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A Case of Thoracic Extraspinal Extradural Tanycytic Ependymoma with Its Radiological and Histopathological Findings

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Abstract

Extraspinal extradural tanycytic ependymoma is a very rare variant of ependymomas that usually tends to be localized in the spinal cord. We report a case of a 52-year-old woman presenting with back pain because of extraspinal extradural tanycytic ependymoma which is located in the paraspinal muscles at the level of the thoracic spine and presented its radiological and histopathological findings.

Keywords: Extraspinal extradural ependymoma, tanycytic ependymoma, thoracic spinal cord

Introduction

Ependymomas account for 60% of all intraspinal glial tumors.[1] They rarely occur in the extradural region and have more tendency to settle in the sacral-sacrococcygeal areas.[2-4] Tanycytic ependymoma is an uncommon fibrillary variant of ependymomas and shows a tendency to originate from the spinal cord.[5,6] Here, we aim to report an extraspinal extradural tanycytic ependymoma (EETE) with its different radiological findings and typical pathological features that are localized in an unusual position such as the paraspinal muscles at the level of the thoracic spine.

Case Presentation

A 52-year-old woman presented with back pain for 6 months. There was no swelling in physical examination although the tenderness with deep palpation was positive in the lower back region. Magnetic resonance (MR) imaging showed a 19 × 12 mm solid mass in the paraspinal region at the posterior level of the right transverse process of the T7 vertebra. The lesion was hypointense on T1-weighted MR imaging and heterogeneous hypointense on T2-weighted MR imaging. After contrast administration, it showed homogeneous enhancement (Figure 1). It had no relationship with the environmental bone structures and the spinal canal. Radiologically, the initial diagnosis was a soft tissue tumor. Surgery was performed and the mass was completely removed. Histopathologically, the tumor cells by forming pseudo rosette formation were seen in some areas but not frequently and the tumor consisted of bipolar spindle cells on the myxoid ground. On immunohistochemical examination, glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA) were positive (Figure 2). The final diagnosis was tanycytic ependymoma (WHO grade 2) containing myxoid areas.

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Discussion

In 1902, Mallory reported the first case of the extraspinal ependymoma,[2] and the primary extramedullary ependymoma of the thoracic spine was reported by De Bonis et al[7] in the literature. Tanycytic ependymoma was described by Friede and Pollak in 1978, as a fibrillary type of ependymoma that arises from the tanycyte cells.[5]

Considering the number of occurrences is high at the lumbar and coccygeal levels, many opinions state extradural ependymoma is originating from the intradural filum terminale or the extradural remnants of the filum or the coccygeal medullary vestige.[3] This opinion is also supported by the existence of associated spina bifida in 20%-30% of the patients.[8] The migration of the mass of ependymal cells may be correlated with the direct extension of an intradural intramedullary mass due to iatrogenic reasons. This opinion not only correlates with the featured position of the tanycytic cells which is from the third ventricle to the thoracic portion of the spinal cord but also implies the existence of a primary tumor which is unlikely in the case considered.[9] Another one is the metastasis and formation of a secondary tumor which is caused by an intradural mass. Although the probability of the occurrence is high in the case of the formation of extramedullary masses due to the easy transport opportunity via the cerebrospinal fluid, an extradural mass originating from sacrococcygeal vestige is unlikely but possible through blood and lymphatic vessels.[10]

Histopathologically, tanycytic ependymomas consist of spindle cells with fibrillary processes, ependymal rosettes-perivascular pseudo rosettes, but they have not seen often-evident perivascular pseudo rosette as in the classic ependymomas, and GFAP positivity.[11] They show low-to-moderate cellularity that is characterized by the spindle cells with nuclear pleomorphism, and mitotic changes are usually absent. According to the WHO, tanycytic

