Case Report

Thoracic dumbbell spinal metastasis secondary to neuroendocrine tumor of unknown origin: Case report and literature review

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ABSTRACT

Background: Dumbbell tumors are typically benign schwannomas, neurofibromas, and meningiomas and only rarely there are malignant variants of these lesions or other malignant histotypes. Here, a 34-year-old male presented with a thoracic spinal dumbbell metastatic neuroendocrine carcinoma of unknown primary origin.

Case Description: A 34-year-old male presented with 2 months of thoracic pain and progressive mid thoracic sensory loss. A post contrast thoracic MRI showed a dumbbell tumor localized between the T7 and T9 levels with extension laterally into the T7-T8 and T8-T9 foramina. The patient underwent a laminectomy for tumor resection following which his pain and gait improved. Histopathologically, the tumor demonstrated multiple rounded small cells with a Ki67 level around 30%, suggesting a malignant metastatic neuroendocrine tumor of unknown etiology.

Conclusion: We successfully treated a 34-year-old male with a T7-T9 malignant spinal dumbbell neuroendocrine tumor of unknown etiology utilizing a decompressive laminectomy.

Keywords: Dumbbell, Metastasis, Spine, Tumor

INTRODUCTION

Dumbbell tumors are typically benign schwannomas, neurofibromas, and meningiomas, with occasional malignant variants. Few dumbbell tumors are metastatic neuroendocrine lesions of unknown etiology.1,3,5,9 Neuroendocrine tumors are epithelial neoplasms originating mainly from the gastrointestinal or bronchopulmonary tracts; they frequently metastasize to lymph nodes, liver, lungs, and bone.2,5 Here, a 34-year-old male presented with a T7-T9 metastatic neuroendocrine spinal dumbbell tumor of unknown primary origin that was successfully decompressed utilizing a laminectomy.
CASE DESCRIPTION

Clinical and radiographic presentation

A 34-year-old male presented 2 months of mid thoracic pain, gait impairment/ataxia, and hypoesthesia from T11 downward. A post contrast thoracic MRI showed a dumbbell tumor between the T7 and T9 (i.e., maximal width T8) levels contributing to severe spinal cord compression [Figures 1a-d]. This lesion was hypointense on T1/T2-weighted images [Figure 1c], hyperintense on STIR sequences [Figure 1d], and intensely enhanced with contrast [Figures 1a and b]. The whole-body PET-CT-scan revealed multiple diffuse osteoblastic lesions, and the high level of a radioactive tracer involving the 8th/9th ribs bilaterally, sternum, pelvis, and C6-C7 and T3 pedicles.

Tumor markers

Blood tumor markers, that included beta-2-microglobulin, alpha-fetoprotein, carcinoembryonic antigen, and human chorionic gonadotropin-beta, were negative. Alternatively, serum neuron-specific enolase levels were high (48.4 µg/L; normal range: 0–16.3 µg/L) consistent with a neuroendocrine tumor origin.

Surgery

The patient underwent a T8 total/partial T7-T9 laminectomies to decompress the spinal cord. The tumor was hyper vascularized and there were significant adhesions between the tumor and the dura mater. Ultimately, a partial resection of the extradural lesion was accomplished. Postoperatively, the patient's gait disturbance and dorsal pain were improved. The 1-week postoperative thoracolumbar MRI documented complete removal of the extradural tumor [Figures 2a and b].

Histology

The histopathological examinations showed multiple round-small cells whose Ki67 level was about 30%, compatible with a neuroendocrine tumor of unknown etiology. Immunostaining showed a positive check for chromogranin and synaptophysin [Figures 3a and b] [Table 1]. The combination of studies allowed for documentation of a well-differentiated neuroendocrine neoplasm (i.e., NEN sec WHO 2019) [Figure 4].

