

Case Report

Primary Central Nervous System Fibrosarcoma

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ABSTRACT

We report a rare case of a young female with primary brain fibrosarcoma, and to the best of our knowledge, we believe that only <50 cases have been reported or described worldwide so far. Fibrosarcoma is a malignant neoplasm, in which histologically the predominant cells are fibroblasts that divide excessively without cellular control and they can invade local tissues or metastasize. Primary central nervous system fibrosarcomas are very aggressive neoplasms and generally have a poor prognosis. This tumor is either from sarcomatous transformation of a meningioma or arises *de novo* within the brain parenchyma. Our patient, a 48-year-old woman, who presented with progressive speech disorder over the period of 4 months, showed a left temporoparietal lesion with surrounding edema and local mass effect. Total surgical resection was achieved. Histopathology revealed classical fibrosarcoma features and secondary screening revealed no other distant lesion as diagnosis of primary brain fibrosarcoma was established. This case is deemed to be extremely rare because most reports claim that recurrence is within 6 months with poor prognosis; however, this patient is currently recurrence-free at 3 years. This would suggest of the possibility for a relook into this disease's course and recurrence rate when complete excision is achieved. Due to extreme rarity of these tumors, more comparative studies will be needed to improve the disease outcome.

KEYWORDS: Central nervous system tumor, fibrosarcoma, malignant, neoplasm, sarcoma

INTRODUCTION

Primary brain fibrosarcomas are very rare, and to the best of our knowledge, we believe that only <50 cases have been reported or described worldwide so far. Sarcomas are a group of spindle cell tumors of mesenchymal cell origin and subdivided into smaller groups according to their predominant cell line. Fibrosarcoma is a malignant neoplasm, in which histologically the predominant cells are fibroblasts that divide excessively without cellular control and they can invade local tissues or metastasize.^[1] Primary central nervous system (CNS) fibrosarcomas are very aggressive neoplasms and generally have a poor prognosis.^[2,3] This tumor is either dura based or may arise within the brain, possibly from leptomeningeal infoldings.^[3]

CASE REPORT

We report an interesting case of a 48-year-old female patient who presented to our center with progressive

speech disorder over the period of 4 months. An initial computed tomography (CT) scan of the brain showed a vague left temporoparietal lesion with surrounding edema and local mass effect. Magnetic resonance imaging (MRI) – T1 and T2 weighted scans [Figures 1 and 2] – was carried out and better visualization of the lesion was obtained. Initial suspicion was of atypical or high-grade anaplastic meningioma due to dural based tumor with irregular borders; heterogeneous tumoral enhancement and peritumoral edema. There was no evidence of distant metastases or other possible primary origin of the tumor.

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Intraoperatively, adjacent skull bone erosion with attachment was noted as the tumor has breached the dura. The tumor was well demarcated and complete excision was done as per Simpson 1 grading. Structurally, the tumor was soft and friable with cystic component as well possible of necrotic material. The eroded bone segment was shaved off with high-speed drill device and placed back. At this stage, resemblance to a high-grade meningioma of WHO Grade 3 anaplastic type was more favorable.

Her histopathology examination result [Figures 3 and 4] reveals that she had primary fibrosarcoma of the brain and was referred to an oncologist who managed her conservatively as she refused radiotherapy and chemotherapy. The patient is now almost 4 years postoperative and she is under our regular follow-up now with imaging being done every 6 monthly to monitor for recurrence. She is recurrence free till now.

DISCUSSION

A rare tumor of the CNS makes it difficult to establish a diagnosis and treatment. Surgery with a complete or maximal excision of the margins should be aimed at resection. This helps the surgeon to determine the further management especially if the lesion comes back to be

as atypical, anaplastic or malignant, in which combined therapy is required. It is ideal to treat all lesions as high grade and aim for complete excision. Establishing a pathological diagnosis is also crucial as primary fibrosarcoma of the brain is extremely rare, and it is occasionally difficult to distinguish from other malignant neoplasms such as sarcomas and metastases,^[4] but it is mandatory to exclude common neoplasms.^[3]

A study by Okeda *et al.* concludes that on the basis of morphological and histochemical findings of their study, fibrous fibrosarcomas in the cranium are of meningotheial derivation and therefore should be included in the category of malignant meningioma.^[5] Gliosarcoma is unlikely if glial fibrillary acidic protein (GFAP) is negative, and if a tumor has dural origin, it is difficult to conclude if the sarcoma has arisen *de novo* or it represents a sarcomatous change in a meningioma. A classical herringbone pattern suggests a fibrosarcoma rather than a meningioma. Epithelial membrane antigen (EMA) immunoreactivity is seen in meningiomas but not in a fibrosarcoma.^[3]

Primary brain sarcomas are rarely curable with surgery and standard radiation therapy as they typically recur locally after treatment. A case report by Fraser *et al.*

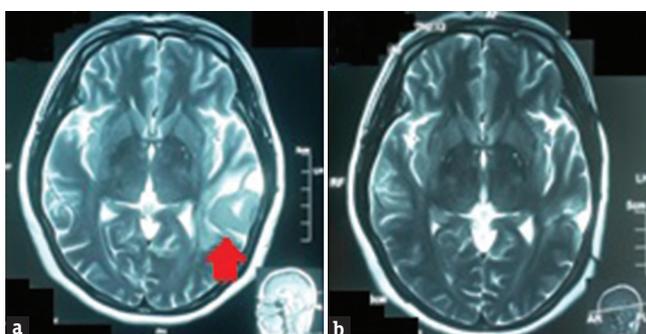


Figure 1: (a) Preoperative axial T2-weighted MRI image (red arrow shows the lesion) (b) Postoperative axial T2-weighted MRI image

