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HISTIOCYT IC SARCOMA OF THE FALX CEREBRI WITH DISTANT METASTASIS

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ABSTRACT:

Histiocytic Sarcoma is an extremely rare CNS tumor and to the best of authors' knowledge, the extra-axial CNS histiocytic Sarcoma involving FalxCerebriin the humans has not been described earlier in the literature.We report an unusual case of22 year old male who presented to the Civil Hospital with symptoms of chronic headache and bilateral lower limb weakness. His MRI Brain showed an aggressive heterogeneously enhancing extra-axialfalxtumor with trans-calvarial invasion seen on interim CT scan and demonstration of distant metastases after resection and craniectomy. The immune-histochemical analysis was strongly positive CD68 and S-100 signifying Histiocytic Sarcoma.

KEY WORDS:

Histiocytic Sarcoma, CNS, Falxcerbri, craniectomy, trans-calvarial invasion.

INTRODUCTION:

Histiocytic sarcoma is a rare, lympho-hematopoietic malignant neoplasm composed of mature tissue histiocytes. It can occur at any age, from infancy to adulthood.The most frequently involved organs are the skin, lymph nodes, and intestinal tract. Involvement of the central nervous system (CNS) as either a part of a systemic disease or as a primary lesion has rarely been described so far1. We present the imaging features and manifestations of Biopsy proven Histiocytic Sarcoma of the Falxcerebri in this case.

CASE REPORT:

A 22 year old male presented to the Civil Hospital in June 2016 with symptoms of chronic headache and bilateral lower limb weakness for last 1 month.There were no known co-morbids. There was hypertonia with positive babinski sign and power was 3/5 on average in all muscle compartmentsof lower limbs.

Magnetic resonance imaging of the Brain was performed on GE Health Care SignaHDxt 1.5 Tesla Scanner which revealed11x7cmaggressive heterogeneously enhancing extra-axial anterior falx mass with extensive peri-lesionalvasogenic edema (Fig.1). The initial differentials aroused in this patient were aggressive falx meningioma / haemangiopericytoma.Before planning for the management, the patient was lost on follow up and presented again in September 2016 with worsening of the symptoms and new onset of seizures.The patient then underwent Contrast CT Brain(Fig.2) performed with 16 slice ToshibaSpiral CT Scanner which re-demonstrated the lesion with significant size increment and duralsino-venous thrombosis involving the superior sagittal sinus, trans-calvarial and scalp invasion.

The surgical resection of the tumor and craniectomy was done.Thehisto-pathological specimen examined at AKUH showed multiple irregular tan white irregular soft tissue fragments. Its microscopic sections revealed neoplastic cells arranged in sheets and nodules. The neoplastic cells are large, show abundant eosinophilia cytoplasm, pleomorphic, vesicular nuclei with variably prominent nucleoli. Multinucleated tumor cells also noted. Intervening areas show moderate to dense inflammation comprising of lymphocytes, plasma cells, neutrophils and eosinophil. Immuno-histochemical stains were performed which show that LCA is Positive & CD68 and S-100 are diffuse strong positive in the neoplastic cells. The biopsy features were consistent with Histiocytic Sarcoma.He later on developed severe malaise, cachexia, altered level of consciousness.

The interim CT scan performed in February 2017showed no recurrence with post-operative subdural collectionmeasuring 1.7cm in depth(Fig.3),extending into falx under the 6.0x5.5cm post-surgical parietal defect; the liverwas enlarged in size measuring 19.2cm with multiple hypodense lesions seen in both lobes of the liver, largest measuring 1.5x1.0cm in segment VIII of liver; in keeping with distant hepatic metastases (Fig.4). The patient was referred to Oncology for induction of chemotherapy.

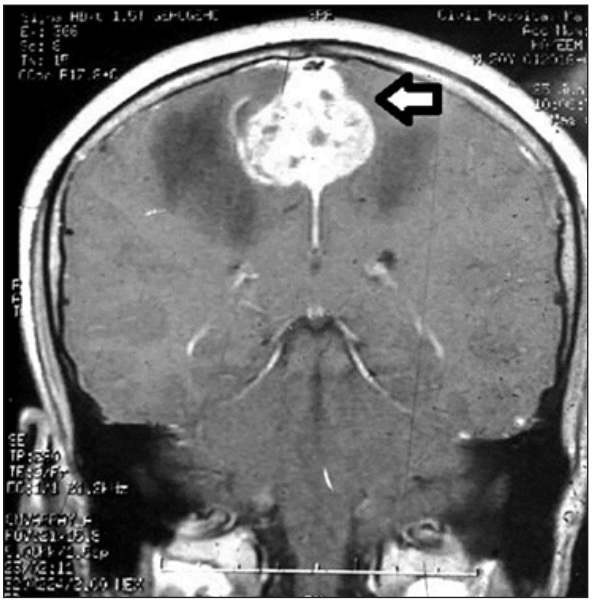

Figure 1:
Coronal MRI Brain Contrast showingFalx lesion with peri-lesional edema(arrow)

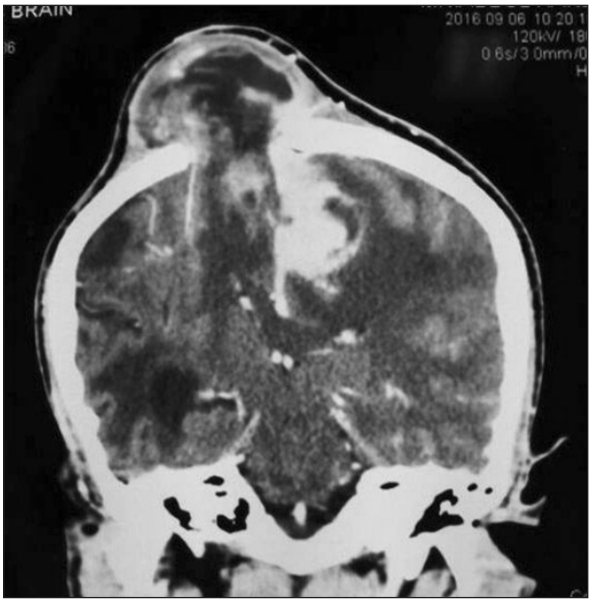

Figure 2:
Coronal CT Brain showing trans-calvarial invasion of the Dural tumor.

