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Case Report

Olfactory groove monophasic sinovial sarcoma and von Recklinghausen's disease: A case report and literature review

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ABSTRACT

Background: Soft-tissue sarcomas are a rare and diverse group of neoplastic lesions. They represent only 1% of malignant tumors in adults and 15% in children. Synovial sarcoma (SS) is a type of soft-tissue sarcoma, accounting for 5–10% of cases, and commonly affecting extremities. Diagnosis, treatment, and prognosis remain challenging especially when localized in uncommon areas, such as intracranial lesions.

Case Description: A 13-year-old male patient with a clinical history of neurofibromatosis Type I (NF1) presenting holocranial headache with jet vomiting and apathy 2 days before admission, without neurological deficits and/or focal findings. On magnetic resonance imaging: an extra-axial infiltrative lesion with contrast uptake at the base of the skull in the olfactory groove topography. After total tumor resection, the anatomopathological examination showed monophasic SS. The patient returned after 6 months with similar symptoms, and the lesion recurred and was reoperated. Unfortunately, 7 months after the second surgery, the patient died.

Conclusion: SS can occur extraarticulously and with a variable clinical presentation and poor prognosis despite adjuvant therapies with radiotherapy and chemotherapy. In individuals with clinical history of NF1, there is still no direct correlation between the two manifestations, although current descriptions are suggestive of a possible interaction.

Keywords: Case report, Intracranial, Monophasic synovial sarcoma, Neurofibromatosis Type 1

INTRODUCTION

Soft-tissue sarcoma comprises a group of rare neoplastic lesions with mesenchymal origin, with about 60 distinct possible diagnoses within this group of tumors. In addition to the complexity and variety of lesions described, this set represents only 1% of malignant tumors in adults and about 15% in children.[3,30] Within this group, different types of lesions can be described and may affect nerve sheaths, blood vessels, muscles, and other connective tissues, among the lesions described is synovial sarcoma (SS). SS is a rare neoplasm that represents about 5–10% of soft-tissue sarcomas, being more common in adolescent and young adults and that, despite the suggestive name related to development in synovia, this type of tumor can affect any part

of the body, especially described in extremities.[24,28] With an incidence of 800–1000 cases described per year in the USA, the diagnosis, treatment, and prognosis of these patients remain challenging.[27]

Cases of metastatic intracranial SS are well described in the literature, and one of the possible sites of neoplastic dissemination is the central nervous system.[20] However, the description of cases of primary intracranial SSs still remains a fairly rare entity.[1,2,12,14-18,21-23,31,32,35,36] The present work is a case report of a final diagnostic patient of monophasic SS (MSS) in olfactory groove and with neurofibromatosis Type 1 (NF1). It is well-described that patients with chromosome 17 disorder with NF1, also known as von Recklinghausen's disease, have a predisposition to the development of central nervous system tumors.[10] However, even with this close correlation between intracranial neoplasms and the NF1 mutation, the existence of research and reports involving primary intracranial synovium sarcomas in patients with NF1 still remained scarce, although translational studies have already demonstrated the possible participation of these mutations in the generation of soft-tissue sarcomas.[7]

MATERIALS AND METHODS

The article presents a case report with a review of the literature. The literature review focuses on case reports of patients with primary intracranial SS. The selection of articles was based on an analysis of the PubMed database using the following keywords: "synovial sarcoma," "soft-tissue sarcoma," and "intracranial synovial sarcoma." To extend the search, the Boolean operators "AND" and "OR" were used to filter articles that were relevant to the study. No filter was used during the search or year limitations were imposed. The search was conducted by one of the researchers using advanced search resources provided by PubMed. To increase the number of publications, the search terms were crossed. The inclusion criteria were limited to case report articles about primary intracranial SS, articles of intracranial metastasis and other types of soft-tissue sarcoma were excluded from the study. Fifteen articles met the inclusion criteria and were used to compose the Table 1. Some of the selected articles were found by referencing other articles that had already been identified during the search and met the same inclusion criteria.

