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Primitive neuroectodermal tumour of the cauda equina

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Abstract. Primary primitive neuroectodermal tumours of the cauda equina are rare. We report a case and review the literature.

Key words: Primitive neuroectodermal tumour – Cauda equina

Primitive neuroectodermal tumours (PNET) are highly cellular central nervous system (CNS) neoplasms which usually occur in children. They are commonest in the posterior cranial fossa where they are conventionally known as medulloblastomas. Only five cases of primary PNET of the cauda equina have been reported [1–3]. We report a primary PNET of the cauda equina in a 47-year-old man.

Case report

A 47-year-old man presented with an 18 month history of backache, a 6 month history of left sciatica and recent onset of perineal anaesthesia and difficulty passing urine. He had restricted straight leg raising bilaterally, weakness of dorsiflexion and plantar flexion of both feet and reduced sensation in the S1–5 distribution on the left and S3–5 on the right.

A myelogram demonstrated a multilobulated intradural filling defect extending from the sacrum to the upper border of L3 (Fig. 1). CT (Fig. 2) confirmed a soft tissue mass within the spinal canal invading the neural arch of L3 and the left 1st sacral foramen. An L3 to S1 laminectomy was performed. A fleshy, vascular mass 5 cm in diameter was found within the dura mater, attached to and extending along the nerve roots, giving an extradural component. A diagnosis of small round cell neoplasm was made on frozen section and smear preparation. The tumour was considered non-resectable because of its vascularity and the nerve root involvement.

Paraffin sections showed sheets of small round/spindle cells with regular nuclei and sparse cytoplasm (Fig. 3). There were occasional but incomplete rosettes and no particular cellular pattern was identified. Mitoses were hard to find but apoptotic bodies were frequently seen. Both fat and nerve in the specimen were infiltrated by tumour. A periodic acid-Schiff stain showed little glycogen. Immunohistochemistry showed the tumour cells were focally positive for antisera to S-100 protein and vimentin but were negative for leucocyte common antigen, epithelial membrane antigen, cytokeratin, glial fibrillary acidic protein, neurofilament protein, leu 7, chromagranin A, PGP 9.5, neurone specific enolase and myoglobin.

Electron microscopy (Fig. 4) showed straight, closely aligned cellular membranes with frequent small intercellular spaces filled with electron-dense material into which small cellular processes projected. Intracellular organelles: mitochondria, endoplasmic reticulum and Golgi apparatus, all appeared prominent. Few specialised cell junctions and no cellular interdigitations were identified. The tumour cells had no regular basement membrane and no cilia or secretory droplets were identified. A diagnosis of PNET was made.

This histological diagnosis prompted CT of the posterior cranial fossa to exclude a primary cerebellar tumour; this was normal. At no stage during the patient's illness did he show any evidence of cerebellar disease.

Megavoltage radiotherapy was commenced and provided pain relief, recovery of bladder function and return of mobility. One year later the patient presented with recurrent back and leg pain and weak legs. A CT myelogram showed almost total obstruction at the level of the L4/L5 interspace caused by a mass occupying the spinal canal and extending into the L5/S1 exit foramen on the left. A large meningocele related to previous surgery was present but unaffected by tumour. Chemotherapy was commenced but two months later severe right sciatica had developed and CT myelography showed further involvement of the sacrum with gross destruction of the left ala and infiltration of the sacroiliac joint, with a large soft tissue mass extending into the pelvis. Palliative radiotherapy was administered but the patient died two months later. Permission for autopsy was not obtained.

Discussion

In 1973 Hart and Early described 23 cases of poorly differentiated cerebral tumours in children with histological features similar to medulloblastoma [4]. They postulated that the tumours reflected the same process occurring in different locations and proposed the name PNET. Rorke [5] observed that biologically and histologically similar tumours can arise in any part of the CNS and probably represent neoplastic transformation of primitive neuroepithelial cells found in the subependymal regions. Histologically PNET consist of small round cells with hyperchromatic nuclei with a high mitotic rate. They may show some

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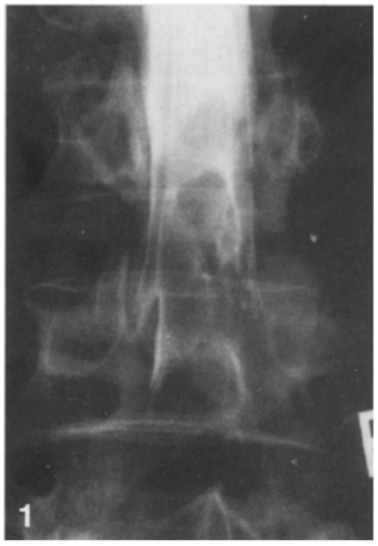