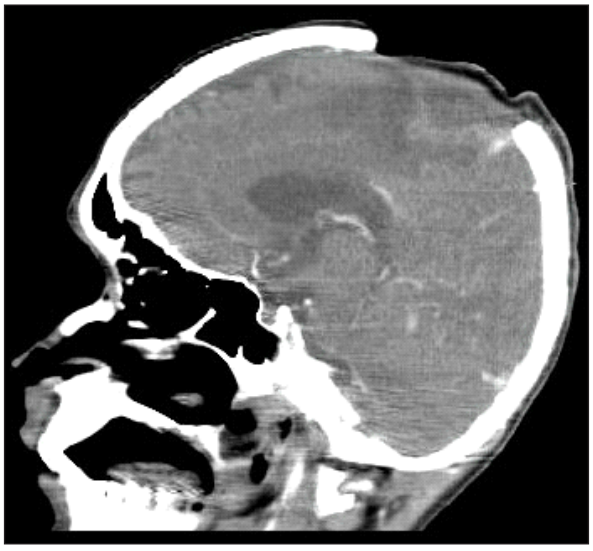

Figure 3:
Sagittal CT Brain showing subdural collection

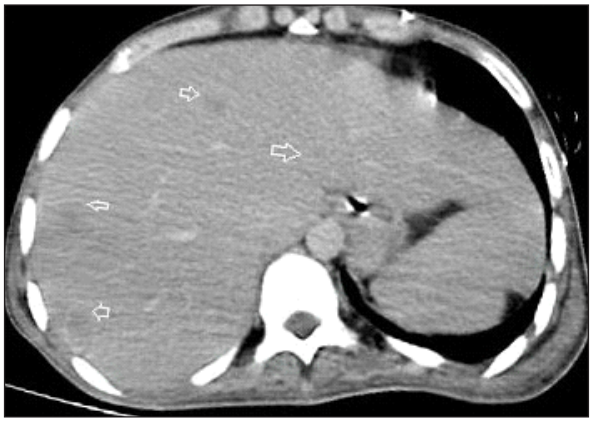

Figure 4:
Axial Contrast CT Abdomen showing liver metastases (white arrows)

DISCUSSION:

Histiocytic sarcoma (HS) is a rare neoplasm composed of tumor cells derived from the monocyte/macrophage lineage. HS has been reported in association with a number of hematological malignancies, including Acute Lymphoblastic Leukemia (ALL) 2. Chalasani S. et al reported a case of 44 years old male, presented with unsteadiness, loss of balance, and left-sided weakness of 2 weeks2. He underwent CT and MRI scan of the brain that showed two intra-axial enhancing lesions: one in the corpus callosum of 3.5 cm, and the other adjacent to the right lateral ventricle of 2.6 cm, both associated with vasogenic edema. His previous history revealed that he had been diagnosed with ALL with cerebrospinal fluid (CSF) involvement 16 years prior.

Diagnosis of HS requires the presence of histiocytic markers and the systematic exclusion of markers of other cell lineages. Primary HS central nervous system tumors are aggressive and generally have poor outcomes.

So H and his colleagues3 in 2015 described a patient with primary CNS histocytic sarcoma involving the cerebral hemisphere and spinal cord, who had been initially misdiagnosed as demyelinating disease. Two biopsies were necessary before a correct diagnosis was made. A histologic examination showed bizarre shaped histiocytes with larger nuclei and nuclear atypia. The cells were positive for CD68, CD163, and S-100 protein.

In 2013, Pérez-Ruiz E et al4 presented case of 41-year-old Caucasian woman with no previous history of disease who started with systemic symptoms such as headache and chills. Magnetic resonance imaging with gadolinium contrast of the brain suggested a 1.5×2cm meningeal mass in the temporal lobe with a non-uniform vasogenic edema. The histological findings revealed a histiocytic sarcoma- comparable to our case, however our case showed the site of tumor at falx.

Devic P et al5reported a case of 43-year-old patient in the International Journal of Medicinewho had complains of progressive ataxia, headache and altered general status lasting for 3 weeks. Brain magnetic resonance imaging (MRI) showed T2hyperintense multifocal lesions involvingcorpus callosum, right middle cerebellar peduncle, right periventricular white matter, cervical and dorsal spinal cord with homogeneous gadolinium enhancement in T1 post contrast sequence.

CNS histiocytic sarcoma is an extremely rare and severe disease and should be suspected when facing rapidly evolving neurological manifestations6. There is no consensus on the most appropriate treatment because of the lack of data and limited number of reported cases.

CONCLUSION:

Involvement of the central nervous system in HS is rare and is usually aggressive. Its rarity can easily lead to a misdiagnosis.Brain biopsy with immunophenotypic study is the only way to confirm the diagnosis and should not be delayed. To the best of authors' knowledge, the extra-axial CNS histiocytic Sarcoma involving FalxCerebri has not been described earlier in the literature.

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Ateeque Ahmed Khan; data collection, data analysis, manuscript writing, manuscript review
Mahnoor Hafeez; data collection, data analysis, manuscript writing, manuscript review