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Case report

Primary dura-based synovial sarcoma of the parafalcine region of brain

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ABSTRACT

Dura-based intracranial neoplasms include a wide range of primary and metastatic tumors, varying in their clinical, radiologic, morphologic, and immunophenotypic characteristics. At this anatomic location, sarcomas are rare, however, they exhibit close morphologic resemblances to meningioma. Herein we describe the third case of primary synovial sarcoma of the parafalcine region in a50-years-old female, who presented with left-sided hemiplegia. The radiologic survey revealed a 5.5 cm × 5.8 cm contrast enhancing dura-based mass at the right parafalcine region with meningeal enhancement and edema in the surrounding areas. Morphologic evaluation exhibited a high-grade spindle cell neoplasm, with focal hemangiopericytomatous pattern. The tumor cells were diffusely immunoreactive for CD99, Bcl2, TLE-1, and vimentin. The Ki-67 proliferation index was 40%. Pancytokeratin was focally positive. Epithelial membrane antigen, progesterone receptor, CD34, S-100, and glial fibrillary acidic protein were negative. Fluorescence in situ hybridization confirmed tumor specific translocation t(X;18)(p11.2;q11.2). Hence, final diagnosis of synovial sarcoma was rendered. Primary meningeal synovial sarcoma should be considered in the differential of aggressive and high-grade dura-based tumors in view of their relative chemosensitivity and future prospect of a molecular target-based therapy. The index case highlights the importance of an extensive pathologic analysis of high-grade mesenchymal lesions of the meninges to arrive at a definitive diagnosis and differentiate such tumors from other usual dura-based tumors, which has important therapeutic and prognostic implications.

1. Introduction

Dura-based intracranial neoplasms include a wide range of primary and metastatic tumors, varying in their clinical, radiologic, morphologic, and immunophenotypic characteristics. A comprehensive diagnostic work up to delineate the origin, morphologic subtype, and grade of these tumors is essential for therapeutic decision-making. Primary and metastatic sarcomas involving the dura are rare. Interestingly, dural sarcomas exhibit close histomorphologic resemblance to meningioma. Synovial sarcoma (SS) is one such dura-based sarcoma and only two cases of primary dura-based SS have been reported in the literature. Herein, we describe another case of primary parafalcine SS presenting with left-sided hemiplegia.

2. Case report

The patient is a 50-year-old female, who presented with left-sided hemiplegia (upper limbs more than lower limbs). Magnetic resonance imaging (MRI) revealed a contrast enhancing mass at the right parafalcine region, which measured 5.5 cm × 5.8 cm (Fig. 1). There was associated tissue edema and meningeal enhancement. The clinical and neuroradiologic impression was that of a meningioma. A stereotactic biopsy was performed, which exhibited a mesenchymal tumor composed of spindle cells arranged

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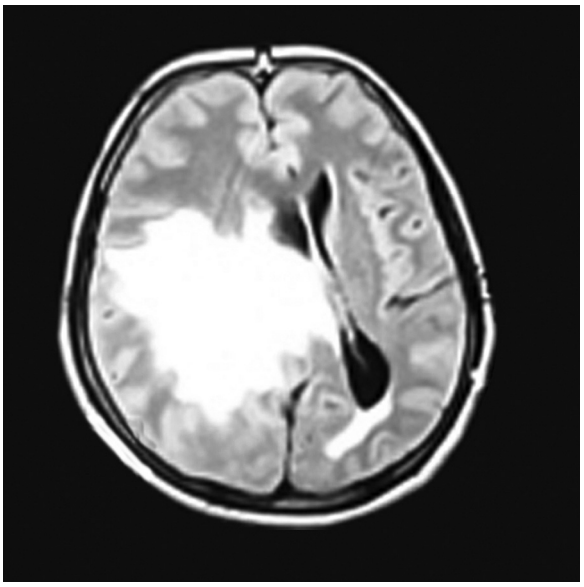

Fig. 1. Magnetic resonance imaging revealed a contrast enhanced right parafalcine mass (5.5 cm × 5.8 cm) causing mass effect with midline shift toward the left.

