Journal of Clinical Neuroscience 21 (2014) 2239–2242

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Case Reports

Malignant peripheral nerve sheath tumor of the cauda equina with craniospinal metastasis

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ARTICLE INFO

Article history:
Received 19 September 2013
Accepted 23 February 2014

Keywords:
Craniospinal metastasis
Malignant peripheral nerve sheath tumor
Spinal tumor

ABSTRACT

Intradural spinal malignant peripheral nerve sheath tumors (MPNST) are extremely rare, with only 20 adult patients reported to our knowledge, and only four primary tumors arising from the cauda equina. A 49-year-old man presented with back pain, constipation, and lower extremity weakness and was found to have a large intradural lesion involving the cauda equina. Imaging of the rest of his neuraxis revealed additional small left temporal lobe, cervical, and thoracic lesions. The patient underwent laminectomy for tumor debulking and biopsy, as gross total resection was not possible due to envelopment of the cauda equina. Histopathology revealed a MPNST with high cellularity, elevated proliferative indices, and nerve fascicle invasion. After the debulking, the patient reported improvement in his symptoms. However, 6 weeks later, the patient began having severe headaches, and his left temporal lobe lesion was found to have increased significantly in size, requiring craniotomy for palliative resection. The authors report the first adult patient with sporadic spinal MPNST with craniospinal metastasis to our knowledge. Imaging of the entire neuraxis is recommended for initial workup of these lesions, which are capable of intradural spread. The best treatment approach is unclear, but total surgical resection should be attempted, barring infiltration and engulfment of the nerve roots or widespread dissemination.

1. Background and importance

Malignant peripheral nerve sheath tumors (MPNST) are a rare form of sarcoma with an incidence of less than 0.001% in the general population [1]. The majority of MPNST occur in patients with neurofibromatosis type 1, who carry a 10% lifetime risk of developing this tumor [2]. Rarely, MPNST may arise from malignant degeneration of a neurofibroma, schwannoma, ganglioneuroma, or pheochromocytoma. They usually involve peripheral nerves of the trunk, extremities, or head and neck area [3].

Primary intradural spinal MPNST are a rare entity, with four pediatric patients and 20 adult patients reported in the literature [4]. Only four of these adult cases have involved the cauda equina primarily. Here we report an adult patient with a primary sporadic MPNST of the spine with evidence of craniospinal metastasis on presentation.

2. Clinical presentation

A 49-year-old man without significant past medical history presented to the Emergency Department with 2 weeks of increased back and leg pain and severe constipation. He had a history of smoking a half-pack per day, but no family history of cancer or neurofibromatosis. On neurologic examination, he had decreased light touch and pinprick sensation in the S1 distribution bilaterally, hypoactive Achilles reflex, diminished rectal tone, and 4/5 strength in his distal lower extremities. No cutaneous stigmata were identified. He underwent MRI of his lumbar spine which showed a large enhancing intradural mass filling the thecal sac below the conus level (Fig. 1A, B). Imaging of his chest, abdomen and pelvis did not reveal any additional pathology. MRI of the remainder of his neuraxis identified areas of enhancement in his left anterior temporal lobe and posterior to the thoracic spinal cord (Fig. 1C, D) and an area of intradural extramedullary nodular enhancement anterior to the cervical spinal cord.

With the plan to debulk the tumor and obtain tissue for diagnosis, the patient was taken to surgery for L4, L5, and S1 laminectomies. Once the dura was opened, as expected, the tumor was found to be tightly investing the nerve roots. The tumor mass was debulked but full dissection of the nerve roots from the tumor could not be performed due to the adherence. Intraoperative cytological imprints and cryostat sections of the specimen revealed cohesive spindle cells, and permanent sections revealed high cellularity and nuclear atypia with penetration and entrapment of nerve fascicles (Fig. 2). The tumor stained strongly for S100 and vimentin with focal cytoplasmic staining for glial fibrillary acidic protein.

