

Frontal Base Gliosarcoma Mimicking as Olfactory Groove Meningioma- A Rare Case Report

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Abstract:

Gliosarcoma is a rare tumour of the brain. It is a type of the glioblastoma. This tumour has high complication rate as well as mortality. In our Institution, a 30 year old female was admitted with the complaints of headache and vomiting and weakness of rt. side of the body. She had history of radiotherapy on the frontal bone at four years age and following enucleation of the left eye. Now her recent MRI showed features of a tumor compatible with olfactory groove meningioma. She underwent craniotomy and her tumour was removed. Her histopathological exam was compatible with gliosarcoma which was confirmed with immunohistochemistry. And she was referred to oncologist for further management.

Gliosarcoma is a rare intracranial tumour which has variable presentation. The aim of this case report is to present a secondary gliosarcoma which had presented as olfactory groove meningioma.

Key Words: Meningioma, gliosarcoma, skull neoplasm, Secondary neoplasm. Irradiation, Frontal base.

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Introduction:

Gliosarcoma is defined as a biphasic neoplasm of glial origin with mixed areas of malignant mesenchymal differentiation¹. The most common presentation of gliosarcomas is as secondary tumors, frequently arising after cerebral radiotherapy to treat a primary glial neoplasm, with very few cases reported of primary origin¹. Gliosarcoma (GS) is a very rare primary mixed tumor in the central nervous system, with a biphasic pattern consisting of glial

(anaplastic astrocytes) and malignant mesenchymal elements².

GS was described for the first time in 1895 by Stroebe and defined as a subtype of glioblastoma by Feigin and Gross in 1955 and Rubinstein in 1956². There are certain features which point toward a distinct clinicopathological behavior of GS namely peripheral location on cerebral lobes, tendency for dural attachment, resemblance to meningiomas,

The study of Singh et al included 16 consecutive patients (mean age: 45.2 years [range 17–70 years], M: F = 7:1) of histopathologically proven GS affecting brain, operated at their institute over 5 year period³. According to Kozak, the incidence of GS is between 1% and 8% of all malignant gliomas and thus represents an exceptionally rare neoplasm.⁴

Gliosarcoma (GS) is a rare variant of glioblastoma (GBM), characterized by a biphasic tissue pattern, with alternating areas of glial and mesenchymal differentiation⁵. GS corresponds to less than 2% of all glioblastomas. It is characterized by displaying both glial and sarcomatous components⁶.

GS are observed as a firm, often times well circumscribed, superficial lesion, with meningeal adhesions. GS invading the skull base with

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accompanying extracranial extension has not been previously documented in primary GS⁵.

Schuss in 2010 had commented that common causes of the extradural extension of gliomas are surgical intervention and radiation-induced damage of the dura mater⁷. He referred to Kawano et al., who considered three possibilities of extracranial extension of gliomas:

1) along the perivascular or dural slit 2) through the cranial or spinal nerves; and 3) direct destruction. Bone invasion might also interfere with local dural blood supply, thus resulting in dural necrosis⁷.

There are multiple neoplastic and non-neoplastic entities that clinically and radiographically mimic meningiomas, these include solitary fibrous tumors, hemangiopericytoma, gliosarcoma, leiomyosarcoma, dural metastases, Hodgkin's disease, plasmocytoma, Rosai-Dorfman disease, neurosarcooidosis, melanocytic neoplasms and plasma cell granuloma⁸.

Radiological features of gliosarcomas are well-demarcated irregular masses (on both CT and MRIs), with a smooth external wall demarcated from the surrounding brain parenchyma regardless of peripheral edema¹.

Case Report:

A 40 years old non diabetic, nonhypertensive right handed female, was admitted to our department with the complaints of progressive headache for one year, and weakness of the right side of the body. She was completely well one year back. She started having headache, which was usually dull in nature and nonspecific. She occasionally had vomiting. Then her headache started to worsen. It had no relation to work, heat or photophobia. In the mean time she noticed that she had weakness of the right side of the body. Her upper and lower limbs were affected but she could continue walking without support. She had no history of seizure. She had History of enucleation of the left eye at the age of 4 years followed by radiotherapy in the frontal region.

On examination, her higher psychic function was normal with no deficit of speech. Her cranial nerves were intact but with loss of olfaction of the left nostril. She had no motor or sensory deficit except for a weakness of the right side. Her muscle power was 4/5 in all groups of muscles of both upper and lower limbs. She had no cerebellar signs or signs of meningeal irritation.

