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Case Report

Primary synovial sarcoma of the brain in a 35-year-old patient: A rare clinical experience

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Abstract

Synovial sarcoma is an aggressive soft-tissue sarcoma with a poor prognosis and has an origin of uncertain histology. It frequently presents as a localised disease, especially near large joints around the knee and thigh. Intracranial disease, which is rare, has been reported as a metastasis from synovial sarcoma. We report the case of a patient with no obvious primary extracranial pathology, suggesting primary intracranial synovial sarcoma. A 35-year-old male presented with altered sensorium for 1 week. Imaging was suggestive of a left frontal high-grade tumour, for which he underwent decompression. Histology showed biphasic synovial sarcoma. Positron emission tomography–computed tomography did not show lesions elsewhere. He underwent radiotherapy adjuvantly.

Keywords: Adjuvant radiation, decompression surgery, immunohistochemistry, primary intracranial, synovial sarcoma

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INTRODUCTION

Synovial sarcomas are very rare, aggressive in nature and generally have a poor prognosis with uncertain histological origin. The pathology of synovial sarcoma is most commonly present locally, especially near large joints around the knee and thigh.[1] The early assumption in our case of synovial sarcoma was probable metastasis from an extracranial site. Nevertheless, later, it was excluded after adequate metastatic workup and was treated as primary intracranial synovial sarcoma.

CASE REPORT

A 35-year-male-patient with no known comorbidities presented with a history of fever, vomiting and altered sensorium for 1 week. There was no history of trauma, seizures and limb weakness. Higher mental function examination showed that the patient was conscious, confused and disoriented. Cranial nerve examinations were within the normal limits. On motor examination, bulk and tone were normal, and power was 4/5 in all groups of muscles. On sensory examination, touch, pain and temperature sensations were normal. Posterior column sensations and deep tendon reflexes were normal. Superficial reflexes were present. Plantar reflexes were bilaterally down going. Signs of cerebellar dysfunction were absent. No neurocutaneous markers were noted. Contrast-enhanced computed tomography (CT) of the brain showed a lesion in the left frontal region, suggestive of a left frontal high-grade tumour (Figure 1).

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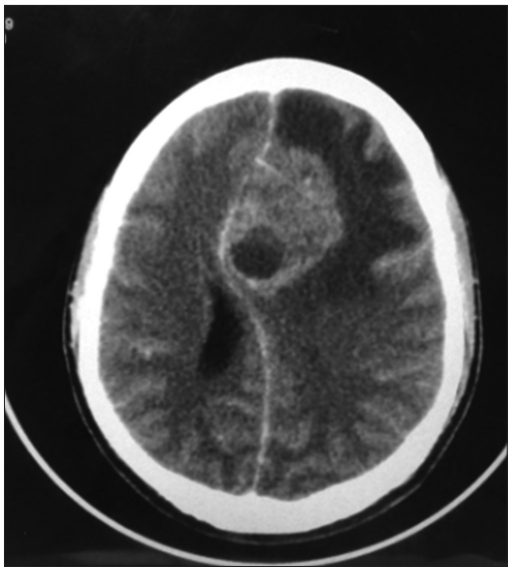

Figure 1: Pre-operative CT brain showing heterogeneously enhancing soft-tissue density lesion in the left frontal region CT = Computed tomography

The patient underwent left frontal craniotomy and gross total decompression of the lesion. Tumour was greyish white, highly vascular, suckable. Squash cytology report was suggestive of glioblastoma. Post-operatively, there was a marked improvement in sensorium. Post-operative CT of the head showed near total decompression of lesion and no operative site haematoma (Figure 2).

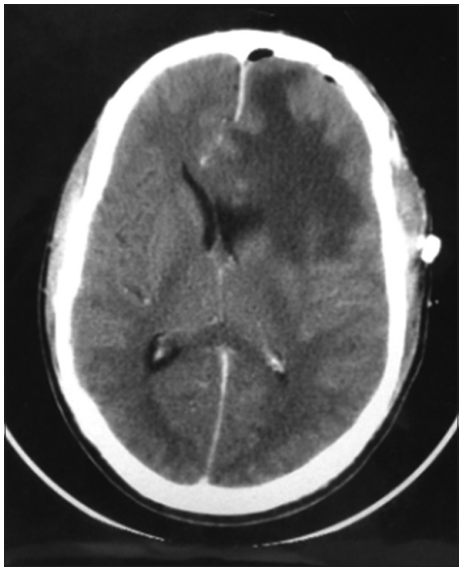

Figure 2: Post-operative CT of the brain showing surgical cavity showing no residual lesion. CT = Computed tomography

Histopathology showed a relatively well-circumscribed malignant mesenchymal spindle cell sarcoma (Figure 3). Immunohistochemistry showed diffuse positivity for