Postoperative course

The patient has started a chemotherapy (i.e., etoposide and cisplatin). After 10 months, his gait ataxia and lower limbs hypoesthesia improved. At that point, he underwent a 68 Gallium-DOTATOC positron emission tomography/computed tomography (PET/CT) that demonstrated stable disease.

DISCUSSION

Tumor presentation

According to the Dumbbell Scoring System, the neuroendocrine tumor presented in this case should be classified as a Grade 5 (size >5 cm, boundary indistinguishable, irregularly lobulated shape, and no osteolytic bone destruction) alternatively, it would belong to Grade 6 of the Eden classification (i.e. due to the multidirectional erosion of the bone). The young age

Table 1: Immunostaining showed a positive check for chromogranin, synaptophysin, CD56, and PanCK. TTF1, Calcitonin, CDX2, and PSA were found negative, excluding a metastatic tumor from a pulmonary, thyroid, colic, or prostatic primary site.

| Immunostaining          |          |
|-------------------------|----------|
| Chromogranin            | +        |
| CD56                    | +        |
| Synaptophysin           | +        |
| PanCK                   | +        |
| TTF1                    | –        |
| Calcitonin              | –        |
| CDX2                    | –        |
| PSA                     | –        |
of our patient, 34, led to the first diagnostic suspicion of a lymphoproliferative lesion (i.e., lymphoma), but this was ruled out when the whole-body PET CT scan documented multiple bone metastatic lesions.[6-8]

**Cases of metastatic neuroendocrine dumbbell spine tumors**

We found only two cases of comparable metastatic neuroendocrine dumbbell tumors of unknown origin in the literature.[4,8] Mori et al. reported a 51-year-old female with the sudden onset of paraplegia due to a dumbbell-shaped metastatic extradural lesion at the T10 and T11 levels accompanied by multiple vertebral metastases from a rectal NET.[4] After total tumor excision, the patient recovered and remained free from disease up to 18 months postoperatively. Saway et al. had a 75-year-old male with metastatic Stage 4 pNET, who presented with a cervical intradural-extradural metastasis at the C1-C2 level; it was effectively treated with a laminectomy and intradural tumor resection [Table 2].[8]

**Markers used to identify the etiology of neuroendocrine tumors**

Neuroendocrine tumors of unknown etiology compromise approximately 10% of all NETs. As they are predominantly undifferentiated, their biological behavior is very aggressive.[1] Urinary 5-hydroxy-indolic acetic can represent a valuable marker in establishing the NET diagnosis.[5] Certainly, the histopathological diagnosis is critical to further define and direct the treatment for Unknown Primary Cancer. Nevertheless, NET metastases carry a very poor prognosis, with 5-year survival rates ranging between 19% and 38%. In our case, the prognosis was supposed to be even

| Authors and year | Number of Patients | Age | Sex | Neurological Symptoms | Surgical Treatment | Tumor Location | Chemotherapy/Radiotherapy | Outcome/Follow-up |
|------------------|--------------------|-----|-----|-----------------------|-------------------|---------------|----------------------------|-------------------|
| Mori et al., 2015 | 1                  | 51  | F   | Paraplegia            | T10-T11 laminectomy, Partial C1-C2 laminectomy with intradural resection | T10-T11         | Unknown                    | Remission at 1.5 years |
| Saway et al., 2020| 1                  | 75  | M   | None                  | C1-C2             | Unknown        | Not specified              |                   |
| Our case, 2021    | 1                  | 34  | M   | Sensory ataxia        | T8 laminectomy, partial tumor resection | T7-T8-T9        | Etoposide and cisplatin    | Stable disease (under treatment) |
poorer (about 11 months) due to the unknown primary.\(^{(3)}\)

In other rare cases, aggressive recurrences may respond to chemotherapy/radiotherapy protocols as well as surgical intervention.

CONCLUSION

Here, we diagnosed a thoracic T7-T9 malignant metastatic spinal dumbbell neuroendocrine metastatic tumor of unknown origin in a 34-year-old male that was successfully treated with a laminectomy followed by chemotherapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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