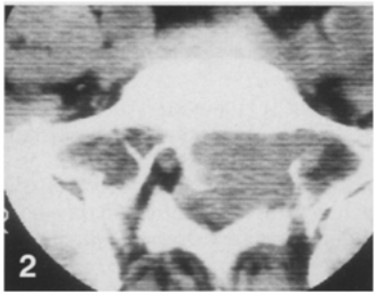

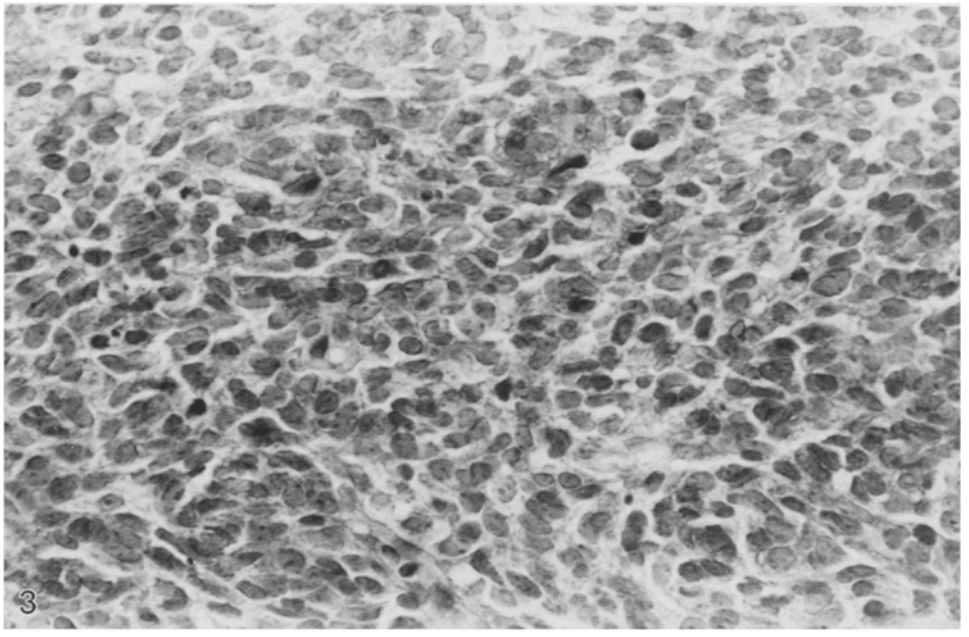

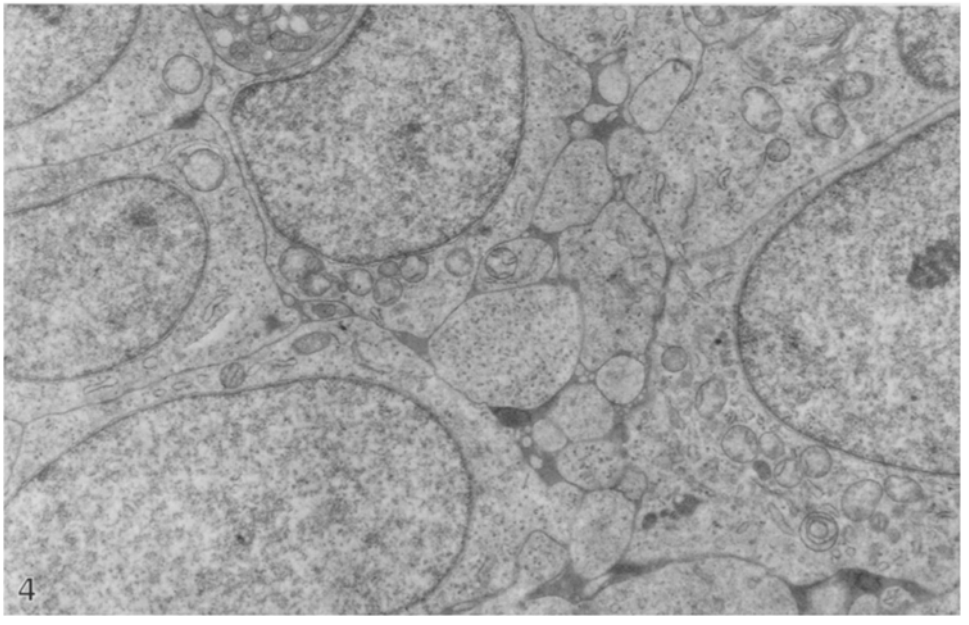

Fig. 1. Posteroanterior projection of a myelogram showing obstruction produced by a multilobulated filling defect with its upper border at L3, which at surgery was primarily intradural

Fig. 2. CT of the lumbosacral spine demonstrating a lobulated soft tissue mass in the spinal canal with bony erosion/destruction and growth through the neural foramina

Fig. 3. Representative histological section of the tumour, showing undifferentiated, slightly spindle-shaped cells with little cytoplasm. Haematoxylin and eosin, × 340

Fig. 4. Electron micrograph of tumour cells showing straight cell membranes with electron-dense material in intercellular space. No desmosomes or basement membrane material are identified, × 59,500

degree of neuronal or glial differentiation but over 90% of the cells must be primitive to meet Hart and Earle's original diagnostic criteria. On electron microscopy the cells lack evidence of specialised intercellular junctions [6].