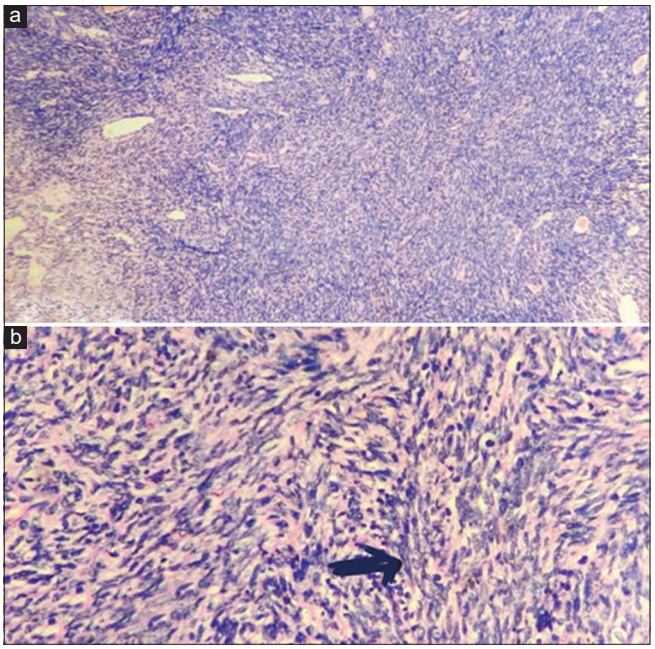

Figure 3: Photomicrographs showing spindle to elongated cells with focal trabecula of epithelial cells (arrow) suggestive of malignant biphasic synovial sarcoma (Haematoxylin and eosin, X100) (a); (Haematoxylin and eosin, X400) (b)

vimentin, variable positivity for cytokeratin (CK) and soluble in saturated 100% ammonium sulphate (S100). Integrase interactor1 (INI1) shows reduced expression. Methylation inhibited binding protein 1 (MIB1) labelling index was 35%–40%. Glial fibrillary acidic protein labels few background reactive stellate astrocytes and is negative in tumour cells. isocitrate dehydrogenase 1 (IDH1) point mutation, which contains an amino acid substitution where arginine is replaced by histidine at position 132 of IDH1 (R132H), epithelial membrane antigen (EMA), smooth muscle actin (SMA), desmin, cluster of differentiation 99 (CD99), FMS like tyrosine kinase1 (FLT1), signal transducer and activator of transcription 6 (STAT-6) and synaptophysin are negative. Alpha thalassemia/mental retardation X-linked (ATRX) shows retained expression. p53 shows variable patchy positivity. Trimethylation of histone H3 at lysine 27 (H3K27me3) shows loss of expression.

He was later referred for adjuvant radiotherapy. Magnetic resonance imaging of the brain before radiotherapy further showed a well-defined intra-axial altered signal intensity lesion of 2.2 cm × 1.8 cm × 3.3 cm in the left frontal lobe in the parasagittal location, the body of the corpus callosum, hyperintense on T1, T2, not suppressed on fluid-attenuated inversion recovery with peripheral enhancement on contrast

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study. Enhancement is also noted along the anterior aspect of the falx, which was suggestive of the residual or recurrent lesion (Figure 4). Whole-body ^{18}fluorodeoxy glucose positron emission tomography-CT (^{18}FDG PET-CT) showed 1.3 cm × 1.2 cm lesion that was metabolically quiescent. Single-positron emission computed tomography showed mildly glucoheptonate (GHA)-avid hypodense lesion in the left frontal lobe of the brain suggestive of residual/low-grade recurrence (Figure 5).

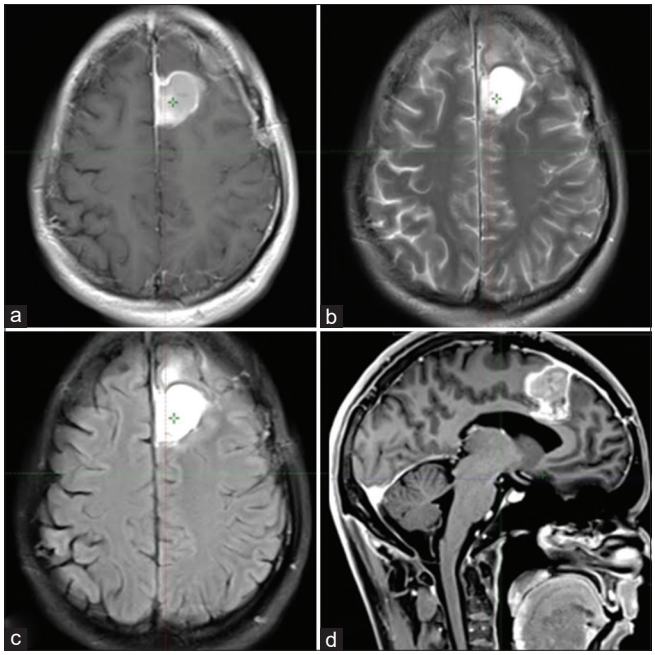

Figure 4: MRI of the brain T1 weighted (a), T2 weighted (b), T2 FLAIR axial view (c), T1 contrast-weighted sagittal view (d) showing residual lesion
MRI = Magnetic resonance imaging; FLAIR = Fluid-attenuated inversion recovery