CASE REPORT

A 13-year-old male patient with a clinical history of NF1 was admitted from work presenting with a holocranial headache associated with jet vomiting and significant apathy for 2 days before admission. On physical examination, there was no neurological deficits or focal findings. On inspection, the presence of multiple café-au-lait spots, at least 6, and several dermal neurofibromas were identified, corroborating the previous diagnosis of NF1. On complementary magnetic resonance imaging investigation, a 4.0 × 3.8 cm infiltrative extra-axial lesion was observed, with contrast uptake, located at the base of the skull in the olfactory groove topography associated with intense peritumoral edema and absence of other lesions [Figures 1 and 2].

Through clinical findings, the surgical approach was adopted through bifrontal craniotomy and interhemispheric subfrontal approach for tumor resection. A spongy, extradural infiltrative lesion in the brain parenchyma and skull base was visualized. After gross total resection, the pathological examination showed MSS in the olfactory groove [Figure 3]. Immunohistochemical analysis is shown in Table 2, and 1st-day postoperative computed tomography scan is shows aggressive gross total resection [Figure 4]. In the outpatient follow-up, there was no evidence of deficits in patient's follow-up.

The patient returned to the hospital with similar symptoms to the first diagnosis (5 months after the procedure), evaluations showed that the lesion had relapsed and hence was reoperated on, in which the MSS was again gross totally resected, and the patient was further referred for oncologic adjuvant treatment with fractionated stereotactic radiotherapy [Figure 5]. Unfortunately, the patient had aggressive recidive after 3 months following the second procedure and palliative care was adopted, progressing to death 42 days after finding this new recurrence.

DISCUSSION

Soft-tissue tumors comprise a broad and diverse group of lesions with distinct clinical characteristics and presentations that differ from mesenchymal tissue, being neoplasms that may present in different age groups, as well as variable

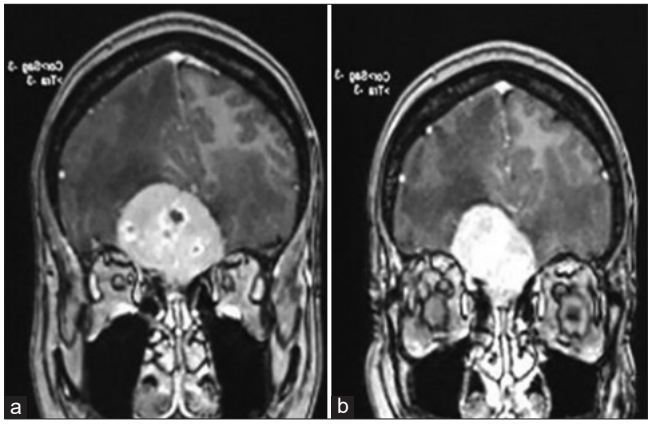

Figure 1: Coronal (a) and axial (b) views of T1-weighted magnetic resonance imaging with gadolinium showing a well-defined expansive lesion, with homogeneous enhancement in the olfactory groove topography.

Table 1: Literature review of primary intracranial synovial sarcoma case reports.