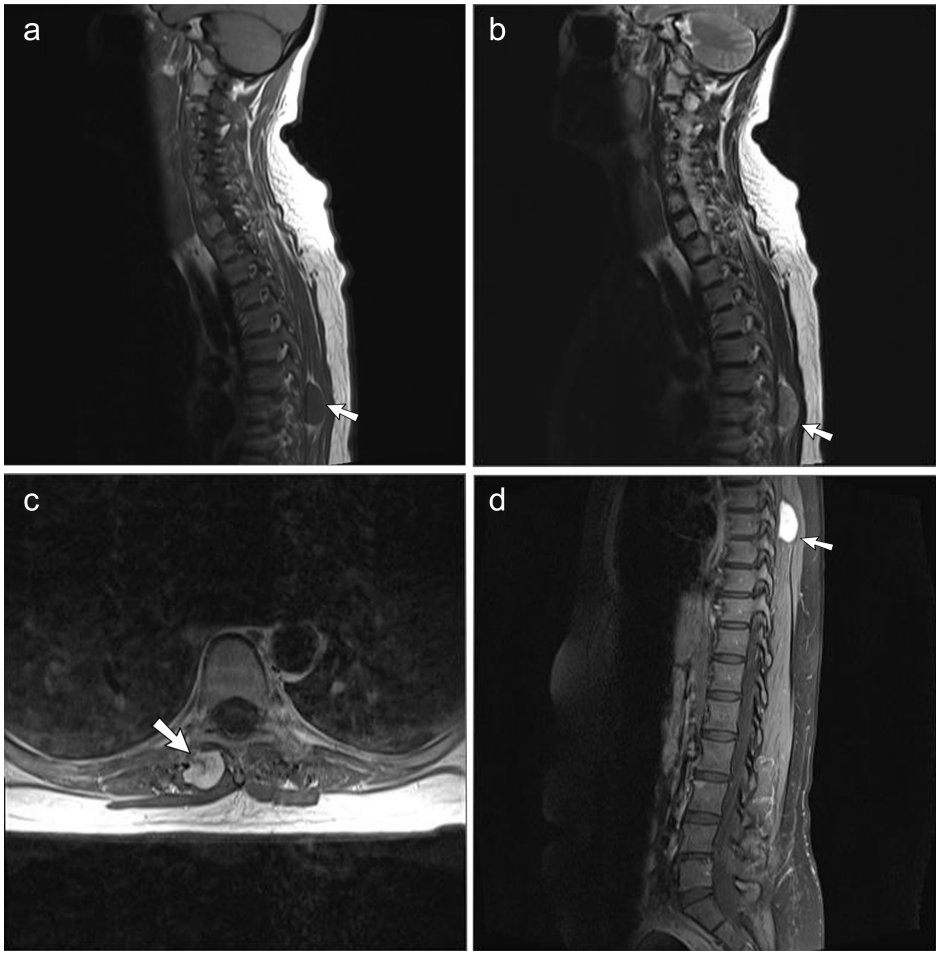

Figure 1. a-d. Sagittal T1- and T2-weighted images demonstrate a mass that is T1 hypointense (a), T2 heterogeneous hypointense (b) in the paraspinal region at the posterior level of the right transverse process of the T7 vertebra. It shows homogeneous enhancement on axial and sagittal contrast-enhanced T1-weighted images (c and d).

ependymomas are grade 2 tumors.[6] On histological examination, they can be confused with other tumors such as schwannomas and pilocytic astrocytomas. Pilocytic astrocytomas are biphasic lesions that consisted of fascicular and microcystic components although tanycytic ependymomas show no biphasic pattern.[6-12] Also tight perivascular conglomerate of spindle cells with ovoid nuclei and lack of Rosenthal fibers are different from pilocytic astrocytomas.[11,12] Schwannomas have typical Antoni A and B structures and nuclear palisading that is known as Verocay bodies. It is unusual to see these structures in tanycytic ependymomas.[6] In comparison with schwannomas, tanyctic ependymoma cells are more uniform and their nuclei are more oval. Immunohistochemical examination is very useful to distinguish these tumors. Tanycytic ependymomas show positivity for GFAP, EMA, and vimentin but rarely for S-100 protein. Schwannomas stain positive for S-100 protein but negative for GFAP.

Spinal ependymomas tend to be hypointense or isointense to the spinal cord on native T1-weighted MR imaging and usually hyperintense on T2-weighted MR imaging. After contrast administration, they show homogenous or heterogeneous enhancement. Due to the cystic formation and hemorrhage, heterogeneous appearance and enhancement may occur. Myxopapillary ependymoma is a subtype of spinal ependymomas, and these tumors present usually as a hyperintense mass on T2-weighted MR imaging because of their mucin component or hemorrhage. Some of the differential diagnoses of EETE are paraspinal soft tissue tumors, schwannomas with extension to the extraspinal space, hematomas, abscess, metastasis, lymphoma, neuroblastoma, or bone tumors. So here, the initial diagnosis was the paraspinal soft tissue tumor. But in our case, the different radiological finding of the lesion was heterogeneous hypointense on T2-weighted MR imaging usually unlike the soft tissue tumors. We thought this appearance might be due to the histological features of tanycytic ependymoma.

