Recurrent Primary intracranial synovial sarcoma,A case report and review of the literature

Manizhe Ataee Kachuee[1], Iman Mohseni[1], Alireza Sahranavard[2], and Zhale Tabrizi[1]

[1]Iran University of Medical Sciences
[2]Isfahan University of Medical Sciences

May 17, 2022

Abstract

Synovial sarcoma(SS) occurs in various parts of the body,predominantly in extremities.It also occurs in organs without synoviom. Intracranial disease, has been reported as metastasis,but primary intracranial SS has been reported rarely. we report a pateint with hemiplegia and a mass on brain CT. pathology showed SS with no extracranial pathology

Recurrent Primary intracranial synovial sarcoma,A case report and review of the literature

1-Ataee Kachuee Manizhe,Department of Radiology,Firouzgar Hospital,Iran university of medical science,Tehran,Iran,Manizhe.ataei64@gmail.com

2-Iman Mohseni,Department of Radiology,Firouzgar Hospital,Iran university of medical science,Tehran,Iran,Imannn24@yahoo.com

3-Alireza Sahranavard,Department of orthopaedics surgery,Alzahra hospital,Isfahan university of medical science,Isfahan,Iran,Dr.sahranavard@gmail.com

4-corresponding: Zhale Tabrizi,Department of Radiology,Iran university of medical science,Tehran,Iran,zh.tb_1991@yahoo.com

Key clinical message

Synovial sarcoma is one of the most common sarcoma of soft tissue,and it mostly occurs in lower extrimities.It can cause metastasic disease especialy to the lungs,bones and lymphnodes.Metastasis to the brain is very rare.Primary intracranial synovial sarcoma has also been reported in extremely rare cases.In this paper,we study the case of a 28-year-old man with primary intracranial synovial sarcoma.

Abstract

Synovial sarcoma(SS) occurs in various parts of the body,predominantly in extremities.It also occurs in organs without synovial structures. Intracranial disease, has been reported as metastasis,but primary intracranial SS has been reported rarely. we report a pateint with hemiplegia and a mass on brain CT. pathology showed SS with no extracranial pathology.

Keywords

Synovial sarcoma,metastatic intracranial synovial sarcoma,primary intracranial synovial sarcoma.

Introduction

Synovial sarcoma is the fourth most common sarcoma of the soft tissue,following liposarcoma,fibrous histiocytoma and rhabdomyosarcoma.With more prevelance in men than women.Most of the SS cases are

located in extrimities,predominantly lower extremites(1, 2). especially near large joints around the knee and thigh(3). Synovial sarcoma has an annual incidence of 2.5 per 100,000(4). Intracranial synovial sarcoma is very uncommom and has been reported as a metastasis lesion from synovial sarcoma in other parts of the body (3, 5-11).In this study, We report a case with no obvious primary extracranial pathology, with recurrent primary intracranial synovial sarcoma,which has been very rarely reported in literature.

Case presentation

The case study is devoted to investigating headache and left hemiplegia in a 28-year-old man since 10 days prior to coming to the emergency room.Neurological examination revealed left sided hemiplegia,without gait instability and ataxia.Brain computed tomography(CT) scan revealed an intra axial mass lesion with central necrotic component and surrounding vasogenic edema in right frontal white matter causing midline shift to the left side.(FIGURE 1) The patient underwent magnetic resonance imaging (MRI) with gadolinium contrast for more evaluation,and the findings were as mentioned bellow:An Intra axial well-circumscribed heterogenous mass with central necrosis and irregular peripheral enhancement and significant surrounding vasogenic edema in right frontal white matter.restriction of peripheral solid component of mass was also seen.(FIGURE 2)

The patient was taken to the operation room and underwent total excision of the mass.Many hours later the left hemiplegia was gradually improved.The histopathological assessment of the mass revealed neoplastic tissue composed of biphasic pattern: large irregular sheets of monotonous cells some with central necrosis set in fibroblastic stroma.Tumoral cells are plump and have very high N/C ratio.Frequent mitotic figures are present.

IHC staining show positive reactivity for TLE1,FEI1 and INI1 negative reactivity for Synaptophysin,Olig 2,SMA,CK CD 99.GFAP staining is nonspecific.Above findings are consistent with Synovial Sarcoma.PET scan didn't show any other metastatic disease. Abdominopelvic CT scan was also normal.