Articles	Age/sex	Symptoms	Location	Treatment	Characteristics	Histological analysis	IHC analysis	Subtype	Outcome
Kleinschmidt-DeMasters *et al.*(1998)[15]	19/ female	Fatigue, nausea, Bell's palsy, amenorrhea, headache, polydypsia, and hirsutism	A 3.5 cm tumor of the third ventricle. The sella showed slight demineralization	Stereotactic biopsy and placement of a ventriculoatrial shunt. Radiotherapy, and debulking of the tumor through the transcallosal approach	Not mentioned	Hypercellular, biphasic epithelial, sarcomatous spindle cell component was composed of fibroblast-like cells	Immunoreactive for vimentin and pancytokeratin focally for epithelial membrane antigen,	Biphasic	Spread of the tumor throughout the subarachnoid space. The patient died 2 months after the surgery
Scheithauer *et al.* (2006)[22]	48/male	Right-sided headache and blurred vision	A 3.5-cm irregular, contrast enhancing mass involving the sella and the right cavernous sinus, right optic nerve and chiasmal compression	Pterional craniotomy with subtotal resection of the mass and decompression the optic nerve	Not mentioned	Uniform cells disposed in sheets and pseudopapillary arrangements. Spindle- and epithelial-appearing tumor. Few mitoses and no necrosis	Cytoplasmic staining for keratin and membrane reactivity for EMA. Presence of the X; 18 translocation (fusion transcript SYT-SSX2)	Biphasic	Neurologically intact for 11 months, with no evidence of regrowth
Horbinski *et al.*[12]	81/male	Right-sided weakness, difficulty with ambulation, and speech, and agitation	A 5×4 cm durabased mass in the left parietal lobe with edema	Left parietal craniotomy with tumor resection except at the most inferior and anterior part of the lesion. Adjuvant radiotherapy	Soft and tan–gray with focal hemorrhage	Hypercellular and composed of plump spindle cells. Mostly arranged in large compact sheets, and fascicles. No epithelioid areas were seen	Positive for CD56, CD99, Bcl-2, p53, and vimentin	Not mentioned	No signs of tumor recurrence after 5 months of follow-up
Katsaros *et al.*[14]	15/ Female	Swelling and pain in the right upper neck	The mass extends from the right petrous bone to the occipital bone and the right atlantooccipital junction with compression of the spinal cord, medulla oblongata and right cerebellar hemisphere	Surgical macroscopically removal through a combined retrosigmoid, transmastoid, infratemporal approach with combined chemotherapy and radiotherapy	Not mentioned	Spindle-cell type	Not mentioned	Monophasic	Pulmonary and bone metastases after 1 year follow-up and 2 months later the patient died
Lin *et al.* (2012)[16]	21/male	Mild headache and general vertigo and weakened visual acuity in his right eye	A mass located at the right anterior cranial fossa associated with multiple cysts. There were infiltration of the right orbital apex and ethmoidal bulla	Surgical removal with adjuvant radiotherapy	The mass was extra-axial and attached to the dura mater. The cyst was found to contain xanthochromic fluid	Uniform oval and rounded cells having a scant cytoplasm with indistinct borders. No glandular elements or epithelioid cells observed in sections	Immunoreactivity for vimentin, CD99, Bcl-2, and S-100 protein. Presence of a t (X; 18) translocation in 50% of the tumor cells	Poorly differentiated	No signs of tumor recurrence after 6 months of follow-up

(Contd..,)

Table 1: *(Continued)*

Articles	Age/sex	Symptoms	Location	Treatment	Characteristics	Histological analysis	IHC analysis	Subtype	Outcome
Xiao *et al.* (2014)[35]	01/male	Crying, gait instability and vomiting	Lesion, measuring 6.0 cm 9 5.5 cm 9 5.0 cm, located in the right cerebellar hemisphere.	Complete surgical removal without further treatment	Not mentioned	Uniform spindle and oval cells with scant cytoplasm and indistinct borders. No glandular elements or epithelioid cells were observed in the sections	Immunoreactivity for vimentin, CD99, and Bcl-2, cytokeratin (AE1/AE3), CK7, and S-100 protein. Presence of a t (X; 18) translocation	Monophasic	No signs of tumor recurrence after 6 months of follow-up
Patel *et al.* (2016)[21]	21/male	Headaches, left homonymous hemianopsia, left hand weakness, and ataxia	Right parietal heterogeneous, hyperdense mass with a large medial hematoma with midline shift to 9 mm	Surgical decompression with posterior excision of the residual tumor. Stereotactic radiotherapy and adjuvant chemotherapy	Not mentioned	Not mentioned	Presence of t (X; 18)(p11.2;q11.2) chromosomal translocation	Not mentioned	Patient without neurological deficits nor tumor recurrence 2 years after the chemotherapy
Sharma *et al.* (2017)[23]	50/ female	Left-sided hemiplegia	A mass at the right parafalcine region with 5.5 cm×5.8 cm. With tissue edema and meningeal enhancement.	Not mentioned	Not mentioned	Spindle cells arranged in short and long interlacing fascicles. Epithelial-looking areas were not present in the sections studied	Immunoreactivity for CD99, Bcl2, TLE1, and vimentin. CK positivity. 90% tumor cells showed SS18 gene translocation	Monophasic	Not mentioned
Akdeniz *et al.* (2019)[2]	60/ female	Nausea, headache, and vomiting	Parieto-occipital mass with a cystic and solid component and peripheral vasogenic edema	Complete surgical removal and adjuvant chemotherapy	Not mentioned	Eosinophilic pleomorphic tumor cells with clear nucleoli and poorly differentiated cytoplasm	Positive for vimentin, S100, NSE, TLE1, BCL-2, EMA, FactorXIIIa, and CD34	Poorly differentiated	Planning for adjuvant radiotherapy
Wang (2021)[32]	35/male	Headache and slurred speech	Mass in the left frontotemporal parietal lobe, approximately 51.3 mm×54.2 mm×60.0 mm. Solid cystic mass with hemorrhage	Surgical removal with adjuvant radiotherapy	Lesion was gray–red, soft, with a rich blood supply. The M2 segment of the middle cerebral artery and its branches were located inside the tumor	Diffused growth with cellular atypia, and mitotic figures	Vimentin (+), Bcl-2(+), CD56 (+), SMA (−), GFAP (−), S-100 (−) and CD34 (−), TLE1 (+), as well as CD99 (−). (SS18) was negative for gene breaks	Not mentioned	16 months later: headaches and tumor recurrence