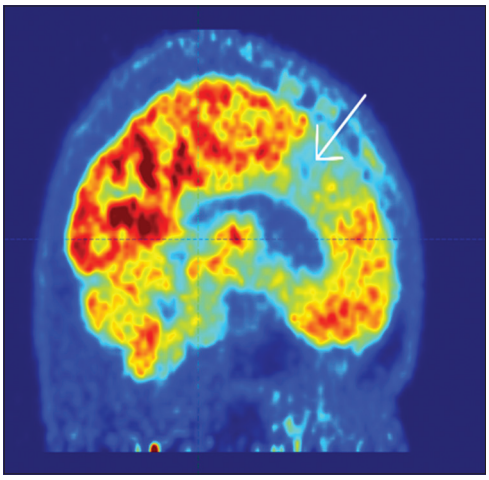

Figure 5: PET-CT of the brain showing a mild to non-FDG-avid lesion (arrow) in the left frontal lobe of the brain
PET-CT = Positron emission tomography-computed tomography; FDG = Fluorodeoxyglucose

The patient treated with adjuvant intensity modulated radiotherapy (IMRT) with a total dose of 60 Gy in 30 fractions, 2 Gy per fraction along with concurrent temozolomide 75 mg/m^2 chemotherapy. He tolerated the entire course of planned treatment well and now he is on regular follow-up and is asymptomatic for up to 2 years post-chemoradiotherapy.

DISCUSSION

Synovial sarcoma (also known as malignant synovioma) is a rare malignancy that affects primarily the soft tissue near the large joints of the extremities. Synovial sarcoma occurs in about 1–2/1,000,000 people a year. They occur most commonly in the third decade of life, with males being affected more often than females (1.2:1).[2] our subtypes of synovial sarcoma have been described: monophasic, monophasic epithelial, biphasic and poorly differentiated types. Synovial sarcoma is a well-defined clinical and morphological entity.[3] However, subsequent immunohistochemical and ultrastructural studies demonstrated that tumour cells do not share characteristics with normal synovium. Although commonly seen in the extremities, it may also arise primarily in a wide variety of organs, including the kidney, heart and lung. A small minority of patients develop symptoms secondary to metastatic lesions before diagnosis of the primary pathology. The rate for metastatic disease in synovial sarcoma ranges up to 33%.[4] Common sites of metastasis include lung, bone and lymph nodes. Most cases with intracranial disease have been reported as metastasis from the primary extracranial site, as in our case, which was initially presumed to be a potential metastatic lesion at the intracranial location. Metastatic workup using whole-body FDG PET-CT was advised to rule out any obvious primary extracranial pathology. This gave rise to the impression of the existence of a possible primary intracranial synovial sarcoma. Synovial sarcoma belongs to the category of soft-tissue sarcoma, which is known for its high FDG avidity, whereas in our case, the lesion appears to be metabolically quiescent, having no FDG avidity, which characterises its rarity of occurrence. The standard treatment for extracranial synovial sarcomas, as described in the literature, is a multimodality approach such as surgical resection with wide resection margins followed by adjuvant therapies, including radiation therapy and chemotherapy, which have demonstrated benefits for local recurrence and overall survival.[3] On the other hand, the treatment and prognosis of intracranial synovial sarcoma appear to be exceedingly unclear because of its rare occurrence. To the best of our knowledge, this case may be the second case report as reviewed in published English literature.

In conclusion, it may be suggested that the possibility of a very rare and aggressive malignant neoplasm like primary synovial sarcoma in the intracranial location should always be confirmed by adequate staging and metastatic workup using functional imaging like whole-body FDG PET-CT and IHC s before proceeding for multimodality treatments such as surgery and concurrent chemoradiotherapy in adjuvant settings. However, in the absence of optimal treatment regimen in published literature, for this rare neoplasm in the intracranial site, experience from the management of its most common extracranial counterpart may be extrapolated as used in our case for achieving a better tumour control and survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

The authors are faculty members/residents of Sri Venkateswara Institute of Medical Sciences, Tirupati, of which Journal of Clinical and Scientific Research is the official Publication. The article was subject to the journal's standard procedures, with peer review handled independently of these faculty and their research groups.

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