorosettes. Haematoxylin and Eosin X 100.

At follow-up three months after surgery, the operation scar looked healthy. Lumbar flexion increased and he could now reach below knee with the tip of his fingers. His right extensor hallucis longus recovered to Grade V.

DISCUSSION

Extrasacral ependymoma is an extremely rare tumour. Mark and Loken⁵ studied 101 patients with histologically confirmed ependymomas over a 22-year period, of these, 48 were intracranial and 53 intraspinal. There was no example of an extrasacral ependymoma in their study. However, Kernohan and Fletcher- Kernohan⁶ reported three pre-sacral ependymomas collected over 26 years in a series of 55 ependymomas, giving a frequency of approximately 5%. Mallory⁷ was the first to describe an extrasacral ependymoma in 1902. By 1979, approximately 40 such cases had been reported in world literature³. The origin of the extrasacral ependymomas is disputed but most workers believe that the origin of these tumours is from "heterotopic" ependymal cells located within this region^{2,6,9}. Extrasacral ependymomas present at two characteristic locations: in the soft tissue posterior to the sacrum or in the pelvis anterior to the sacrum in the retro-rectal space³. Those in whom the tumour arose posterior to the sacrum presented with a local mass which was occasionally painful while those

whose tumour was pelvic in location presented predominantly with bowel and bladder dysfunction. The extrasacral ependymoma, being a rare lesion, is often mistaken for other inflammatory, congenital, neurogenic and osseous lesions in this area, like tuberculosis, pionicidal cyst, neurofibroma and chordoma. Histopathological examination is the only way to confirm the diagnosis. In contrast to most of the reported cases in which the ependymomas were either papillary or myxopapillary, our case revealed a typical cellular ependymoma, with no papillary structures. The average survival time for the group of patients on whom information is available is approximately ten years³. Postoperative survival tends to be long. These tumours characteristically recur locally. Systemic metastases have been noted in several reported cases^{3,9-11}. Early diagnosis is, therefore, important. Complete surgical removal should be attempted in all cases. If not feasible, then radiation therapy should be employed³.

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