(Contd..,)

Table 1: *(Continued)*

Articles	Age/sex	Symptoms	Location	Treatment	Characteristics	Histological analysis	IHC analysis	Subtype	Outcome
Aggad *et al.* (2022)[1]	48/male	Severe headache	Frontal lobe	Complete surgical removal by a left frontal approach and adjuvant radiotherapy	Grayish red lesion, soft, well-vascularized mass, attaching to the dura	Dense, poorly differentiated and cells with an eosinophilic cytoplasm. Focal collagenic rearrangements	A transcript of fusion SS18-SSX2 was found	Monophasic	9 months later: lesion recurrence and seizures, with total resection. 6 months after second surgery: no recurrence nor symptoms
Manizhe *et al.* (2022)[17]	28/male	Headache and left hemiplegia	Intra axial mass lesion and vasogenic edema in the right frontal white matter	Complete surgical removal. Adjuvant radiotherapy	Central necrotic component and edema causing a midline shift to the left side	Irregular sheets of monotonous cells some with central necrosis set in the fibroblastic stroma	Positive reactivity for TLE1, FLI1, and INI1 and negative reactivity for Synaptophysin, Olig 2, SMA, CK and CD99	Biphasic	1 month later: recurrence and hemiplegia. Total resection was performed and any neurological signs
McCool *et al.* (2022)[18]	27/male	Seizures	Left ventricular trigone mass with surrounding oedema	Left occipito-temporal craniotomy, resection and adjuvant radiotherapy	Not mentioned	Fascicles of monotonous basophilic spindle cells. Vaguely lobular architecture	Positivity for EMA, bcl-2 and CD99. Presence of an SS18 gene rearrangement	Not mentioned	Recurrence of the tumor in the same site, total resection and adjuvant radiotherapy
Vora *et al.* (2022)[31]	31/male	Severe headache, right-sided weakness, and difficulty in speaking	Left frontal intra-axial heterogeneously tumor with an area of hemorrhage. Significant mass effect, and perilesional edema	Surgical removal with adjuvant radiotherapy	Solid cystic components and areas of hemorrhage with thrombosed veins in between	Spindle cell with large areas of necrosis. Tumor cells were arranged in fascicles and had scant and ill-defined cytoplasm. High mitotic activity	Staining TLE1 and focally for desmin. CD-99 positive. Positive for both translocations SYT-SSX1 and SYT-SSX2	Monophasic	Patient died 11 months after surgery
Zhang *et al.*[36]	7 female (15–47 yo) 9 male (5–65 yo)	12 (headache). Vomit, seizuere, ataxia, náusea, difficult to speech were the main symptoms	Frontal, temporal, occpital and parietal lobe, cerebellum, anterior and middle skull base, third and lateral ventricles, and sellar region	Surgical only (4) Surgical+radiotherapy (10), Surgical+radio-therapy +chemo-therapy (2)	Solid (11) Cystic (5)	Not mentioned	All cases SS18-SSX translocation were detected. Positive markers variable	Biphasic (9) Monophasic (7)	Overall survival between 6 and 24 months

IHC: Immunohistochemical, yo: Years-old, TLE1: Transducer-like enhancer of split 1, EMA: Epithelial membrane antigen, SMA: Smooth muscle actin, GFAP: Glial fibrillary acidic protein, NSE: Neuron-specific enolase. The numbers in parentheses indicate the number of cases that present the described condition. Therefore, among the 16 cases reported in the line, the values in parentheses indicate how many of them present the mentioned characteristics.