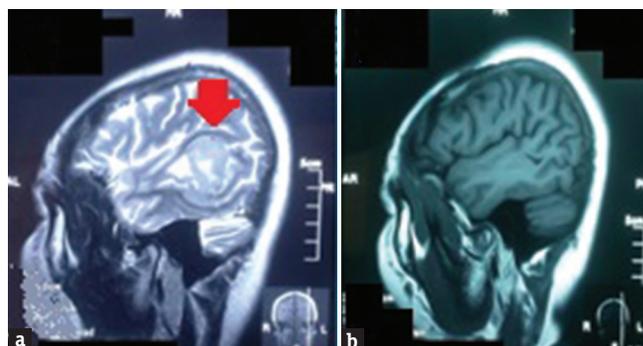


Figure 2: (a) Preoperative sagittal T2-weighted MRI image (red arrow shows the lesion) (b) Postoperative sagittal T1-weighted MRI image

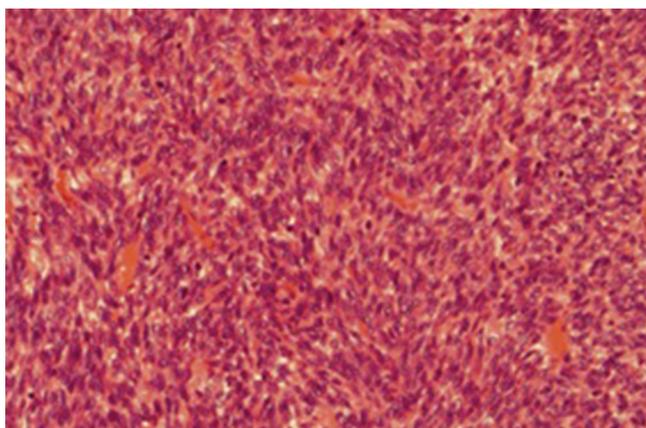


Figure 3: Herringbone pattern



Figure 4: Positive stain with vimentin

suggested that boost intracavitary brachytherapy after surgical excision and standard radiation therapy can play a critical role in preventing local recurrence and early death in patients with primary brain sarcomas. This case report also highlights the point that, despite being a highly recurrent aggressive tumor, the correct surgical approach and adequate tumor-free margin during resection can still ensure a recurrence-free interval despite no radiotherapy is given.^[6,7]

This rarity of this report is due to recurrence-free period of this highly malignant tumor which is now up to 2 years. Most reports show recurrence within 6 months. We emphasize the importance of treating every operable brain lesions as malignant and aiming for complete excision with minimal or no complication and aim for appropriate postoperative adjuvant chemotherapy and radiotherapy.

It is also reported that there is possibility of sarcomatous transformation of meningiomas in the pathogenesis of fibrosarcoma formation, but it usually does not present with classical herring bone pattern on histopathology.^[4,6,7] Moreover, it is usually derived from meningotheial derivative type; however, intraoperatively, the tumor was more representative of anaplastic meningioma which prompted the necessity for complete Simpson 1 excision of tumor, but the final histopathology proves toward a primary fibrosarcoma of a *de novo* origin.^[4,7]

CONCLUSION

In our case, a complete resection with tumour-free borders was achieved and this proved adequate as patient is well without any recurrence 3 years after surgery despite refusing radiation therapy. This also goes against the reported survival and recurrence rate of primary brain fibrosarcoma which would suggest of the possibility for a relook into this disease's course and recurrence rate when complete excision is achieved. Dealing with rare brain tumors requires the neurosurgeon to be on the right track from the time the patient is seen and diagnosed. Fibrosarcomas are malignant tumors; hence, precise and quick diagnosis with complete or maximal surgical excision which is followed by accurate histopathological interpretation and postoperative combined therapy should give patients a longer disease-free interval.

Due to extreme rarity of these tumors, we are unable to report case series which would reflect better on the overall survival and mortality among these patients.

Radiological findings [Figures 1 and 2]

- Figure 1:
 - Preoperative axial T2-weighted MRI image (red arrow shows the lesion)
 - Postoperative axial T2-weighted MRI image
- Figure 2:
 - Preoperative sagittal T2-weighted MRI image (red arrow shows the lesion)
 - Postoperative sagittal T1-weighted MRI image.

Histopathological findings [Figures 3 and 4]

Microscopic examinations revealed a cellular tumor composed of spindle cells arranged in intersecting fascicles with herringbone pattern as well as storiform arrangement. Nuclei show minimal pleomorphism with ill-defined cytoplasmic borders. There is no necrosis seen and mitotic activity is not increased [Figure 3]. Immunohistochemical staining is positive for vimentin [Figure 4] and negative for other stains such as EMA, cytokeratin MNF 116, cytokeratin 7, CD34 antibody, S100 protein, desmin, smooth muscle actin, and GFAP. Proliferative index (Ki-67) is also not raised. Thus, the interpretation by a neuropathologist is low-grade fibrosarcoma.

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Conflicts of interest

There are no conflicts of interest.

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