University Journal of Pre and Para Clinical Sciences

ISSN 2455–2879 **2021, Vol.7(1)**

A RARE CASE REPORT ON GLIOSARCOMA

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Abstract : Gliosarcoma is a rare malignant glial tumour considered as a variant of Glioblastoma. The clinical presentation, age distribution and treatment modalities are similar to that of glioblastoma. The differentiating features rest on the tumour location, different radiographic appearance, infrequency of EGFR mutation, tendency for extracranial metastasis and poor survival rate.

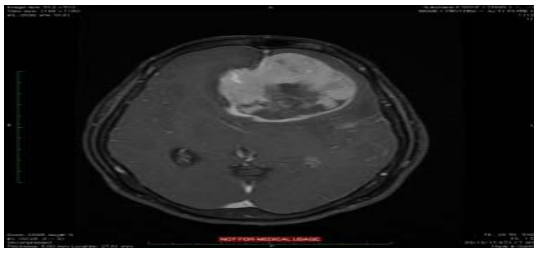
Keyword : Gliosarcoma, Glioblastoma, Reticulin, GFAP

INTRODUCTION:

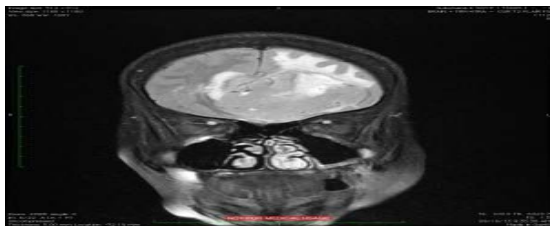
Gliosarcoma was first described by Heinrich Stroebe in 1895 as a brain tumour demonstrating both glial and mesenchymal components. Later it gained importance only by the detailed analyses of Feigin and his colleagues in 1955 and is named as Feigin's tumour. Under 2007 World Health Organisation Classification, Primary Gliosarcoma is placed as a grade IV neoplasm and a variant of Glioblastoma. Primary Gliosarcoma is defined as a well circumscribed tumour with biphasic glial and metaplastic mesenchymal components.

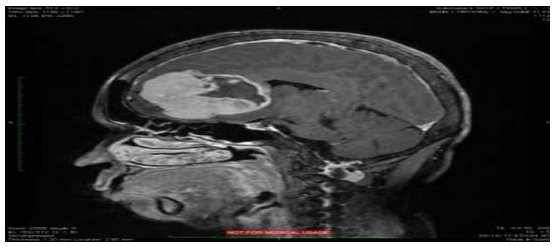
CASE REPORT:

56 years old female presented with weakness, confusion and disorientation. MRI imaging studies revealed an intracranial mass in the anterior cranial fossa with mass effect and edema in frontal lobes. Tumour showed focal areas of enhancement. The patient underwent gross total resection of the tumour. The tumour was received in piecemeal and measured about 6 x 5 x 2 cm in size. Cut surface was grey white, firm with focal areas of necrosis. MRI Imaging Studies of brain showing a Frontal mass lesion

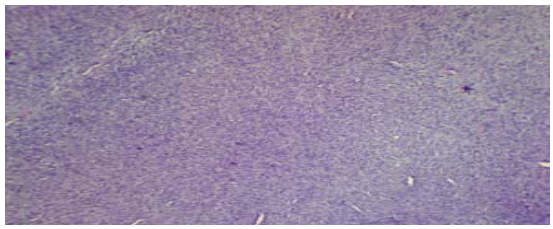


MRI Brain

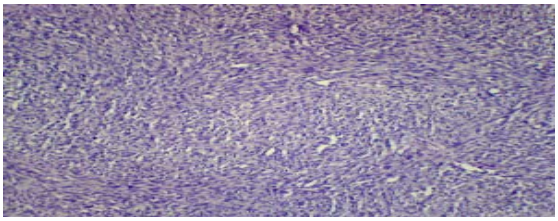




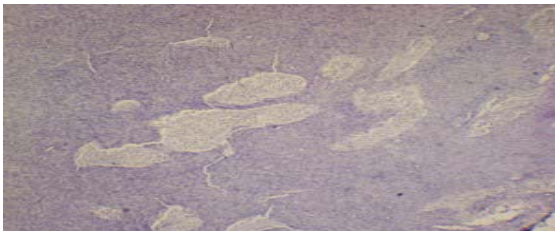
Histologic sections showed a markedly cellular spindle cell neoplasm arranged in fascicles. The tumour cells were pleomorphic and mitotically active. Intermingled among these cells were islands and nodules of glial cells with somewhat larger nuclei. Areas of necrosis and vascular proliferation were noted. Immunohistochemistry showed positive immunostaining for GFAP in gliomatous areas which was absent in spindle cell areas. Reticulin staining show increased deposition of intercellular reticulin in spindle cell foci which was not seen in glial nodules.