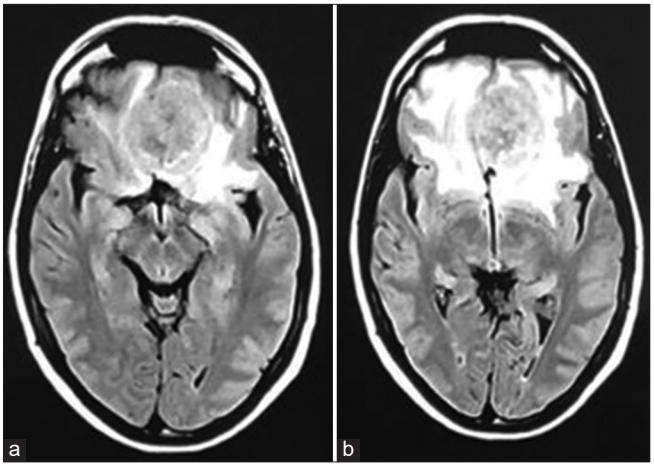

Figure 2: Axial view (a and b) of Fluid-attenuated inversion recovery (FLAIR) showing intense peritumoral brain edema.

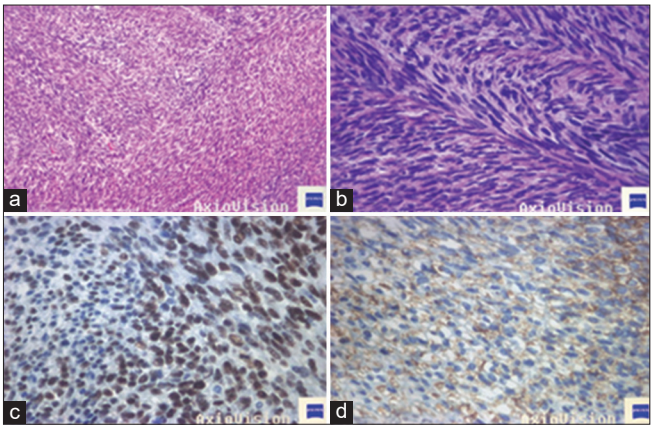

Figure 3: (a) 100XX - Monophasic synovial sarcoma with fibrosarcoma-like areas, spindle-shaped cells with moderate nuclear pleomorphism arranged in long and intercalated fascicles. (b) 400XX – Hyperchromatic, fusiform nuclei cells arranged in long bundles. (c) TLE-1 400XX – Strong and diffuse nuclear expression antibody in neoplasia. (d) CD99 400XX – Cytoplasmatic and diffuse membrane expression antibody.

topography, prognosis, treatments, and histopathological analysis.[25] Within this complex group of neoplasms, there are SSs, malignant tumors of partial epithelial differentiation and which can affect several anatomical locations. Presenting an occurrence in the age group close to 15–40 years, which may also affect the pediatric and old age group, with the smallest part of the cases described after 50 years.[5]

The clinical presentation of this condition varies according to topography, and the extremities, joints, or bursa are the most frequent sites of involvement, although the temporomandibular joint and knee are the most injured.[9] However, despite this rare description, the identification of primary intracranial SSs is a limited diagnosis described in the literature, and only 30 cases were found in 15 articles

