Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pathology

Intracranial Solitary Fibrous Tumour: Case Report

Ihssan Elouarith[1*], Leila Benbella[1], Fouad Zouaidia[1], Ahmed Jahid[1], Zakia Bernoussi[1], Kaoutar Znati[1]

[1]Pathology Department, Ibn Sina Hospital, Mohammed V University, Rabat, Morocco

DOI: 10.36347/sjmcr.2023.v11i08.008 | **Received:** 19.06.2023 | **Accepted:** 25.07.2023 | **Published:** 02.08.2023

***Corresponding author:** Ihssan Elouarith
Pathology Department, Ibn Sina Hospital, Mohammed V University, Rabat, Morocco

Abstract **Case Report**

Intracranial solitary fibrous tumor is a rare mesenchymal tumour. The diagnosis is based on the histological study given the clinical and radiological character that can simulate other benign or malignant pathologies especially meningioma. We report the case of a patient with an intracranial solitary fibrous tumor. We aim to discuss the clinical, radiological, histological and immunohistochemical features of Intracranial solitary fibrous tumor as well as the new grading system reported in the fifth edition of the WHO classification of central nervous system tumours.

Keywords: Solitary fibrous tumor – Skull - Immunohistochemical study.

INTRODUCTION

Solitary fibrous tumor is a mesenchymenl, fibroblastic neoplasm characterized by NAB2-STAT6 gene rearrangement. Intracranial localization is rare and few cases have been reported since its first description. We present a rare case of an intracranial solitary fibrous tumour in a 53-year-old patient, in whom the radiological appearance suggested a meningioma.

CASE REPORT

53-year-old male patient with no particular history. He consulted in our structure for headaches becoming disabling and resistance to analgesics for 6 days. A cerebral CT scan was performed, it showed a left parieto-frontal cerebral mass measuring 5.2x1cm. A subsequent brain MRI showed a well limited lesion isointense on T1-weighted imaging and hyper-intense on T2-weighted imaging. After the intravenous administration of the contrast agent gadopentetate dimeglumine, the mass shows homogeneous enhancement. Based on these radiological data, a meningioma was suspected. The patient underwent complete surgical resection of the mass.

The surgical specimen was then subsequently sent for anatomopathological study. We received the resected specimen measuring 5,5x1,4cm. It was a well-limited, non encapsulated masse. This mass has a gray-white appearance with fibrous and firm consistency on the cut surface.

The histological study showed a tumoral proliferation of variable cellularity. In the hypercellular zones, the cells are of haphazard arrangement. They are ovoid in appearance, fusiform in places, and devoid of cytonuclear atypia. The stroma is fibrous and comprises large and dilated vessels with a thickened and hyalinized wall. The hypocellular areas are rich in fibrous structures with myxoid changes. No tumor necrosis or mitotic figures were observed. In view of this aspect, an immunohistochemical study was carried out. It showed positivity of the tumor cells by the STAT6 marker, but negative staining by the anti-EMA and anti-CD34 markers. The KI67 proliferation index is very low. A diagnosis of a solitary fibrous tumor was retained. It is classified as grade 1 according to the 2021 WHO classification.

The post-operative follow-up and after 6 months of the patient was unremarkable and the patient is currently disease-free.