The patient was discharged 1 week after surgery without any neurological problems.

He underwent radiotherapy 1 month after his discharge.

4 months later,the patient came back to the neurology emergency room with left hemiplegia,he underwent CT scan findings revealed recurrence of the previous tumor(FIGURE3) ,so he was taken to operation room again and underwent total excision of the mass.The histopathological assessment showed Synovial sarcoma recurrence.PET scan was done again and there were no evidence of primary synovial sarcoma anywhere else or any other metastasis.The patient was discharged one week later without any neurological sign and symptoms.

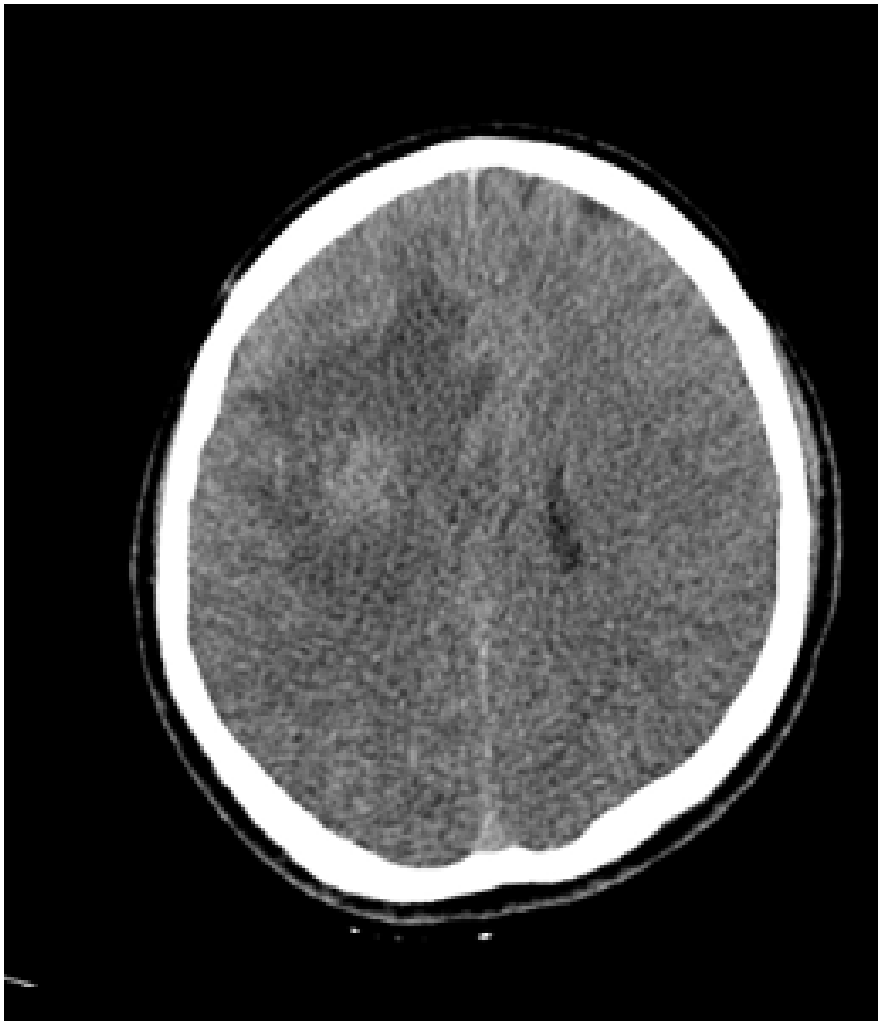
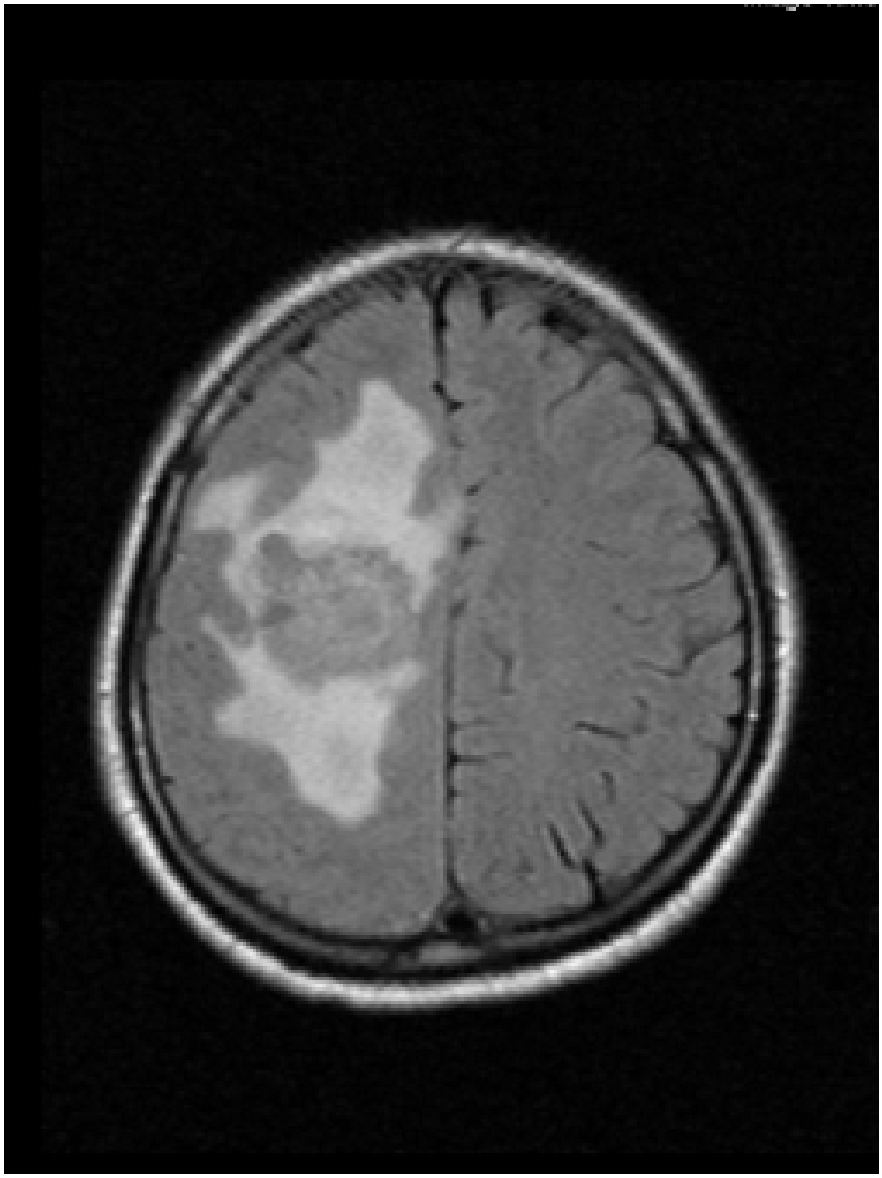
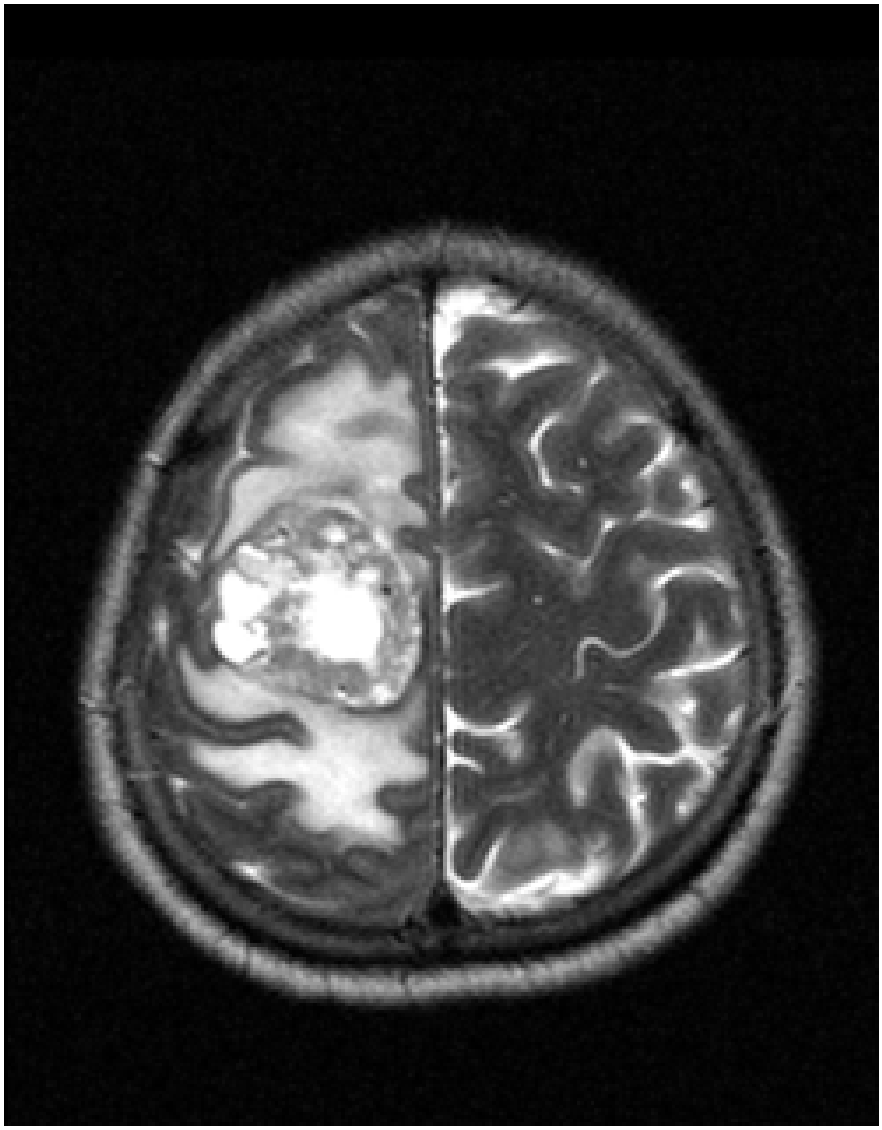
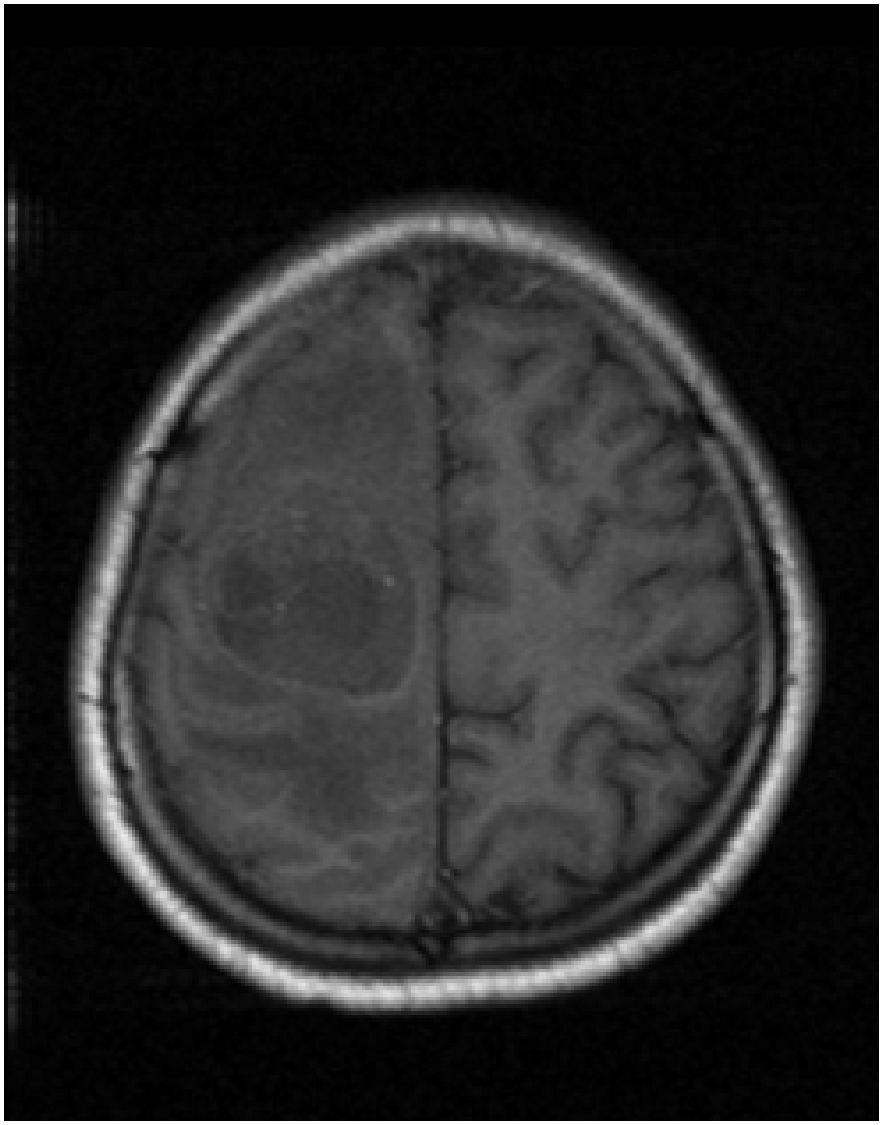