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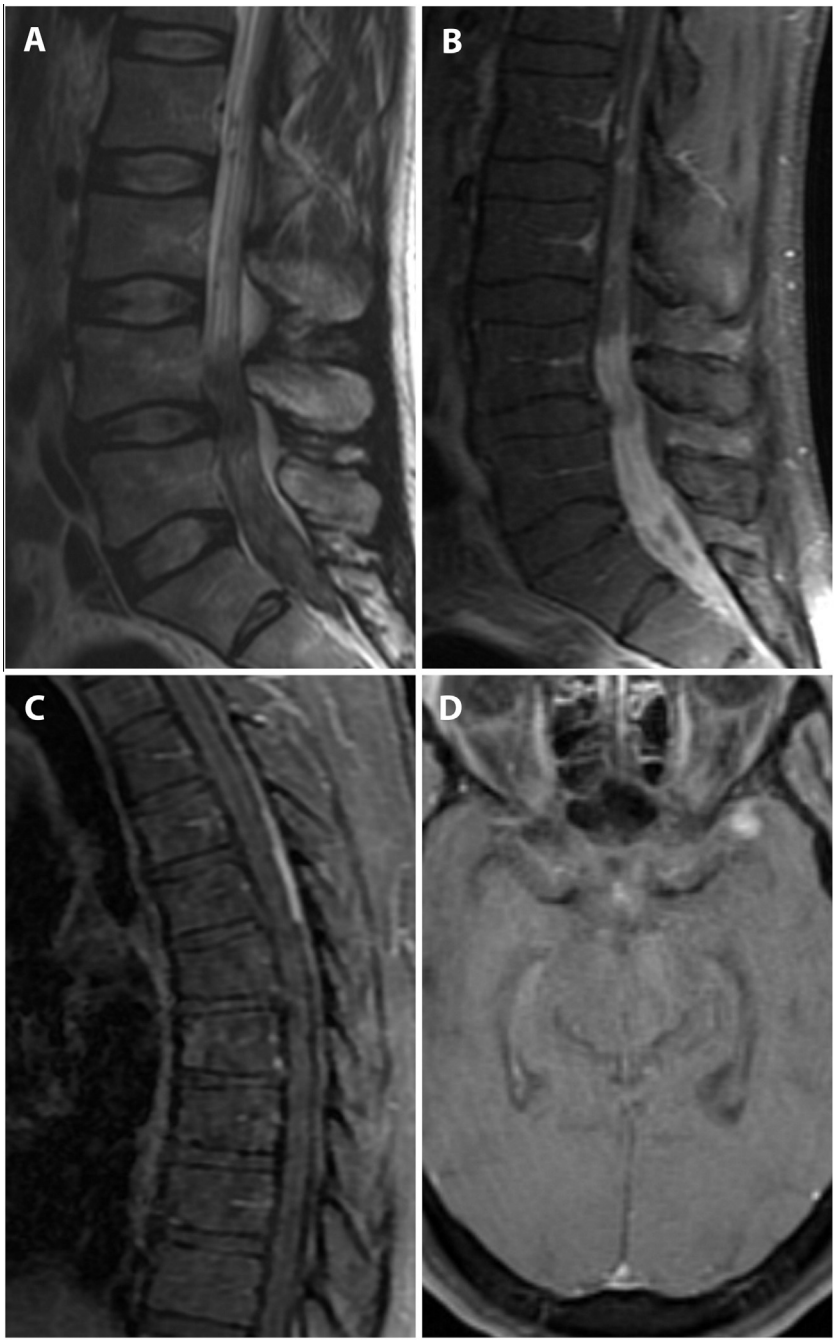

Fig. 1. MRI of the lumbar spine shows a diffuse infiltrative intradural process on (A) T2-weighted and (B) T1-weighted post-contrast sagittal sequences. Imaging of the rest of the neuraxis revealed intradural contrast-enhancing lesions in (C) the thoracic spine (sagittal T1-weighted post-contrast) and (D) the left temporal lobe (axial T1-weighted post-contrast).

The Ki67 stain revealed elevated proliferative index (7–10%) in large atypical cells throughout the tumor. Histopathology was consistent with MPNST.

Postoperatively, the patient reported improvement in his pain and his constipation. Permanent pathologic examination was consistent with MPNST. Six weeks later, he began having severe headaches. MRI of the brain showed that the small left temporal lobe metastasis had increased markedly in size and was now causing significantly mass effect (Fig. 3). His cervical and thoracic lesions showed only slight interval growth. He underwent a palliative craniotomy for resection of the temporal lesion. Histopathology was consistent with MPNST.

3. Discussion

Primary intradural MPNST of the cauda equina are very rare, with only four adult patients previously reported in the literature to our knowledge [4]. Generally, optimal treatment of MPNST requires total resection with clear margins – 5 year survival is 67% in cases of clear margins compared to 22% with positive margins [5]. However, the same oncologic resection principles can rarely be applied in spinal MPNST due to the location in the spinal canal and involvement of neural tissue. Adjuvant radiotherapy may improve rates of local control [5], so even after total resection in spinal MPNST, adjuvant radiotherapy should be considered.