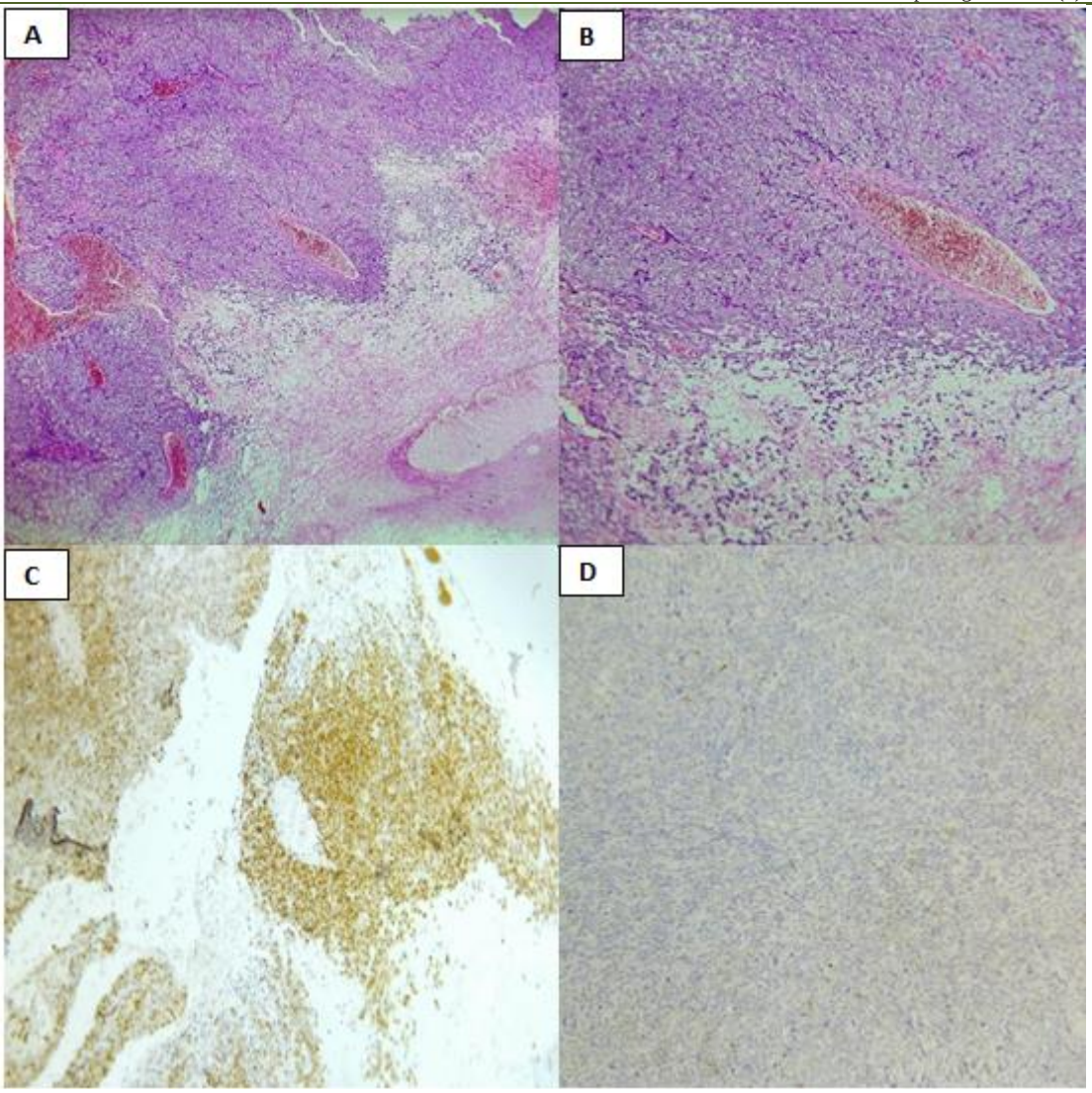

Figure 1: Histological appearance and immunohistochemical profile of a solitary fibrous tumor : A and B- Tumoral proliferation with variable cellularity, made up of ovoid to spindle-shaped cells and comprising large vessels with thick and hyalinized walls. (HE_40 et _100) C- Tumor cells are positive for the STAT6 marker (x100) D- but negative for the EMA marker (x100)

DISCUSSION

Solitary fibrous tumor is a mesenchymenl neoplasm originate from fibroblastic cells and characterized by NAB2-STAT6 gene rearrangement. It was first defined in 1931 by Klemperer and Rabin [1].

This tumor is most often located at the thoracic level, specifically in the pleura and the mediastion. Other extrathoracic localizations, including intracranial ones, remain rare.

In 1996, it was first reported that Solitary fibrous tumor can arise in the central nervous system, and since then cases of published Solitary fibrous tumor have increased [2].

