An unusually aggressive clinical behavior in a case of atypical subependymal giant cell astrocytoma

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Sir,

Subependymal giant cell astrocytoma (SEGA) is a benign tumor that mostly arises in the wall of the lateral ventricle. [1] Though it has been classified as a low grade, WHO Grade 1 tumor, aggressive lesions with metastasis and intratumoral hemorrhages have been described. [2],[3] High grade features like mitoses, focal necrosis, and endothelial proliferations are distinctly rare, but few cases have been reported in the literature with these unusual findings of high-grade tumor, termed as atypical SEGA. [4],[5] Despite anaplasia in histology, the clinical behavior of these patients have been benign, same as typical SEGA. A few cases of atypical SEGA may behave aggressively. [5] We report a case of atypical SEGA mimicking malignant glioma with an unusually aggressive clinical course.

A 5-year-old male child presented with 1 month history of worsening headache with right hemiparesis and facial weakness. Magnetic resonance imaging (MRI) showed a large lesion in the left posterior thalamic region abutting the atrial wall with peripheral enhancement with edema [Figure 1]a and b. There were no markers of tuberous sclerosis complex (TSC). The patient underwent a parietooccipital craniotomy and near total resection of the tumor through transcortical, transventricular route [Figure 1]c and d. Immediate postoperative period was uneventful. The histology showed spindle-shaped to large polygonal tumor cells with many multinucleated giant cells scattered in between [Figure 2]. Large necrotic areas were observed. Few mitotic figures were identified with Ki-67 labelling index around 2-3%. The tumor also showed glial fibrillary acidic protein (GFAP) positivity. The overall features were suggestive of SEGA with atypical features. Within 3 weeks of primary surgery, the patient presented with altered sensorium and progressively worsening right hemiparesis. MRI showed a small recurrent lesion with hydrocephalus. Ventriculoperitoneal shunt was placed following which the sensorium improved. In view of the early recurrence and high-grade histological features, conformal
Radiotherapy was given. A month later, the clinical condition worsened and repeat imaging showed increase in size of lesion with invasion of adjacent structures [Figure 1]e and f. Because of the poor general condition, reexcision of tumor could not be offered and the patient subsequently died within a fortnight. (Figure 1) (Figure 2)

SEGAs are low-grade tumors that seem to arise from the ependymal layer lining the ventricular walls. They usually arise near the foramen of Monro, though other ventricular locations have been described. It has been linked with TSC, though occasionally, they can occur sporadically. [1] It most frequently occurs in the first 2 decades of life. Owing to its periventricular location they frequently obstruct the ventricular pathway producing symptoms of raised pressure with other clinical manifestations being seizure, mental retardation, cognitive disability, and visual disturbances. Computed tomography (CT) shows uniformly dense lesion with occasional peripheral calcifications. These are commonly homogenous lesions with occasional cystic changes that appear isohypointense on T1-weighted (T1W) and hyperintense on T2W images with marked contrast enhancement. Histologically it is composed of three types of cells: Swollen gemistocytic cells, fibrillated spindle cells, and giant ganglion-like cells. [1] There are large polygonal cells with abundant cytoplasm arranged in sheets, clusters, or pseudorosettes with different degrees of vascularization, angiocentric pattern, and calcifications. Few cases may show anaplastic features like nuclear pleomorphism, mitosis, necrosis, microvascular proliferation, and increased Ki-67 index; but traditionally these high-grade features have been said not to impact the diagnosis or prognosis. [4] These high-grade histologic features may easily lead to misdiagnosis of glioblastoma. Absence of atypical small cells, low mitotic count, and Ki 67 index as compared to glioblastoma have been suggested to be the best discriminative features. [4] In spite of presence of necrosis and mitosis, the biological behavior of these atypical tumors is not aggressive and does not mandate radiotherapy and chemotherapy. However, cases of atypical SEGAs have been described in young children with a more rapid growth pattern due to malignant change and hemorrhage. [4],[5]

In our case the tumor recurred very rapidly and the clinical course was almost similar to malignant tumors. In contrast to the traditional belief, our patient showed concordance between high-grade histological features and the clinical outcome. So it is difficult to prognosticate these atypical SEGAs and the clinical course may vary in different cases. Utmost caution should be exercised in managing these patients. Those with aggressive course should probably be treated as other malignant gliomas.

References