Intraosseous malignant peripheral nerve sheath tumor with focal epithelioid differentiation of the thoracic spine

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The authors describe an extremely rare case with malignant peripheral nerve sheath tumor (MPNST) with focal epithelioid differentiation presenting as an intraosseous lesion of the spine. A 75-year-old woman presented with progressive paraplegia caused by epidural mass arising from the posterior element of the T7 vertebra. At surgery, the lesion was noted to originate from the T7 vertebra and separate from the dura and spinal nerve roots. The patient died of tumor metastases to the lungs six months after the initial presentation. Histological diagnosis was MPNST. However, the tumor also contained cystic structures lined by epithelioid cells, requiring differentiation from synovial sarcoma. From the histological and immunohistochemical features, as well as the absence of SYT-SSX fusion gene expression, the diagnosis of MPNST with focal epithelioid differentiation was made. This is the first case report of intraosseous MPNST of the spine with a peculiar biphasic appearance.

Key words: Epithelioid differentiation, intraosseous tumor, malignant peripheral nerve sheath tumor, spine

Introduction

Malignant peripheral nerve sheath tumor (MPNST) of the spine is a rare spinal tumor and its rate among primary spinal tumors of Schwann cell origin was reported to be 2-3%. The spinal MPNSTs usually arise from spinal nerve roots and cause secondary bony changes. Intraosseous MPNST is extremely rare and to the best of our knowledge, only two such cases in the spine have been reported in the literature.

Aberrant epithelial or epithelioid differentiation in MPNSTs is rare but well-recognized. Epithelioid MPNST is defined as a tumor predominantly composed of large epithelioid cells and in most cases, they are strongly positive for S-100 protein and do not form glandular or cystic structures. It is estimated that 5% or fewer MPNSTs belong to this group.

In this report, we present a rare case of MPNST arising from the intraosseous region of the thoracic spine. The case report is the first documentation of intraosseous MPNST with peculiar biphasic differentiation occurring in the spinal column.

Case Report

A previously healthy 75-year-old woman presented with a two-week history of severe back pain, progressive hypesthesia and motor weakness of both legs. The magnetic resonance imaging (MRI) of the thoracic spine revealed that an ill-defined epidural mass arising from the intraosseous lesion of the posterior element of the T7 vertebra severely compressed the spinal cord. A metastatic spinal lesion was mostly suspected but extensive radiographic examinations including total body computed tomography scan revealed no evidence of other primary neoplasms. Due to progressive paraplegia, the patient underwent emergency decompression surgery one day after the admission. The lesion involved the lamina, right pedicle and bilateral transverse processes. The epidural tumor was thoroughly resected and the posterior stabilization was performed using the Luque titanium rods with sublaminar cables. The epidural tumor was easily detached from the dura. Bilateral T7 spinal roots were identified separate from the tumor mass. The postoperative histological diagnosis was MPNST.

Postoperatively, the symptoms of the patient were relieved dramatically. However, the patient’s symptoms recurred in six weeks postoperatively. The tumor grew rapidly thereafter despite local radiotherapy. Six months after the initial clinical presentation, the patient died from respiratory failure due to direct invasion of the mass into the thorax and multiple lung metastases.
Intraosseous MPNST of the spine

Miyakoshi et al.: Intraosseous MPNST of the spine

Autopsy was performed to reconfirm the previous histological diagnosis.