PNETs usually occur in children or young adults. Medulloblastomas have a five year survival rate of 64% [7] but at other sites PNET have an average survival of less than two years [8]. They spread by means of local invasion, leptomeningeal extension and subarachnoid seeding of the neuraxis.

The commonest tumours of the cauda equina are meningioma, neurofibroma and ependymoma. Spinal tumours with small cells in adults are most commonly lymphoma, sarcoma or metastases. The histological diagnosis in our case was difficult, in part because of the patient's age. The morphological features were not those of meningioma, schwannoma or melanoma. This was confirmed by electron microscopy which also excluded ependymoma, malignant schwannoma and small cell carcinoma of the lung. The immunohistochemical findings excluded lymphoma and glioma. The paucity of glycogen and the scarcity of specialised cell junctions made Ewing's sarcoma an unlikely possibility. The absence of myofilaments and the age of the patient helped to exclude rhabdomyosarcoma. The tumour did however fulfill the pathological criteria for PNET [8].

Most cases of PNET involving the spinal cord do so by means of secondary "drop" metastases, deposited from an intracranial primary tumour, along the dorsal nerve roots, particularly those of the cauda equina [9]. The presence of

Table 1. Summary of published cases of PNET of cauda equina

Case	Age	Sex	Presentation	Metastases	Survival	Reference
1	24	Male	?	Lung	10 months	1
2	24	Male	Sciatica	Leptomeningeal	18 months	2
3	56	Male	Back pain, leg weakness	No	Alive	2
4	39	Male	Leg weakness	Leptomeningeal	42 months	2
5	26	Female	Sciatica	No	Alive	3
6	47	Male	Sciatica	Local	16 months	Present case

an intracranial tumour can be excluded by means of modern imaging methods. Primary spinal PNET is rare: including this case, 13 cases have been described, 9 in adults and 4 in children [1–3, 10–13]. Six of the tumours in adults occurred in the cauda equina (Table 1); these patients have tended to be older, with an average age of 36 years and are predominantly male. The age range is not highly unusual, as medulloblastomas have been described in patients up to 67 years of age [14], but the sex distribution is unexpected. The usual presentation is backache and lower limb weakness or sciatica.

Little has been written about the radiological findings of spinal PNET. The three cases described by Kosnik et al. [10] all had widened pedicles on spine radiographs, and that described by Liu et al. [3] caused enlargement of the spinal canal and sacral neural foramen, demonstrated by myelography, CT and MRI. Our patient showed erosion and expansion of the spinal canal and extension through the sacral neural foramina, demonstrated by CT.

As already mentioned, the differential diagnosis of a lesion of the cauda equina includes meningioma, neurofibroma, epidermoid and ependymoma. Meningioma and neurofibroma are the commonest intradural spinal tumours in this region. Meningiomas erode bone in less than 10% of cases and are calcified in up to 10% [15]. They tend to occur in older patients than neurofibromas and are more common in women. Neurofibromas are four times as likely to erode bone and may extend through the intervertebral foramen, but rarely calcify [16]. Both tend to have a smooth outline. Epidermoids and ependymomas are less common. The former may have fat characteristics on CT or MRI and be associated with congenitally abnormal vertebrae. Ependymomas are intramedullary tumours, which may expand the distal spinal cord. Metastases, including "drop" metastases, must always be considered in the differential diagnosis of an unusual mass occupying the spinal canal. They may produce an irregular arachnoid and glial contour, similar to arachnoid scarring, or can mimic an intradural extramedullary tumour, with single or multiple discrete nodules [16]. The tumour we report had radiological appearances suspicious of malignancy. PNET of the cauda equina is so rare that metastatic disease would have to be considered the most likely radiological diagnosis.

PNET of the cauda equina is aggressive and local recurrence or leptomeningeal spread are common; distant metastases were described in one case [1]. Despite treatment with surgery, radiotherapy and, in two cases, chemotherapy, four of the six patients have died within three and a half years. The optimal treatment of primary spinal PNET is unknown, but surgery followed by radiotherapy has been advocated, based upon experience with PNET at other sites [12].

Radiologists should be aware of PNET as a cause of malignant small cell tumour of the spine. Although primary spinal PNET is sufficiently rare to require investigation to exclude the possibility of "drop" metastases from medulloblastoma, the condition is a real entity, with a predilection for the cauda equina, particularly in older men.

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