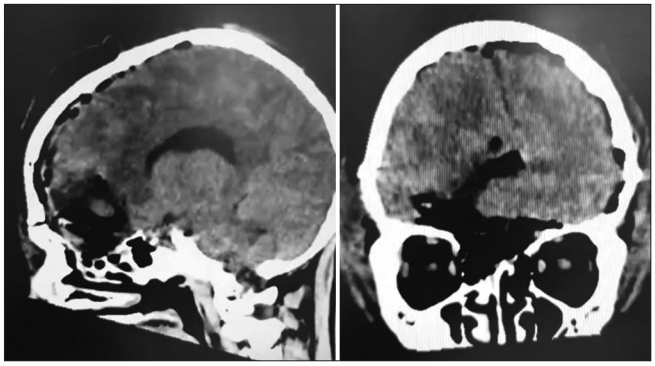

Figure 4: First-day postoperative computed tomography scan showing gross total resection of the tumor.

Table 2: Immunohistochemical panel.

ANTIBODIES	RESULTS
S 100 (POLYCLONAL)	NEGATIVE
FLAT MUSCLE ACTIN (1 TO 4)	NEGATIVE
DESMIN	NEGATIVE
MYOGENIN	NEGATIVE
EMA	NEGATIVE
CHROMOGRANIN A	NEGATIVE
CD99	POSITIVE
GFAP	NEGATIVE
TLE1	POSITIVE
BETA CATENIN (POLYCLONAL)	NEGATIVE
FL 1	NEGATIVE

TLE1: Transducer-like enhancer of split 1, Highlighted in Red: The results obtained in the IHC.

[Table 1] during the review carried out in this article. In the case described, in addition to the confirmation of a primary intracranial SS, it was a patient with NF1, a condition well described for committing the nervous system and predisposing to the formation of tumor lesions.[10] Nevertheless, the correlation between primary intracranial SS and NF1 is not yet well established, and there have been no similar cases reported in the literature to date, although translational studies have already tried to describe this relationship.

Clinical and surgical management

Among all 30 cases collected in this article, of which 14 are described in Table 2 and the others were gathered from the study Zhang *et al.*,[36] 21 (70%) patients had headache as main symptoms, some reporting progressive headache. Despite this, symptoms such as hemiplegia, seizures, and difficulty in speech and ambulation are also described. The tumor location is quite variable, with cases described in the frontal, parietal, occipital lobe, as well as the third ventricle. Although one of the cases described in Table 1 did not show the treatment

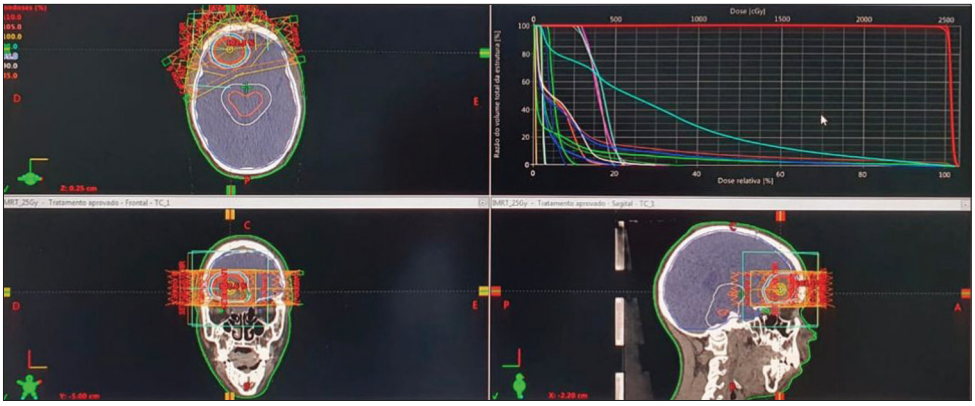

Figure 5: Fractionated stereotactic radiotherapy with intensity modulated radiation therapy and localization by image-guided radiotherapy. Total dose: 2.500 cGy in 5 fractions. Daily dose: 500 cGy.

performed, all the other 29 cases needed a surgical approach for lesion exeresis. In addition, 22 (73%) of the cases were indicated for adjuvant radiotherapy, and 5 (16.6%) required associated adjuvant chemotherapy. Among the 14 cases described in Table 1, seven presented recurrence or death. Among the 16 reported in the article Zhang *et al*.,[36] overall survival ranged from 6 to 24 months.