Her MRI of Brain with contrast had shown a contrast enhancing lesion attached with the frontal base at

the olfactory groove and the falx on the left anterior cranial fossa. There was cleavage between the brain and the tumour on T2 weighted image. Her CT scan showed a rounded mass without calcification which enhanced with contrast. (Fig: 1)

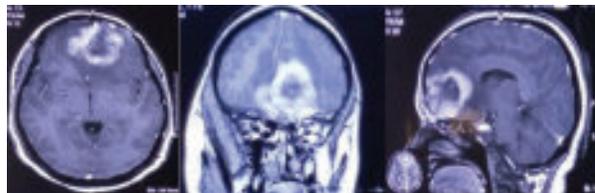


Fig:1- Pre operative Axial, coronal and sagittal contrast MRI Images

She had undergone a left frontal craniotomy. The periosteum was tightly adherent over the area of the tumour and also the bone was thickened. The Left frontal bone was removed and dura was incised. The left frontal lobe was gently retracted and the tumour was seen. It was grayish in colour, firm, not suckable, with a cleavage from the brain, moderately hemorrhagic. It was attached to the base of the frontal bone more along the olfactory groove and also with the falx. At places, it was very hard and had to be cut with the knife. It was totally removed. Involved dura was excised and duroplasty was done with G-patch. The bone was replaced. A subgaleal drain was placed and wound was closed in layers.

Her postoperative period was uneventful.

Her histopathological report stated that it was a gliosarcoma (Fig 2). Immunohistochemistry: it was positive for GFAP and vimentin. Therefore, the diagnosis was gliosarcoma.

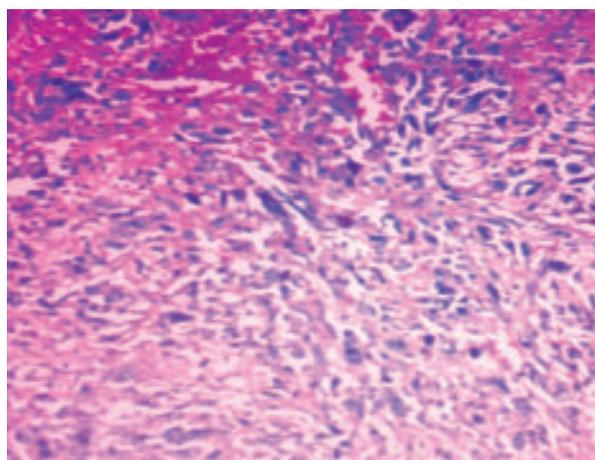


Fig.-2 (a): Histopathological exam: Showing sarcomatous and gliomatous components

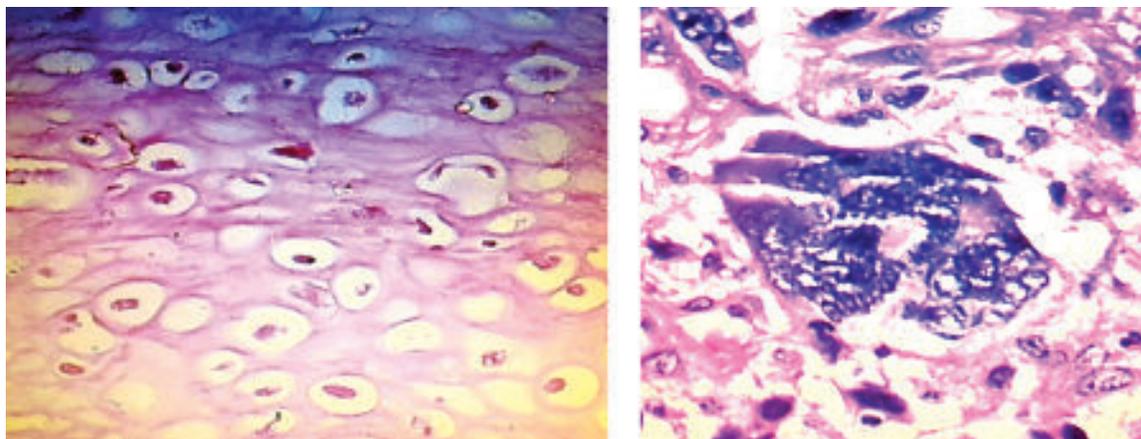


Fig.-2 (b): Histopathological photograph showing cartilaginous component and some gliotic cells

She was discharged on the 8th POD with advice for consultation with oncologist. But after one month she came with the complaints of elevation of the bone flap on the left side. An urgent CT scan was done and it showed a huge tumour of the right side involving the falx and extending from the olfactory groove up to the planum sphenoidale. This time she urgently undergone craniotomy of the right side. And the total tumour, involved dura, falx and adjacent gliotic tissue was excised. Duroplasty was done. Wound was closed in layers. In the third post operative day she had CSF collection under the skin and successfully

treated with lumbar drainage. Her lumbar drain was removed on the ninth post operative day. Her stitches were also removed on the 10th POD and then she was discharged with advice to consult with oncologists.

