Case Report

Primary intracranial synovial sarcoma: A case report and review of literature

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INTRODUCTION

Primary intracranial synovial sarcoma (PrISS) is an unusual mesenchymal tumor within the intracranial space. Synovial sarcoma is an aggressive soft-tissue sarcoma that is generally seen in middle-aged patients, around the knee area, with a slight male predominance.¹¹ Unlike the systemic synovial sarcoma, PrISS carries a worse prognosis.¹⁸ Like their systemic counterparts, PrISS is classified into biphasic and monophasic variants based on the presence of epithelial and/or spindle cell components. The monophasic type is the more common subtype in systemic synovial sarcomas, though tumor progression is often poorly differentiated in either variant. Over the years, molecular cytogenticstics has proven to play an important role in confirming diagnosis in these tumors.
CASE REPORT

A 31-year-old male presented to us with severe persistent headache, right-sided weakness, and difficulty in speaking for 2 weeks at the height of the COVID pandemic. His admission GCS was E3V4M6 with Grade 4 power in the right upper and lower limbs. MRI brain showed the left frontal intra-axial heterogeneously enhancing tumor with an area of hemorrhage. Significant mass effect, perilesional edema, and midline shift were also noticed [Figure 1]. With a working diagnosis of high-grade glioma, surgery was planned. During his preoperative surgical workup, he was found to be COVID-19-positive. However, as patient’s clinical and neurological condition was deteriorating, it was deemed to be an emergency and gross total excision was performed. Intraoperatively, the tumor was friable with both solid cystic components and areas of hemorrhage with thrombosed veins in between. Clear plane between the brain and tumor was absent. A frozen section was not sent as the patient was COVID-positive and surgery was performed in specially designated operating room for COVID patients. Postoperatively, his recovery was smooth. His weakness and speech improved gradually.

Histopathological examination [Figure 2] revealed a spindle cell tumor with brisk mitosis and large areas of necrosis. Tumor cells were arranged in fascicles and had scant and ill-defined cytoplasm. High mitotic activity averaging 15–16/hpf was noted. IHC showed tumor cells staining for TLE-1 and focally for desmin. CD-99 was also diffuse and strongly positive. GFAP, STAT-6, SOX-10, EMA, S100, CD34, ATRX, CD31, NKX2.2, Caldesmon, and Bc2 were negative in the tumor cells. IHC and histological findings were suggestive of primary monophasic intracranial synovial sarcoma. For confirmation, the block was sent for fluorescent in situ hybridization (FISH) testing of SYT-SSX2 translocation, but was consistently reported negative in two reputed laboratories. In view of the classical histological features, further testing with RT-PCR was done and was positive for both translocations SYT-SSX1 and SYT-SSX2, confirming the diagnosis of synovial sarcoma.

Whole body PET CT was done to exclude a metastasis confirming the diagnosis as a PrISS. Postoperative imaging revealed a small residual disease. He received radiotherapy for the same (60 Gray in 30 fractions). At 6 months of follow-up, he was noticed to have tumor progression in spite of radiotherapy. [Figure 1] Patient died at 11 months after first surgery.

Figure 1: MRI imaging with gadolinium contrast depicting solid cystic tumor with dural attachment at convexity. (Top Row) Follow-up MRI at 6 months depicting tumor progression (Bottom Row).
DISCUSSION

Clinical symptoms and signs of PrISS depend on intracranial location of the tumor and may include dysphagia, pain, hoarseness, headache, or at times a palpable mass. The known sites of intracranial involvement are sellar region, skull base, cerebellum, and brain parenchyma. The preoperative diagnosis of PrISS is difficult and it may be thought to be an atypical meningioma, hemangiopericytoma, or a high-grade glioma. Synovial sarcoma may sometimes be confused with malignant meningioma because of their similar histological and immunohistochemical appearance (vimentin+, EMA+, and cytokeratin+). Most synovial sarcomas strongly coexpress CD99 and TLE-1, a pattern is highly distinct because it is generally not encountered in anaplastic meningiomas. FISH and RTPCR methods are routinely used for detection of the characteristic chromosomal translocation SYT-SSX t (X; 18) (p11.2; q11.2) including two fusion types SYT-SSX1 (X11p.23) and SYT-SSX2 (X11p.21).