FIGURE1.Brain computed tomography(CT) scan revealed an intra axial mass lesion with central necrotic component and surrounding vasogenic edema in right frontal white matter causing midline shift to the left side.







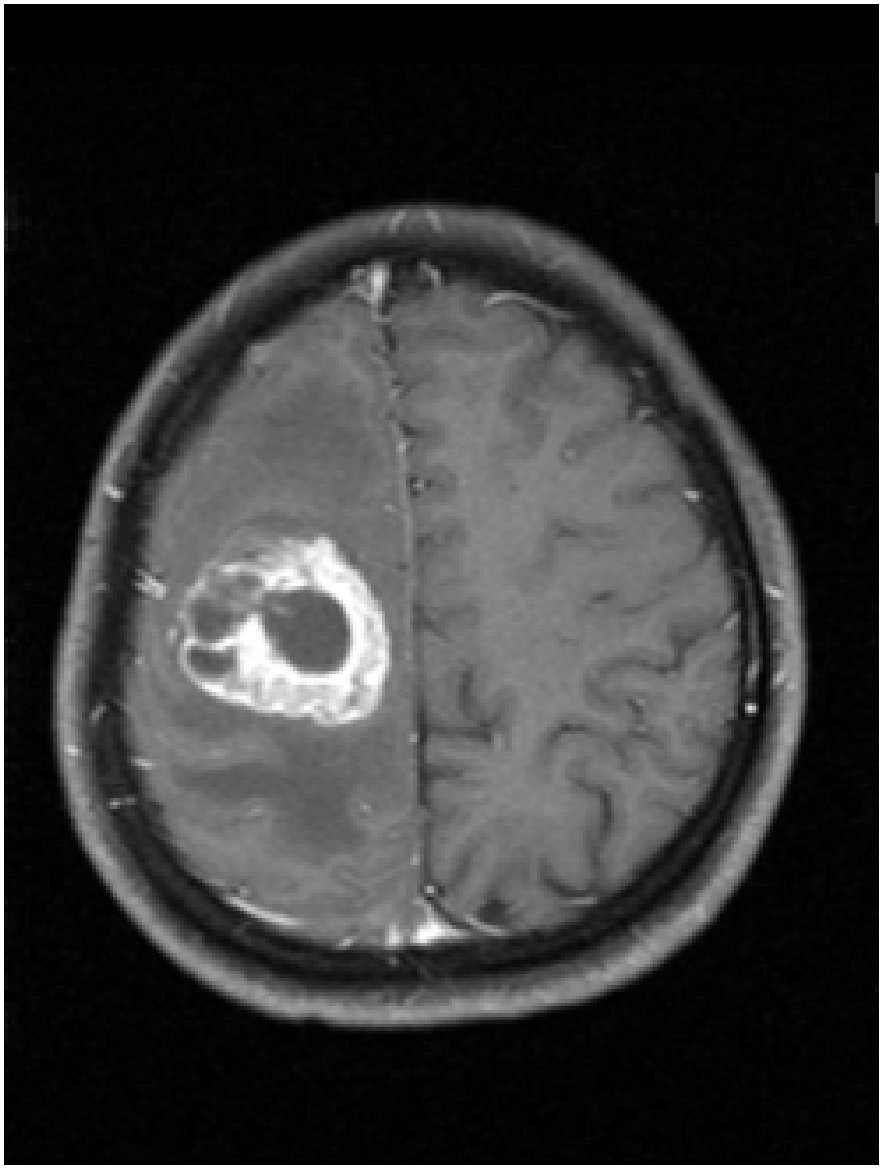
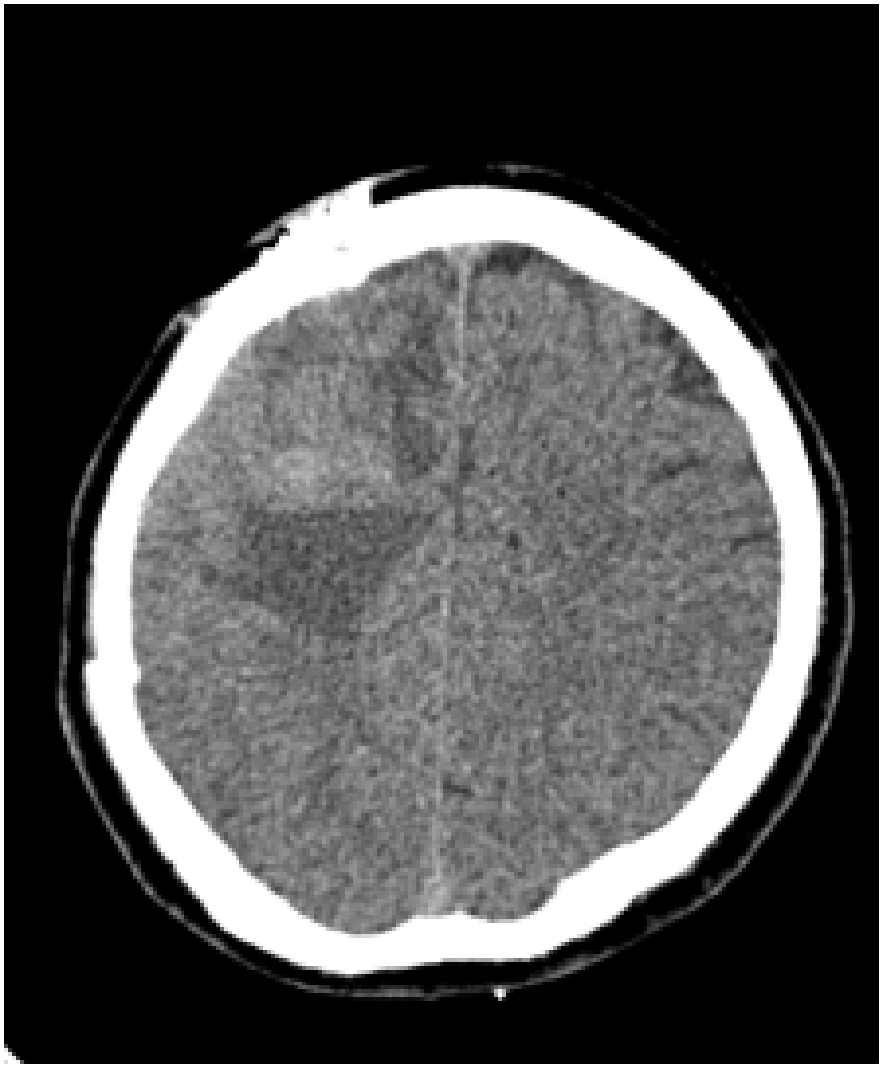