Laboratory diagnosis and analysis

In general, the macroscopic analysis of the lesion is evident that it is a heterogeneous, multinodular mass, well adhered to adjacent planes, infiltrative, and irregular. In addition, intracranial lesions present with a high aggressiveness and a rapid decline in neurological function, and dural adherence is described in most cases reported in the literature.[9,31] The histological analysis of SS comprise a wide spectrum of lesions that divide similarities in clinical evaluation, immunohistochemistry, and genetics. This analysis divides the classification of SSs into two groups: single-phase and biphasic sarcomas, although other subtypes of rarer lesions are also described, such as poorly differentiated sarcomas and myxoid types.[29] Single-phase sarcomas, spindle cell type, are the most common subtype of SS, characterized by the presence of only a spindle cell, presenting nuclei with homogenously dispersed chromatin, ovoids, and sparse amphophilic cytoplasm.[8] These characteristics make differential diagnosis with several other types of soft-tissue tumor, such as hemangiopericytoma and fibrosarcoma, for example. Meanwhile, on the contrary, the classification of the biphasic variant is characterized by the union of fibroblast-like spindle cells and epithelial cells and may vary in terms of concentration of these components, although they usually remain proportional.[8,28] Among the cases that mentioned the sarcoma subtype, described in Table 1 and by Zhang *et al*.,[36] 12 were described as single-phase and 12 as biphasic, in addition to two described as poorly differentiated.

Some markers are essential for identification and differentiation of lesion types. Cytokeratin CK19 And CK7, EMA, BCL-2, CD34, CD99, S-100, and transducer-like enhancer of split 1 are some useful examples for the identification and differentiation of SS from other lesions.[13,19,34] The analysis of genetic translocations is useful in identifying possible molecular abnormalities that predisposes to tumor formation.[6] In the case of SSs, we have a description of translocation t(X; 18) (p11.2; q11.2) in both single-phase and biphasic. It occurs so that there is a fusion of the SS18 genes (18q11) merges with the SSX genes (SSX1, SSX2) (present in Xp11) with the generation of the SS18-SSX fusion, characteristic by its tumorigenes.[6,26]

Correlation with NF1

NF1 is a disease of autosomal dominant genetic origin characterized by the mutation of the NF1 gene on chromosome 17q11.2. It was first described by Frederick von Recklinghausen and whose responsible gene was identified in 1990.[34] This is a variable clinical condition in which individuals with NF1 gene mutations may present different presentation of the disease. The best-known clinical presentations are coffee-with-milk stains and neurofibromas, benign tumors that may arise in the peripheral nervous system.[11,33] This condition is well described and clinically diagnosed, being an important factor in the generation of lesions involving the nervous system.[10] However, the correlation between the mutation of the 17q11.2 gene and the development of sarcomas is still a point to be discussed, being a source of current analyses.

Although the typical description of the NF1 gene mutation is linked to changes in the peripheral nervous system, there are descriptions of NF1 alterations related to the generation of lesions of the soft-tissue tumor group, such as pleomorphic liposarcomas myxofibrosarcomas, lesions of sarcomas subtypes.[4] In the study developed by Dodd *et al*.,[7] although it is a study in mice, induces the addition of the NF1 gene in

an attempt to simulate a patient with subtypes of sarcoma that present this mutation, revealing, from histological analyses, the development of soft-tissue sarcomas after this induction. However, these are still early analyses of these points, despite translational studies and already described cases of soft-tissue sarcomas and NF1 deletion, the correlation between primary intracranial SS and NF1 is not yet feasible, but there are indications of a possible correlation of these clinical conditions.

CONCLUSION

The present case report is an unusual intracranial presentation of primary MSS, associated with a patient with NF1 disease. It is a rare condition described in the literature, with complex diagnosis and essentially surgical treatment, with adjuvant radiotherapy alone or associated chemotherapy. The prognosis of these patients is poor, with reduced survival and several reports of recurrence. Despite the description of other cases that correlate the NF1 gene mutation and the presence of soft-tissue sarcomas, the description of cases correlating SSs and von Recklinghausen's disease is still incipient and further investigation should be performed to properly address its relationship.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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