Due to extradural localization being close to the lymphatic and vascular structures, these tumors are more prone to metastasis to distant organ systems such as bones, liver, and lungs.[13] The first treatment choice of EETE is surgery. If metastasis, local recurrence, and incomplete resection are present, adjuvant radiotherapy may be suggested. The prognosis of ependymomas is bound up with age, localization, and size of resection.[14] Tanycytic ependymomas show a tendency to be in the spinal cord and have a better prognosis than cerebral lesions.[6]

Extraspinal extradural tanycytic ependymoma is a very rare type of ependymoma that settled in an unusual localization. The different side of our case was an uncommon type of ependymoma besides an unusual localization and also atypical radiological findings, especially heterogeneous hypointense on T2-weighted MR imaging.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

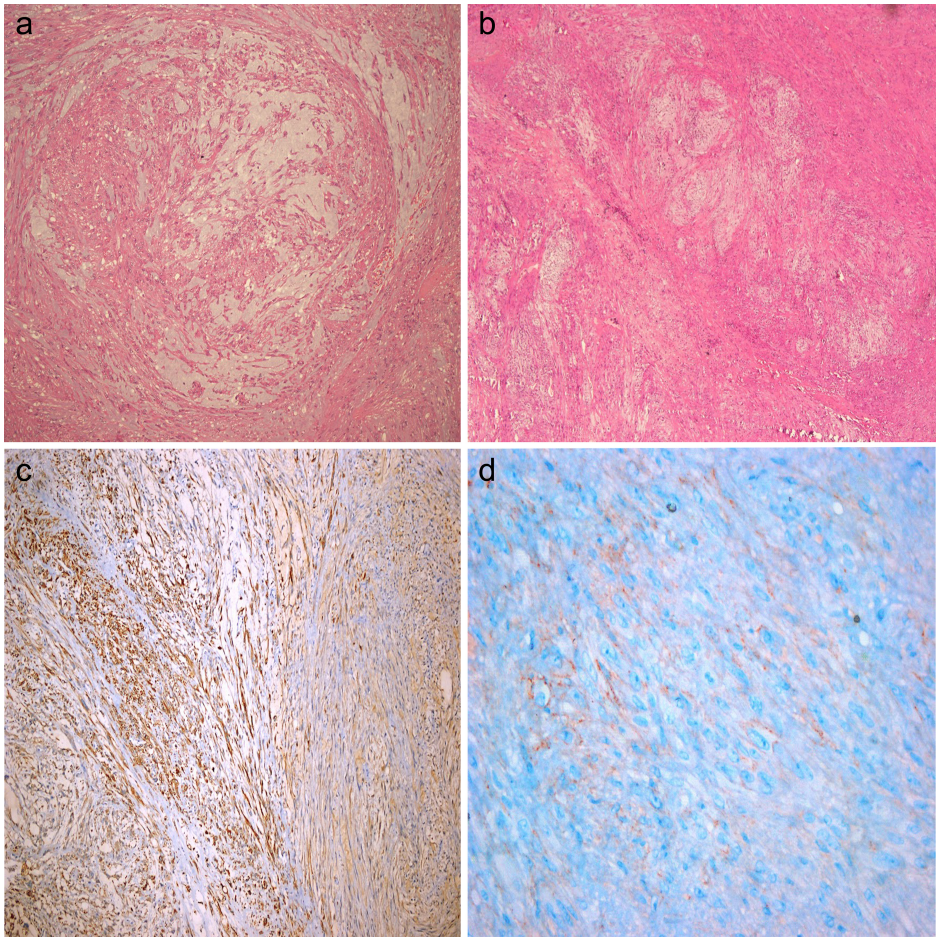

Figure 2. a-d. Hematoxylin and eosin staining revealed that the tumor cells by forming pseudo rosette formation are seen in some areas but not frequently and the tumor consists of bipolar spindle cells on the myxoid ground, magnification, ×100 (a); magnification, ×40 (b). Immunohistochemical staining for GFAP (c) and EMA (d) is positive. GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen.

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