Discussion:

Malignant gliomas account for 35–45% of all adult brain tumors, and approximately 85% are glioblastomas. So, glioblastomas account for 29.7–38.2% of all adult brain tumors. Gliosarcoma constitutes approximately 2% of all glioblastoma, and accounts for 0.59–0.76% of all adult brain tumor². It typically affects older men, with onset between the fourth and sixth decades of life and a male/female ratio of 1.8/1, although some cases of infantile gliosarcoma have also been describe². Our patient was a female in her 30s.

It is normally located in the supratentorial region with a slight preference for the temporal lobes, although it can also affect the frontal, parietal and occipital lobes². Our patient presented with a frontal lobe tumour in the olfactory groove.

In Sing's series, patients in group A (GS with meningioma like appearance) had radiologically well circumscribed masses that showed more or less homogenous albeit strong contrast enhancement (with or without dural tail) and were firm, well demarcated from surrounding brain at surgery³. It was also in our patient.

The criteria used for diagnosis of radiation induced tumors in general include (1) the tumor appear in the area covered by irradiation, with a significant time interval (years) between irradiation and the appearance of the new tumor, (2) the tumor must be

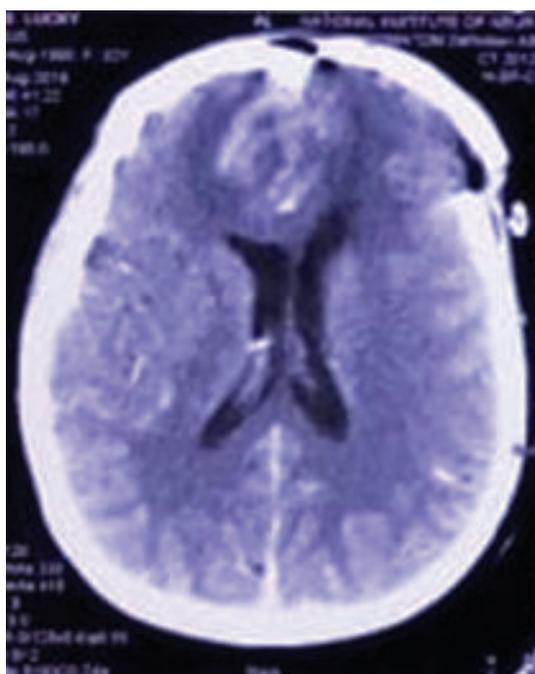


Fig.-3: Post operative CT scan Showing recurrence of tumour (1 month following 1st surgery)

absent prior to irradiation, and (3) the new tumor has to be histologically different than the first tumor³. Our patient had a history of previous irradiation at the age of 4 years for a lesion in the left eye.

Damodaran have noted that 100% of secondary GBMs tended to be peripherally placed as compared to 67% of primary GS³. In our patient, it is also peripherally placed. Radner has also commented that in rare cases, glioblastoma and, more often, gliosarcoma, may infiltrate dura and bone⁹.

According to Sade, intraoperatively, gliosarcomas (like meningiomas) can be firm, well-circumscribed, and adherent to the dura, or they (like astrocytomas) can exhibit irregular boundaries with the surrounding cerebral tissue, which can lead to limited resection¹⁰. In the present case, intraoperatively observed features were more similar to those of aggressive meningioma.

Charfi had commented that sarcomatous proliferation is more heterogenous; it commonly displays fibrosarcoma or pleomorphic sarcoma (previously called malignant fibrous histiocytoma) or leiomyosarcomatous patterns. Occasionally, other lines of differentiation have been described, such as the formation of cartilage, osteoid, vascular, smooth and striated muscle, and lipomatous tissue¹¹. In the present case there were portions of hyaline cartilage in histopathological section. (Fig: 2)

Conclusion:

Gliosarcoma is a rare tumour of the brain, which is a subtype of glioblastoma. Imaging of the tumour may not be definitive as this tumour may have similar features of meningioma. In our patient the tumour had the features of olfactory groove meningioma. But histopathological report as well as immunohistochemistry confirmed it as gliosarcoma. Therefore, we have to keep gliosarcoma in mind in

the differential diagnosis of meningioma especially in patients who had previous irradiation.

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