FIGURE2. magnetic resonance imaging (MRI) with gadolinium contrast showed an Intra axial well-circumscribed heterogenous mass with central necrosis and irregular peripheral enhancement and significant surrounding vasogenic edema in right frontal white matter.restriction of peripheral solid component of mass was also seen.



. FIGURE 3.Brain CT scan showed recurrence of previous tumor

Discussion

Synovial sarcoma is a form of soft tissue sarcoma that occurs mostly in young and middle-aged adults and has more tendency to occurs in extrimities,however,it can occurs in children and old individuals and involve other parts of the body (12, 13).It is believed that 5-10 percent of soft tissue sarcomas are synovial sarcoma.The tumor is closely related to tendon sheath,tendon,joint capsule and bursal structures (14).It may also arise primarily in other parts of the body,such as heart,kidney and lungs (3).There are four subtypes of synovial sarcoma,as mentioned bellow: monophasic, monophasic epithelial, biphasic, and poorly differentiated(round cell tumors)(3, 15). Classical synovial sarcoma has a biphasic morphology and it's pathological findings are sheets of spindle cells and sharply segregated epithelial cells forming gland-like areas. A second form of synovial sarcoma is monophasic containing only a sarcomatous component. Most of the synovial sarcomas are of these two forms mentioned above (4).Because of the tumor resemblance to synovial tissue under light microscope,it was named "Synovial Sarcoma" by Sabrazes in 1934 (16).However,the cellular origin of this tumor is possibly from neural crest-derived malignant peripheral nerve cells (3). Chromosomal translocation of t(X;18)(p11.2;q11.2) can be investigated in over 95% of synovial sarcomas (4). the treatment of choice for primary tumors is excisional surgery,while metastatic synovial sarcoma can't be treated with