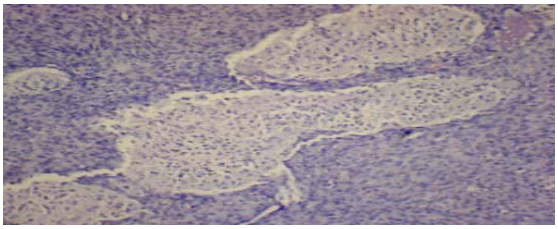
Gliosarcoma- Cellular spindle cell component in fascicles (H&E,10X)



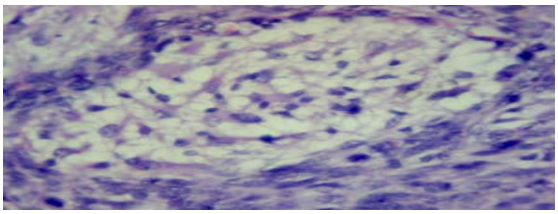
Pleomorphism and increased mitosis in spindle cell component(H&E,40X)



Islands and nodules of glial cells interspersed with spindle cell component (H & E, 4X)

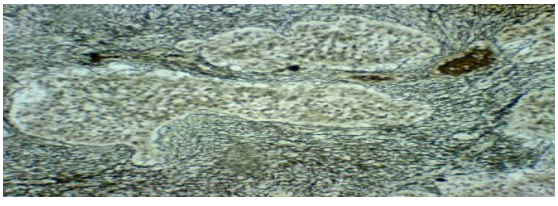


Glial component (H & E, 10X)

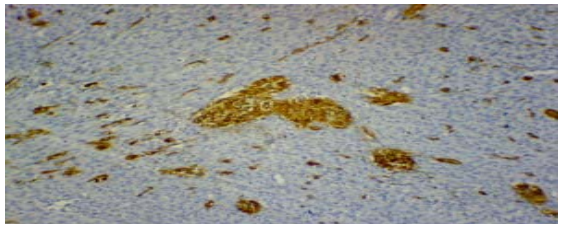


Glial cells with larger nuclei (H & E, 40X)

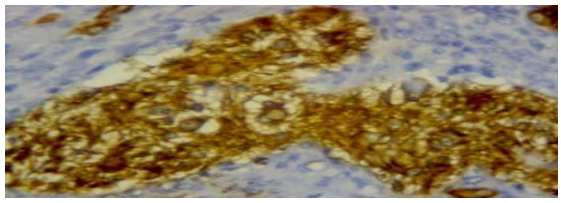
Reticulin rich areas of spindle cells with separate reticulin free islands of glial cells (Low power view)



Reticulin positivity (High power view)



Islands and nodules of GFAP glial cells (Low power)



GFAP Cytoplasmic Positivity- High power

DISCUSSION:

Gliosarcoma is a rare glial neoplasm accounting for 1.8 – 2.8% of the glial tumours and 2 – 8% of Glioblastoma. Tumour commonly presents in the 6th to 7th decade with slight male preponderance (M:F 1.4-1.8 :1). The common clinical features are that of a rapidly growing intracranial mass lesion with aphasia, headache, seizures, hemiparesis and cognitive decline, which clinically mimics Glioblastoma. The difference is that Gliosarcoma never occurs infratentorially and have predilection for temporal lobe with few reports showing frontal lobe predominance. The pathogenesis of gliosarcoma is unknown. Few theories are proposed. Initially, it was thought that the sarcomatous component arose from neoplastic transformation of hyperplastic blood vessels in a preexisting malignant glioma. This concept was supported by positive reactivity of sarcomatous component to vascular endothelial markers like Factor VIII, vWF and CD34. Another theory suggested monoclonal origin of both components with sarcomatous component arising through aberrant mesenchymal differentiation. Both components shared common genetic alterations like p53 and PTEN mutations, p16 deletion and CDK4 amplification. Less common are gains on chromosome 7, 20q, 9q and X and losses on chromosome 9p, 10 and 13q. EGFR amplication is seen in about 50% of glioblastoma cases, whereas it is much lower about only 8% in gliosarcoma. The currently proposed hypothesis is that GBM promotes differentiation of local or circulating mesenchymal stem cells into sarcoma. Gliosarcoma exhibits typical biphasic pattern showing gliomatous and mesenchymal differentiation. The glial component is similar to that of glioblastoma. The mesenchymal portion consists of fibrosarcoma like areas or malignant spindle cells with nuclear pleomorphism and mitotic activity. Differentiation towards cartilage, bone or muscle may be seen. The sarcomatous component should not be of minimal distribution and it should be well demarcated from the glial portion. Immunohistochemistry shows reactivity for factor VIII-related antigen or *Ulex europaeus I*agglutinin (UEA-I) suggesting vascular origin. GFAP shows positivity only in gliomatous area, whereas the spindle cell foci demonstrate reticulin reactivity in these GFAP negative areas. Usually, cerebral gliomas including glioblastoma rarely metastasize. But extracranial metastasis is well demonstrated in gliosarcoma which show mostly sarcomatous component in these sites. Most common sites are lung and liver, but also seen in cervical lymphnode, spleen, kidney, adrenals, skin and bone marrow. The treatment comprises of surgical resection, followed by

radiotherapy and chemotherapy. Compared to glioblastoma, the prognosis is poor with a median survival of 4 months in untreated patients and 6 to 11 months in treated individuals.

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