Histologically, the primary tumor consisted of sheets of tumor cells with alternating areas of hyper- and hypocellularity [Figure 3A]. The majority of the tumor cells were atypical spindle cells arranged in vague fascicles [Figure 3B]. Although some tumor cells had abundant eosinophilic cytoplasm, there was no evidence of rhabdomyoblastic features, such as cross striations. Cystic spaces containing serous fluid were scattered within the tumor and some of the spaces formed malignant cystic or glandular structures lined by epithelioid cells with marked nuclear atypia [Figure 3C]. Both spindle and epithelioid tumor cells were strongly positive for vimentin. Nuclei of some spindle cells were faintly positive for S-100 protein [Figure 3D]. The epithelioid cells lining the cystic spaces and adjacent spindle cells were occasionally weakly positive for AE1/AE3 and CAM5.2 but they were negative for epithelial membrane antigen and chromogranin A. The tumor cells were negative for α-smooth muscle actin, desmin, CD31, CD34 and HMB-45. The percentage of MIB-1-positive nuclei to all nuclei was 38%. The SYT-SSX fusion genes, which are specific to synovial sarcoma, were not detected by reverse transcription-polymerase chain reaction using RNA isolated from the tumor tissue. From these histological, immunohistochemical and cytogenetic findings, we finally diagnosed this tumor as MPNST with focal epithelioid differentiation. The diagnosis was approved by an experienced pathologist in this field in an independent institute. The metastatic tumors in the lungs consisted of sheets of spindle cells similar to those seen in the primary tumor.

Discussion

MPNSTs are relatively uncommon tumors of cells of peripheral nerve sheath origin. Secondary infiltration of bone is a well-recognized feature, but primary intraosseous MPNSTs appear to be extremely rare. To the best of our knowledge, only two cases with primary intraosseous MPNST of the spine have been reported in the literature. Khan et al. reported intraosseous MPNST that arose in the C2 vertebra of a 40-year-old woman. The patient underwent tumor excision and stabilization of the cervical spine, but died of pulmonary metastases one year later. Gnanalinghan et al. reported a 59-year-old woman with intraosseous MPNST of the T3 vertebra causing spinal cord compression. The patient underwent decompression surgery with spinal instrumentation and survived at one-year follow-up despite developing metastatic deposits in the spine and femur.

In general, the prognosis of primary spinal involvement of MPNSTs is not good. These lesions usually arise from the spinal nerve roots and cause secondary bony changes (MPNSTs at paraspinal regions are a known entity). Their one-year and five-year survival rates were 76% and 23%, respectively. Local recurrence occurs in 71% of cases. Metastases, most often to the lung, were reported in 33%. The present case with intraosseous spinal MPNST with neurological deficit was characterized by a more dismal clinical outcome. Incomplete removal of the tumor with microscopic spread and ineffectiveness of
radiation resulted in poorer prognosis in this case. Complete en-bloc resection is impossible for intraosseous spinal MPNST like the present case because of the diffuse tumoral infiltration into the bone and extradural space.

The intraosseous primitivity of a nerve sheath tumor is undoubted when, in the absence of a relationship with a nerve trunk, radiographically and macroscopically the tumor lies wholly within a bone. However, the bone involvement when extensive like the present case and the previously reported two cases with intraosseous spinal MPNST[3,4] cannot be surely recognized as primary or secondary. Similarly as the previously reported cases,[3,4] the diagnosis of primary intraosseous MPNST in the present case was made with intraoperative findings of the lesion that was noted to originate from the vertebral bone and separate from the dura and spinal nerve roots.

Histologically, divergent differentiation in MPNST occurs in approximately 15% of cases.[10] The epithelioid feature of the tumor in our case required us to differentiate it from sarcomas with biphasic differentiation, especially synovial sarcoma, which may originate from the spines or paraspinal regions. Morrison et al.[11] described a rare case of paraspinal synovial sarcoma containing multiple cystic structures. However, in the present case, the histopathological diagnosis of MPNST was favored because of our findings of the histological features with alternating areas of hyper- and hypocellularity, focal positivity for S-100 protein and the absence of expression of the SYT-SSX fusion gene. The present case is the first reported case with intraosseous MPNST of the spine with aberrant epithelioid differentiation.

Figure 3: Photomicrographs of the primary tumor. The tumor consisted of sheets of atypical spindle cells with alternating areas of hyper- and hypocellularity (A and B: H/E, original magnifications 20× and 100×, respectively). Cystic spaces containing serous fluid were scattered within the tumor and some of the spaces were lined by epithelioid cells with marked nuclear atypia (C: H/E, 200×). Nuclei of some spindle cells were positive for S-100 protein (D: 400×)

References


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