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LETTER TO THE EDITOR

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Spontaneous regression of optic chiasmatic glioma in pediatric patients: When to intervene?

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Full Text

Sir,

A four-year-old girl presented with excessive weight gain and progressive diminution of vision for six months. At three months of age, she had history of enlargement of head and down-turning of eyes. Imaging evaluation (CT scan and MRI) revealed enhancing suprasellar mass with parasellar extension, moderate hydrocephalus and involvement of optic chiasm [Figure 1],[Figure 2],[Figure 3]. She was diagnosed with optic pathway glioma (OPG) and was advised treatment but her parents refused. However, she gradually improved symptomatically without any specific treatment. She had deterioration of visual symptoms and hence was again evaluated at four years of age with MRI scan [Figure 1],[Figure 2],[Figure 3], and the examination revealed significant regression of the solid enhancing mass with resolution of hydrocephalus, indicative of spontaneous regression of OPG. Right frontal craniotomy drainage of cystic component and excision biopsy from the solid component was done. Histopathology was pilocytic astrocytoma and there was no feature of necrosis. At presentation, she had preserved neurological function and was observed closely with regular imaging. OPGs are low grade tumours involve optic nerve, optic chiasm, hypothalamus or in few cases may involve the entire optic pathway. [1] Optic nerve, optic chiasma and retrochiasmatic lesions have progressively higher rates of complications and death in that order. [1] Major symptom of OPG is visual loss and main aim of treatment is to preserve vision. There are controversies regarding diagnosis and management of OPGs having spontaneous regression. MRI scans

show dramatic regression of tumor especially in pediatric patients. Radiological interpretation is critical in diagnosis of spontaneous regression. Solid contrast enhancing mass may reduce, while flair/edema may increase. [2],[3] Although these tumors are slow growing indolent LGG, clinical course and natural history is variable making treatment decision difficult. Management option varies from observation only with regular follow-up, surgery either in the form of biopsy, partial debulking to complete excision, chemotherapy or radiotherapy (RT) at the time of progression. Close clinical observation with neuro-ophthalmological examination and serial radiological imaging (MRI scan) evaluation is the first accepted treatment of choice in pediatric patients without clinical progression. [3],[4],[5] Surgical excision is reserved in cases of unilateral optic nerve tumors with poor or no vision, severe disfiguring proptosis and for symptomatic relief of hydrocephalus. OPG with progressive disease are sometimes treated with RT in children over six years and progression free survival is 70-80% in different series, though not demonstrably different than the natural history of these tumors. [1] Late effects of RT are optic neuropathy, endocrinopathy, cataracts, secondary malignancies, developmental abnormalities, neurocognitive dysfunction and vasculopathy (Moya Moya syndrome). New and emerging RT modalities and techniques such as fractionated stereotactic radiotherapy (SCRT) may reduce RT-induced toxicities but remain relatively contraindicated in those under six years of age. Chemotherapy (such as 'Packer's regimen,) is advocated by some in progressive OPGs to delay RT in very young patients under six years. [6] Major dilemma occurs in young patients with stable or persistent but regressing disease and optic pathway glioma task group recommendations are useful in such situations. [4],[5] OPG presenting with partial spontaneous regression need individualized management option depending upon tumor location, radiographic/clinical status, NF1 and risk-benefit of treatment, with all treatments withheld at least until evidence of re-growth is made. As per our knowledge, the present case is the first report that has histological confirmation after regression of the tumor. Previous published biopsy proven cases had biopsy samples prior to regression. The present case demonstrated that regression may show a type I pilocytic tumor still "unremarkable" without extensive areas of necrosis or other unusual traits in its appearance following regression and also without yet undocumented transformation to another lower grade of tumor. Regression of tumor is the inherent property of certain tumors and regression occurs without any transformation. In summary, spontaneous regression has recently become a demonstrated fact and this need to be taken into account when addressing these tumors. Since no effective treatment exists (except for surgery for exophytic components of symptomatic lesions), it is reasonable to simply wait further when spontaneous regression is noted, as exemplified by this case. {Figure 1} {Figure 2} {Figure 3}

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