The exact frequency of intracranial Solitary fibrous tumor has been very difficult to determine because of its consistent nomenclature. Thus, because of its rarity, Solitary fibrous tumor has been grouped with mesenchymal tumors in recent statistical reports, such as the report published in 2019 by the central brain tumor registry of the united states [3].

Solitary fibrous tumor most often affects elderly patients with a peak incidence in patients aged between 50 and 70 years. However 18% of patients were under 40 years old. No significant gender difference has been reported. The most common intracranial locations are represented by skull base, falcine and parasagittal locations. Spinal locations of Solitary fibrous tumor are estimated at around 10%. However, pineal gland, sellar region and cerebelopontine angle are rare localizations [3].

The clinical symptoms of solitary fibrous tumor are not specific but depend on its location. Thus, patients with solitary fibrous tumor localized in the

cerebral hemisphere may present a headache, nausea, vomiting, epileptic seizure and many other symptoms. On the other hand, intracranial pressure as well as mass effect can occur in case of large tumor [4, 5].

Radiologically, the diagnosis of solitary fibrous tumor is very difficult and is often confused with meningiomas, given the absence of specific radiological signs of this tumour. Generally, a brain computed tomography scan shows a solitary mass with irregular but clear boundries and generally with uniform density. However, sometimes, areas with lower density may include necrosis, while areas including some collagen fiber or rich in fusiform cells may be respectively slightly higher or higher in density [6]. On MRI, solitary fibrous tumour (SFT) is usually isointense on T1-weighted images, and show iso or hypointensity on T2-weighted images. After the intravenous administration of the contrast agent, the mass shows homogeneous enhancement [7, 8]. In a few reports, some authors have revealed non-specific radiological signs on MRI, which may point towards the suspicion of SFT. This is the case, for example, of the commonly named yin-yang sign which is presented, on postcontrast T2-weighted scans, as an alternation of zones of hyperintensity and hypointensity [9, 10]. This appearance probably reflects the histological appearance of the TFS characterized by a variations of cellularity and by a characteristic vascularization called “staghorn vascularity” [11].

Histologically, the features of intracranial solitary fibrous tumour are similar to those in other locations. The tumor consists of haphazardly arranged fusiform spindled to ovoid monomorphic cells with indistinct cell borders. Cells may also be arranged in short and ill-defined fascicles. They are admixed with dilated, branching, hyalinized and thin walled vessels. They are called staghorn-like or hemangiopericytoma-like vasculature.

SFT is also characterized by a variable histological appearance with alternating hypercellular and hypocellular zones which is essentially made up of an abundant stroma rich in collagen fibers. Myxoid stroma, lipomatous component and giant cells may be present.

In the 5th edition of the WHO classification of the central nervous system tumours (2021), histological grading was reported. It is based on mitotic activity and the presence of tumor necrosis. Thus, we distinguish:

CNS WHO grade 1: <2,5 mitosis/mm2

CNS WHO grade 2: >= 2,5 mitosis/mm2 without necrosis

CNS WHO grade 3: >= 2,5 mitosis/mm2 with necrosis

Immunohistochemically, CD34 is generally diffusely positive in grade1 TFS. However this marker is non-specific and may be positive in other types of tumors. STAT6 is a highly sensitive and specific marker which allows differentiating SFT from a large number of tumors that pose the problem of differential diagnosis, such as meningiomas.

Other markers, such as SMA, desmin and EMA may be focally and weakly positive [3].

The main differential diagnosis of intracranial SFT includes different meningothelial and soft tissue tumors, including Fibrous meningioma, Ewing sarcoma, monophasic synoial sarcoma and malignant peripheral nerve sheath tumour. Immunohistochemistry plays a key role in distinguishing between these different neoplasms. Thus, unlike TFS which is CD34 and STAT6 positive, fibrous meningioma and Ewing's sarcomas are STAT6 negative and typically express EMA and CD99 respectively. On the other hand, a positive EMA and TLE1 and/or an SS18 rearrangement detected by FISH, argues rather in favour of a monophasic synovialosarcoma. Finally, malignant peripheral nerve sheath tumours are CD34 and STAT6 negative but they focally express PS100 and SOX10.