excisional surgery but chemotherapy and radiotherapy may help them (17).In some patients,synovial sarcoma is diagnosed when it reach out other organs such as lungs,bone and lymphnodes(5, 8, 18).Intracranial synovial sarcoma is very rare and in most cases are reported as a metastasis from synovial sarcoma (19). Synovial sarcoma presenting as an intracranial has been reported very rarely,as mentioned bellow:

1-Kleinschmidt-DeMasters et Al reported the case of a 19-year-old woman with primary synovial sarcoma of the third ventricle (20),

2- Bettio et Al reported a 36-year-old man with a large intracranial tumor involving the cranial base, with invasion to the sellar region and sphenoidal sinus (21) .

3-Mohit Patel et Al reported a primary intracranial synovial sarcoma as a right parietal heterogeneous, hyperdense mass with a large medial hematoma in a 21-year- old man (19) .

4-Yang-Yang Wang et Al reported a 35- year-old man with left intracerebral lesions with hemorrhage and left middle cerebral artery arteriovenous malformation, respectively (22).

The clinical manifestations of primary intracranial Synovial Sarcoma are nonspecific,such as headache, nausea, vomiting, hemiplegia.Radiologic features of primary synovial sarcoma are also nonspecific and the final diagnosis is made by pathological and immunohistochemistry features (22).

Our patient was a 28-year-old man with headache and left hemiplegia.Neurological examination revealed left sided hemiplegia,without gait instability and ataxia.Brain computed tomography(CT) scan revealed an intra axial mass lesion with central necrotic component and surrounding vasogenic edema in right frontal white matter causing midline shift to the left side.The patient underwent magnetic resonance imaging (MRI) with gadolinium contrast for more evaluation,and the findings were as mentioned bellow:An Intra axial well-circumscribed heterogenous mass with central necrosis and irregular peripheral enhancement and significant surrounding vasogenic edema in right frontal white matter.restriction of peripheral solid component of mas was also seen.The patient was taken to the operation room and underwent total excision of the mass.Many hours later the left hemiplegia was gradually improved.The histopathological assessment of the mass revealed neoplastic tissue composed of biphasic pattern: large irregular sheets of monotonous cells some with central necrosis set in fibroblastic stroma.Tumoral cells are plump and have very high N/C ratio.Frequent mitotic figures are present.

IHC staining show positive reactivity for TLE1,FEI1 and INI1 negative reactivity for Synaptophysin,Olig 2,SMA,CK CD 99.GFAP staining is nonspecific.Above findings are consistent with Synovial Sarcoma.PET scan didn't show any other metastatic disease.Abdominopelvic CT scan was also normal.

Conclusion

Synovial sarcoma is one of the most common sarcoma of soft tissue,and it mostly occurs in lower extrimities.It can cause metastasic disease especialy to the lungs,bones and lymphnodes.Metastasis to the brain is very rare.Primary intracranial synovial sarcoma has also been reported extremely rare.In this paper,we present a 28-year-old man with primary intracranial synovial sarcoma.It can misdiagnosed with other intracranial masses.So it is important to confirm the diagnosis with pathology and resect the tumor and start radiotherapy treatment if needed.

ACKNOWLEDGMENTS:

None.

Patient consent: Written informed consent for publication was obtained from the patient.

ETHICAL APPROVAL: All ethics standards and consent were obtained for this manuscript

conflicts of Interests: The Authors declare that there are no competing interests.

Data availability statement

Data sharing not applicable – no new data generated Data sharing is not applicable to this article as no new data were created or analyzed in this study.

Refrences

1. Ladenstein R, Gadner H, Treuner J, Koscielniak E, d'Oleire F, Keim M, et al. Synovial sarcoma of childhood and adolescence. Report of the german CWS-81 study. Cancer. 1993;71(11):3647-55.

2. van der Heide HJ, Veth RP, Pruszczynski M, Wobbes T, van Hoese QG, Lemmens JA. Synovial sarcoma: oncological and functional results. European Journal of Surgical Oncology (EJSO). 1998;24(2):114-9.