in short and long interlacing fascicles. The tumor cells depicted moderate nuclear pleomorphism and inconspicuous nucleoli and were having scant cytoplasm. Hemangiopericytomatous "staghorn" vascular pattern was observed. The mitotic activity was high including a few atypical mitotic figures. Mitotic figures were 40 per 10 high-power fields (Fig. 2A). Epithelial-looking areas were not present in the sections studied. There was a clear demarcation between the tumor and the brain parenchyma. Meningothelial whorls or features of meningothelial differentiation were not identified. There was absence of necrosis. Based on the cytoarchitectural features, the differential diagnoses considered were solitary fibrous tumor (SFT), meningioma, and meningeal sarcoma (primary or metastatic). A battery of immunohistochemical stains, including pancytokeratin (CK), vimentin, epithelial membrane antigen (EMA), progesterone receptor (PgR), CD99, B-cell lymphoma 2 (Bcl2), transducer-like enhancer of split-1 (TLE-1), CD34, S-100, glial fibrillary acidic protein (GFAP), and Ki-67 was performed. The neoplastic cells revealed diffuse and moderate to strong immunoreactivity for CD99 (membranous), Bcl2 (membranous and cytoplasmic), TLE-1 (nuclear), and vimentin (cytoplasmic). CK revealed strong and focal positivity in the neoplastic cells in a cytoplasmic and membranous fashion. EMA exhibited focal and strong positivity. The Ki-67 proliferation index of 40% was observed in the most proliferative zone of the tumor (Fig. 2B–G). EMA, PgR, CD34, S-100, and GFAP were negative. Fluorescence in situ hybridization (FISH) assay was performed on formalin-fixed, paraffin-embedded sections using dual color break-apart probe within the *SYT* or *SS18* gene on 18q11.2 location. Total of 235 cells were scored, of which 90% tumor cells showed *SS18* gene translocation (Fig. 2H). Keeping in view the clinical and pathologic findings, final diagnosis of meningeal synovial sarcoma was given.

3. Discussion

Early in the 20th century, researchers coined the term SS for those mesenchymal lesions which had a microscopic similarity to the synovium and a propensity to arise adjacent to the joints. With better understanding of the disease and diagnostic breakthroughs, SS now exhibits a wide anatomic distribution, including thigh, abdominal wall, head and neck, including intracranial region, mediastinum, abdominal cavity, lung, pleura, kidney, and gastrointestinal tract [1]. Hence this disease is no longer considered to originate from the synovium or have any relationship with it. Smith et al. in 1987 established the origin of this tumor from an unknown multipotent stem cell having the capability to differentiate into mesenchymal and/or epithelial structures [2]. As the origin of the tumor is still unknown, World Health Organization classifies SS as a malignant tumor of uncertain differentiation and discourages the earlier used synonyms such as tenosynovial sarcoma, synoviosarcoma, synovial cell sarcoma, malignant synovioma, and synovioblastic sarcoma [3]. SS is considered to be a disease of adolescent and young adults but can occur in any age group. 58% of the cases occur between 10 years and 40 years of age and 77% cases occur before 50 years. In the head and neck region, SS occurs between 5 years to 55 years, with a median of 29 years and a mean of 30.6 years [4]. In the head and neck region, SS is a rare mesenchymal lesion and comprises 3–5% of all sarcomas occurring in this region. SS has no gender preference, however, a slight female preponderance is known, with a female to male ratio of 1.6:1 [3,5]. The clinical presentation of non-meningothelial mesenchymal tumors depends on the anatomic location and the size of the tumor. The commonest symptoms are nausea, vomiting, headache, loss of sensation of different parts of the body, loss of memory, convulsions and difficulty in muscle movements, balance, posture, speech, and hearing. Neuroradiologic survey has an important role in discerning the origin of the tumor from the meninges and visualization of the extent of the tumor. Presence of multiple small and spotty radiopacities caused by focal calcification is the most characteristic radiologic feature of SS elsewhere in the body [6]. However, this feature was not seen in the index case. A computerized tomography (CT) scan identifies this feature in about one third of cases [7]. MRI shows variation in intensity and enhancement with frequent septations. Most tumors are relatively iso- or hypointense compared to muscle and iso- or slightly hyperintense compared to fat [8] (Table 1).

The macroscopic appearance of these tumors depends on their differentiation and it generally resembles the corresponding extracranial soft tissue tumors. While slowly growing SS tend to be sharply circumscribed, round, or multilobular, with a smooth glistening pseudocapsule,rapidly growing tumors are poorly circumscribed and exhibit a variegated, friable, or shaggy appearance, with hemorrhage, necrosis, and cyst formation. Meningeal sarcomas are known to invade the brain parenchyma and usually have a firm texture. Histologically, SS are divided into two major categories: biphasic and monophasic, based on the presence of epithelial and/or spindle cell components. Monophasic fibrous type is the most common subtype. Poorly differentiated round cell SS arranged in a pericytomatous pattern has also been described. It represents the tumor progression that can occur in either monophasic or biphasic tumors [9].

Considering the site and morphology of the tumor, the present case had a number of histopathologic differentials, which included meningioma, solitary fibrous tumor (SFT), including its malignant form, and other sarcomas. Absence of morphologic architectural patterns of meningioma and negative staining for EMA and PgR argues against a meningioma [10]. Abdelzaher et al. in their study of 87 meningiomas showed a 100% expression of EMA, concluding the diagnostic efficacy of this marker in diagnosing meningioma [11]. Negativity for PgR also strengthened the results as nuclear positivity of PgR is seen in 86–90% cases of meningioma [12]. The characteristic appearance of SFT was investigated carefully, but in the sections submitted the spindle cells were not disposed in wavy fascicles between prominent eosinophilic bands of collagen [13]. The "staghorn" vascular pattern separating the tumor into small lobules was the key morphologic feature in the present case. Hence sarcomas with this feature were sought for. SSs are known to exhibit hemangiopericytoma-like pattern in approximately 10–20% cases. Multiple sections were then studied to