Surgical resection is the treatment of choice. Systemic treatment with chemotherapy or radiotherapy is used if necessary. The surgical strategy depends on both the location and the size of the tumour. However, radical resection is recommend to prevent tumour recurrence and metastasis [12].

It has been demonstrated by two studies carried out on patients with cerebral TFS proven by their expression of STAT6 or even by the detection of the NAB2-STAT6 gene fusion, that TFS has a high potential for recurrence and metastasis [3].

CONCLUSION

Intracranial solitary fibrous tumor is a rare neoplasm that can be differentially diagnosed with other mesenchymal tumors, mainly meningioma. Through our work, we highlight the role of the pathologist in the positive diagnosis of this rare entity and especially given the non-specificity of the radiological aspects.

REFERENCES

1. Klemperer, P. R. C. B. (1931). Primary neoplasms of the pleura. A report of five cases. *Arch Pathol*, *11*, 385-412.
2. Carneiro, S. S., Scheithauer, B. W., Nascimento, A. G., Hirose, T., & Davis, D. H. (1996). Solitary fibrous tumor of the meninges: a lesion distinct from fibrous meningioma: a clinicopathologic and immunohistochemical study. *American journal of clinical pathology*, *106*(2), 217-224.
3. WHO Classification of Tumours Editorial Board. (2021). *World Health Organization Classification of Tumours of the Central Nervous*

System. 5th ed. Lyon: International Agency for Research on Cancer.

4. Chen, H., Zeng, X. W., Wu, J. S., Dou, Y. F., Wang, Y., Zhong, P., ... & Wang, X. Q. (2012). Solitary fibrous tumor of the central nervous system: a clinicopathologic study of 24 cases. *Acta neurochirurgica*, *154*, 237-248.
5. Vassal, F., Manet, R., Forest, F., Camdessanche, J. P., Péoc'h, M., & Nuti, C. (2011). Solitary fibrous tumors of the central nervous system: report of five cases with unusual clinicopathological and outcome patterns. *Acta neurochirurgica*, *153*, 377-384.
6. Sun, L. J., Dong, J., Gao, F., Chen, D. M., Li, K., Liu, J., ... & Yang, X. P. (2019). Intracranial solitary fibrous tumor: Report of two cases. *Medicine*, *98*(17), e15327.
7. Clarençon, F., Bonneville, F., Rousseau, A., Galanaud, D., Kujas, M., Naggara, O., ... & Chiras, J. (2011). Intracranial solitary fibrous tumor: imaging findings. *European journal of radiology*, *80*(2), 387-394.
8. Weon, Y. C., Kim, E. Y., Kim, H. J., Byun, H. S., Park, K., & Kim, J. H. (2007). Intracranial solitary fibrous tumors: imaging findings in 6 consecutive patients. *American journal of neuroradiology*, *28*(8), 1466-1469.
9. Kim, K. A., Gonzalez, I., McComb, J. G., & Giannotta, S. L. (2004). Unusual presentations of cerebral solitary fibrous tumors: report of four cases. *Neurosurgery*, *54*(4), 1004-1009.
10. Lawlor, M. W., Nielsen, G. P., & Louis, D. N. (2008). Malignant solitary fibrous tumour of the meninges with marked amianthoid fibre deposition. *Neuropathology and applied neurobiology*, *34*(5), 569-572.
11. Bisceglia, M., Dimitri, L., Giannatempo, G., Carotenuto, V., Bianco, M., Monte, V., ... & Magro, G. (2011). Solitary fibrous tumor of the central nervous system: report of an additional 5 cases with comprehensive literature review. *International Journal of Surgical Pathology*, *19*(4), 476-486.
12. Hori, E., Kurimoto, M., Fukuda, O., Takahashi, C., Nagai, S., Oya, T., & Endo, S. (2007). Recurrent intracranial solitary fibrous tumor initially diagnosed as hemangiopericytoma. *Brain Tumor Pathology*, *24*, 31-34.