Dural attachment and tumor origins

PrISS is an aggressive tumor associated with rapid neurological deterioration. Although “synovial” sarcoma has a predilection for particular areas, it is not actually associated with any synovial structure. Despite its name (a misnomer), it is no longer considered to be histologically derived from the synovium but rather from primitive mesenchymal cells. MRI picture may often be similar to high-grade gliomas, except for the dural attachment which has been noticed in all reported cases till date. [Table 1] Dura is embryological mesenchymal in origin and neoplastic transformation of mesenchymal tissue or embryonic mesenchymal cell rests may lead to PrISS.

Review of the literature [Table 1]

To the best of our knowledge, till date, 27 patients have been reported to have PrISS in English literature. Average age of patients reported till date is 34.9 years. There was no sex predilection. The lesion locations mainly included the frontal convexity dura (n = 6), parietal convexity dura (n = 4), cerebellum (n = 3), anterior skull base (n = 3), sellar region (n = 3), petroclival region (n = 2), and temporal lobe dura (n = 2). Occipital lobe, lateral ventricle, third ventricle, and parafalcine locations were observed in one case each. [Table 1] Unlike its systemic counterpart where monophasic variant is more predominant, both subtypes have been reported equally for PrISS. Supratentorial location was predominant and dural attachment was noted in all cases reported. FISH has been predominantly used for cytogenetic confirmation of the chromosomal translocation. RTPCR is rarely used for confirmation of the translocation. To the best of our knowledge, this case is the first such report in PrISS where FISH was negative and RTPCR was positive.

Molecular cytogenetics: FISH versus RTPCR

Heuvel et al. compared the diagnostic accuracy of RT-PCR and FISH for synovial sarcomas. They verified SYT-SSX1/SSX2 gene fusions and FISH analysis for SYT gene breaks on 50 specimens of formalin-fixed and paraffin embedded synovial sarcomas. RTPCR had a sensitivity of 94%; FISH had a sensitivity of 82%. A combined sensitivity of 100% was noted. If FISH is negative, RT-PCR has to be done to rule out diagnosis. Although the sensitivity of an RT-PCR test is greater than that of FISH, a FISH test is cheaper and capable of diagnosing gene fusion in majority of the cases, retaining it to be the first choice to detect the gene fusion. However, if FISH test is negative, RT-PCR test is mandatory.

Intratumoral hemorrhage in PrISS

PrISS has been reported to be a mimicker of spontaneous intracranial hemorrhage. Our case also presented with intracranial bleed with rapid neurological deterioration. Abnormal perilesional edema and contrast-enhancement on MRI helped are suspecting presence of intracranial tumor. Intratumoral hemorrhage in these patients is most likely to be due to abnormal neoplastic neovascularization in tumor progression. However, our patient was COVID-19-positive and a vasculitic thrombotic event secondary to the viral infection could have precipitated the hemorrhage in the neoplastic cells.
Treatment and outcome

The treatment for systemic synovial sarcoma is surgical resection with wide margins followed by radiotherapy. Wide margins are difficult to achieve in PrISS in comparison to systemic synovial sarcomas. Standard treatment for PrISS is not yet optimized though, adjuvant therapies including radiation have demonstrated benefits for local recurrence and prognosis. Long-term outcomes in PrISS are unknown due to paucity of data. The overall survival for patients with systemic (not intracranial) localized synovial sarcoma was 51% at 10 years. In contrast, PrISS has an overall survival of 15.5 months only with surgery and radiation based on a series of 16 patients. Our patient had a survival of only 11-month postdiagnosis in spite of radiotherapy.

CONCLUSION

PrISSs are unusual aggressive dural-based mesenchymal tumors with poor prognosis. They are seen most commonly...
in supratentorial compartment in 3rd or 4th decade of life. Radiological appearance of a supratentorial solid cystic lesion with dural attachment and intratumoral hemorrhage should raise suspicion for PrISS. Molecular cytogenetics including FISH and RTPCR are required for confirming the diagnosis, though FISH seems to have lower sensitivity than RTPCR and can yield false negative results.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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