PLEOMORPHIC XANTHOASTROCYTOMA WITH ANAPLASTIC FEATURES- A RARE CASE REPORT AND REVIEW OF LITERATURE

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PRESENTATION OF CASE

A 42-year-old healthy woman complained of headache, imbalance while walking with slurring of speech with weakness in right upper and lower limbs with deviation of mouth evolving over 15 days. CT brain plain and contrast study revealed a heterogeneous density mass lesion in left parietal lobe with perilesional oedema with a midline shift of 8-10 mm towards the right side. Patient was given symptomatic treatment for three days and there was relief of focal neurological symptoms.

DIFFERENTIAL DIAGNOSIS

- Glioblastoma Multiforme
- Atypical Meningioma
- Pleomorphic Xanthoastrocytoma
- Gliofibroma
- Gliosarcoma

CLINICAL DIAGNOSIS

Glioma

PATHOLOGICAL DISCUSSION

Histological examination showed that lesion comprised of predominantly malignant spindle cell component (85 - 90%) and focal pleomorphic areas with bizarre nuclei and abundant eosinophilic cytoplasm. Eosinophilic granular bodies were seen. Mitosis in spindle cell area with necrosis with strong staining for p53 and increased Ki-67 suggestive of high grade and aggressive tumour and possibility of gliosarcomatous transformation was seen. Staining for GFAP was positive in pleomorphic area and negative in spindle cell, Vimentin and p53 were positive. CK (Epithelial Marker) was positive in pleomorphic area. Ki-67 was positive in 30 - 35% cells and negative for p40, SOX-10 and Desmin. Lesion was turned out to be pleomorphic xanthoastrocytoma with anaplastic features with gliosarcomatous transformation.

Pleomorphic xanthoastrocytoma (PXA) is an uncommon tumour, constitutes less than 1% of all astrocytic glial neoplasms, was first reported in 1979.[1,2,3] PXA commonly occurs in young patients and manifests itself first as seizures followed by focal neurological deficits.

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The role of radiotherapy or chemotherapy has not yet been established, because of the relative infrequency of this disease.[2,4] PXA is classified as grade II tumour in the WHO classification of tumours of the CNS.[3,5] In literature 9 to 20% PXA may undergo malignant change at recurrence or may display at the time of initial presentation. Malignant transformation is mainly associated with high mitotic activity and necrosis. The criteria for PXA with anaplastic features are five or more mitotic activity per 10 high-power fields, necrosis, microvascular proliferation, marked cellular anaplasia and high Ki-67 labelling indices.[2] PXAs with anaplastic features management is highly controversial, as very sparse literature is available.[2]

It usually arises within the first three decades of life with no gender predilection. These tumours are ordinarily located in cerebral hemispheres without dural involvement. PXA is one of the desmoplastic glial tumours of the brain. Microscopically, its most conspicuous properties are pleomorphic and in part xanthomatous tumour cells that contain intracytoplasmic lipid. Monstrous giant cells, cell atypia, nuclear irregularity, hyperchromatism and mitotic activity and necrosis can also be seen.[6] Development of glioblastoma has been reported in some patients with PXA.[2,4] Anaplastic PXA shares several clinical and histologic features with other desmoplastic neuroepithelial neoplasms such as gliofibroma, gliosarcoma, desmoplastic infantile ganglioglioma and desmoplastic cerebral astrocytoma of infancy.[6] Various molecular genetic changes are associated with PXA,[2,7,8] which were entirely different from the other diffusely infiltrating astrocytomas. The CDK4, CDKN2A, EGFR and MDM2 genes are well known to occur in diffuse infiltrating astrocytic glioma PXA.[8]

DISCUSSION OF MANAGEMENT

Working diagnosis of Glioma was made and left parietal craniotomy and gross total resection was performed. Post-operatively, patient underwent 25 cycles of radiotherapy and patient is under regular follow-up.

First and foremost, because of the uncommonness of PXA with anaplastic features, the role of standard post-operative therapy has not been established. Eighteen cases of PXA showing anaplastic features at the first presentation have been reported in the literature till now.[6,9,10,11,12]

In Gianni et al study the significance of the mitotic index, the presence of necrosis and the extent of resection were analysed and found that the mitotic index and the extent of resection were the main predictors for recurrence free survival and overall survival rates.[2]

Data to support the role of adjuvant treatment are scanty and sparse. Chemotherapy for PXA has been generally considered ineffective. Many regimens including carboplatin/VP-16, cyclophosphamide, vincristine and

temozolomide were used in the treatment of PXA with anaplastic features, but none worked well.[2,4]

The case reported by Koga et al showed control upto certain extent with stereotactic radiosurgery for the disseminated nodules of PXA with anaplastic features at each relapse. By the use of repetitive stereotactic radiation, the patient was free from neurological deficit for 50 months despite frequent tumour recurrences within a short duration. In Koga et al case report with institution of stereotactic radiosurgery, survival was prolonged upto 66 months even with multiple nodular dissemination for several times.[10]

Macaulay et al[13] addressed the role of adjuvant radiation therapy in these tumours and reported a trend towards better recurrence-free survival with use of adjuvant radiation. However, the difference in overall survival was not statistically significant despite a long follow-up period of fifteen years.

Similarly, Papahill et al[14] reported that the survival curve of patients receiving adjuvant irradiation was not significantly different from those not subjected to it. The extent of surgical resection, which is an independent prognostic factor for survival was not available in either of the studies. There could be an element of selection bias with the less favourable prognostic subset of PXAs (Comprising either subtotal excised or those having adverse histopathological features such as high mitotic index, necrosis, endothelial proliferation) being subjected to adjuvant radiotherapy, while those with gross total excision and favourable histopathological features were kept on observation. Despite this bias, the use of adjuvant radiation therapy was associated with similar overall survival even after long periods of follow-up. Hence, adjuvant radiation therapy should be offered to all unfavourable PXAs.[15]

FINAL DIAGNOSIS

Pleomorphic xanthoastrocytoma with anaplastic features.

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