3. Nuwal P, Dixit R, Shah NS, Samaria A. Primary monophasic synovial sarcoma lung with brain metastasis diagnosed on transthoracic FNAC: Report of a case with literature review. Lung India: Official Organ of Indian Chest Society. 2012;29(4):384.

4. Scully SP, Temple H, Harrelson JM. Synovial sarcoma of the foot and ankle. Clinical Orthopaedics and Related Research (1976-2007). 1999;364:220-6.

5. Przkora R, Vogel P, Ullrich OW, Knüchel R, Jauch KW, Bolder U. Synovial sarcoma—unusual presentation with cerebral hemorrhage. Archives of orthopaedic and trauma surgery. 2003;123(7):376-8.

6. Flannery T, Kano H, Niranjan A, Monaco III EA, Flickinger JC, Kofler J, et al. Gamma knife radiosurgery as a therapeutic strategy for intracranial sarcomatous metastases. International Journal of Radiation Oncology* Biology* Physics. 2010;76(2):513-9.

7. Yoshida S, Morii K, Watanabe M, Saito T. Brain metastasis in patients with sarcoma:: An analysis of histological subtypes, clinical characteristics, and outcomes. Surgical neurology. 2000;54(2):160-4.

8. Siegel HJ, Dunham WH, Lopez-Ben R, Siegal GP. Intracranial metastasis from synovial sarcoma. Orthopedics (Online). 2008;31(4):405.

9. Grossman R, Ram Z. Recursive partitioning analysis (RPA) classification predicts survival in patients with brain metastases from sarcoma. World Neurosurgery. 2014;82(6):1291-4.

10. Kaufman J, Tsukada Y. Synovial sarcoma with brain metastases. Report of a case responding to supervoltage irradiation and review of the literature. Cancer. 1976;38(1):96-9.

11. Baptista AM, Camargo OPd, Croci AT, de Oliveira CRG, Azevedo Neto RSd, Giannotti MA, et al. Synovial sarcoma of the extremities: prognostic factors for 20 nonmetastatic cases and a new histologic grading system with prognostic significance. Clinics. 2006;61(5):381-6.

12. Tateishi U, Hasegawa T, Beppu Y, Satake M, Moriyama N. Synovial sarcoma of the soft tissues: prognostic significance of imaging features. Journal of computer assisted tomography. 2004;28(1):140-8.

13. Shmookler BM. Retroperitoneal synovial sarcoma: a report of four cases. American Journal of Clinical Pathology. 1982;77(6):686-91.

14. Rajwanshi A, Srinivas R, Upasana G. Malignant small round cell tumors. Journal of Cytology/Indian Academy of Cytologists. 2009;26(1):1.

15. Rajwanshi A, Srinivas R, Upasana G. Malignant small round cell tumors. J Cytol 26: 1–10. Article; 2009.

16. Nosrati A, Shafiee S, Aazam E. Case report: A 27 years old woman with brain metastases of synovial sarcoma. International Journal of Medical Investigation. 2016;5(2):69-72.

17. Murray PM. Soft tissue sarcoma of the upper extremity. Hand clinics. 2004;20(3):325-33.

18. Rong R, Doxtader EE, Tull J, de la Roza G, Zhang S. Metastatic poorly differentiated monophasic synovial sarcoma to lung with unknown primary: a molecular genetic analysis. International journal of clinical and experimental pathology. 2010;3(2):217.

19. Patel M, Li L, Nguyen HS, Doan N, Sinson G, Mueller W. Primary intracranial synovial sarcoma. Case Reports in Neurological Medicine. 2016;2016.

20. Kleinschmidt-Demasters BK, Mierau GW, Sze C-I, Breeze RE, Greffe B, Lillehei KO, et al. Unusual dural and skull-based mesenchymal neoplasms: a report of four cases. Human pathology. 1998;29(3):240-5.

21. Bettio D, Rizzi N, Colombo P, Bianchi P, Gaetani P. Unusual cytogenetic findings in a synovial sarcoma arising in the paranasal sinuses. Cancer genetics and cytogenetics. 2004;155(1):79-81.

22. Wang Y-Y, Li M-L, Zhang Z-Y, Ding J-W, Xiao L-F, Li W-C, et al. Primary intracranial synovial sarcoma with hemorrhage: A case report. World journal of clinical cases. 2021;9(29):8871.