Rare Occurrence of Synovial Sarcoma Originating from Dura Mater

Nadiye AKDENIZ,1 Mehmet KÜÇÜKÖNER,1 Muhammet Ali KAPLAN,1 Zuhat URAKCI,1 Halis YERLİKAYA,1 Oğur KARHAN,1 Fatma TEKE,2 Abdurrahman IŞIKDOĞAN1

1Department of Medical Oncology, Dicle University, Faculty of Medicine, Diyarbakır-Turkey
2Department of Radiation Oncology, Dicle University, Faculty of Medicine, Diyarbakır-Turkey

SUMMARY

Primary or metastatic dura mater sarcomas are rarely seen, and dural synovial sarcomas are quite rare. Here we report the case of a 60-year-old man who presented with headache, nausea, and vomiting and was found to have a 63×33×40-mm mass with solid and cystic components in parieto-occipital lobe. The mass was resected completely, with no residue detected on postoperative magnetic resonance imaging. On pathological evaluation, a vimentin-, S100-, transducer-like enhancer of split 1-, BCL-2-, and EMA-positive and desmin-, PR-, CD99-, PANCK-, and GFAP-negative synovial sarcoma with Ki-67 proliferation index of 12% was detected. Postoperatively, chemotherapy has been started, and it will be followed by radiotherapy. Primary meningeal-derived sarcomas are rare, and differential diagnosis with other meningeal mesenchymal tumors is based on the findings of morphological and immunohistochemical analyses.

Keywords: Dura mater; rare tumor; synovial sarcoma.

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Introduction

Dura mater-originated neoplasms comprise a wide range of primary and metastatic tumors based on their clinical, radiological, morphological, and immunophenotypical features. Treatment decision is based on tumor origin, morphologic type, and degree. Primary and metastatic dura mater tumors are rarely seen, and dural synovial sarcomas are quite rare.[1] There have been few reports on dura mater-originated synovial sarcoma; here we report the case of a patient with primary dural synovial sarcoma.

Case Report

A 60-year-old woman presented to us with nausea, headache, and vomiting. No abnormalities were detected on neurological examination, and her biochemical parameters were within normal range. Contrast-enhanced magnetic resonance imaging revealed a parieto-occipital mass 63×33×40-mm in diameter with a cystic and solid component and peripheral vasogenic edema that had caused right lateral ventricle compression (Fig. 1). The mass was resected completely, and no residue was detected on postoperative MRI.

Differential diagnosis of dural-origin fusiform and epithelioid tumors comprises hemangiopericytoma, fibrous meningioma, and synovial sarcoma. In the present case, on immunohistochemical examination, the tumor was found to be positive for vimentin, S100, NSE, transducer-like enhancer of split (TLE)-1, BCL-2, EMA, FactorXIIIa, and CD34 and negative for CD30, LCA, desmin, PR, STAT-6, CD99, pancytokeratine,
Discussion

Synovial sarcomas comprise 5%–10% of soft tissue sarcomas.[2] The occurrence of head and neck synovial sarcomas is even less, and they comprise 3%–5% of total sarcomas. Slight female predominance is seen, with the woman to man ratio being 1.6:1.[3,4] These tumors are mostly observed in individuals aged 15–35 years; however, their occurrence may range from being present at birth until the age of 89 years.[2]

Synovial sarcomas are not derived from synovium; they originate from primitive mesenchymal stem cells and can be seen in regions without synovium.[5] Synovial sarcomas are mostly seen on extremities; however, they may be distributed widely across the head and neck, mediastinum, abdomen, lung, pleura, kidney, and gastrointestinal tract.[6] These tumors have been reported to exist intracranially in the dura mater and base of the skull, indicating that they may be present anywhere in the body.[7]

Clinical presentation of meningothelial mesenchymal tumors includes their anatomic location and diameter. The affected patients mostly present with nausea, vomiting, headache, paresthesia, memory impairment, convulsions, disequilibrium, aphasia, and hearing loss. Neuroradiological examination has reportedly revealed meningeal-originated tumor and a range of dissemination.[8]

The appearance of these tumors is related to differentiation and resembles extracranial soft tissue sarcomas. Rapidly progressive tumors are poorly demarcated, fragile, hemorrhagic, necrotic, and cystic.[9] Histologically, synovial sarcomas are differentiated into four subtypes: monophasic, monophasic epithelial, biphasic, and poorly differentiated types.[10,11] The present case was classified as poorly differentiated synovial sarcoma because cystic, solid component and intratumoral hematoma were observed.

Based on the location and morphological features, the tumor in the present case was histopathologically different from meningioma, solitary fibrous tumor, and others sarcomas.[12] In addition to 100% immunoreactivity for EMA, 86%–90% nuclear positivity PgR is observed in meningiomas.[13,14] In the present case, contrary to EMA positivity, PgR was negative. Solitary fibrous tumors may also be located in central nervous system. They are negative for cytokeratin and EMA, while diffuse positivity is observed with CD34.[2] Although the present case was positive for CD34 and negative for cytokeratin, positive immunoreaction for EMA was inconsistent with the solitary fibrous tumor. Approximately 10%–20%
of synovial sarcomas have the same pattern as hemangiopericytomas.[1] Like peripheral nerve sheath tumors, 30% of monophasic and poorly differentiated synovial sarcomas show nuclear and cytoplasmic S-100-positive immunoreaction.[15] The immune reaction to EMA and cytokeratin is negative in malignant peripheral nerve sheath tumors; thus, these tests may be used in differential diagnosis.[16] In the present case, similar to malignant peripheral nerve sheath tumors, S-100 was immunoreactive, but unlikely EMA was immunoreactive either. Recently, high expression of TLE-1 has been observed in synovial sarcomas. In the present case, TLE immunoreactivity was positive; this feature is specific and sensitive for synovial sarcomas.[17] Furthermore, in this case, Ki-67 proliferation index was 12%. However, because synovial sarcoma-specific chromosomal translocation (X; 18) (p11; q11) was seen, diagnosis was made based on the findings of histological and immunohistochemical tests in the present case.[9]

Dura mater and leptomeningeal synovial sarcomas are very rarely seen, so optimal systematic treatment has not been established.[9] Wide surgical resection, radiotherapy, and doxorubicin-based chemotherapy may prevent local recurrence of the tumor and improve prognosis.[10, 18, 19]

Conclusion

In conclusion, primary meningeal synovial sarcomas have been seldom diagnosed, and case-based presentations are present. They can be differentiated from mesenchymal meningeal tumors on the basis of morphological and immunohistochemical features.

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References

12. Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH. Solitary fibrous tumor of the meninges:


