Letter to the Editor

A rare case of pinealoblastoma in adult with complete response to treatment

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Dear Editor,

Supratentorial primitive neuroectodermal tumors (SPNETs) usually arise from cerebrum or pineal gland and accounts for about 2.5% of pediatric brain tumors.[1] SPNET arising from the pineal gland is known as pinealoblastomas, which are highly malignant embryonal tumors usually affects children and adolescents and are very rare in adults.[2]

A 42-year-old male patient presented with a history of a headache and blurring of vision for 15 days. There was no history suggestive of any focal neurological deficits. Neuroimaging [Figure 1] showed a well-defined mass lesion of size 2 cm × 1.5 cm in the posterior aspect of the third ventricle along the hypothalo-pineal axis possibly a pineal gland tumor.

He underwent craniotomy and maximal safe resection of the tumor. Histopathological analysis showed pinealoblastoama (the WHO Grade IV) with MIB index of 30%. Immunohistochemistry was done which showed synaptophysin and chromogranin positivity. The postoperative imaging showed residual mass lesion with involvement of medial thalamus. The patient received craniospinal irradiation with vincristine-based concurrent chemotherapy (five cycles). Imaging at re-evaluation showed near-complete resolution of the tumor [Figure 2]. The patient received six cycles of adjuvant chemotherapy with vincristine, procarbazine, and lomustine. The imaging at the completion of the treatment showed complete resolution of the lesion with no residual abnormalities.

Pinealoblastomas in an adult is a rare entity. In a retrospective review of 48 patients with the pineal gland tumors over 10 years, which included 35 children and 13 adults, pinealoblastoama was diagnosed only in six (12.5%) cases.[3] In a review of treatment outcomes in pinealoblastoama from tertiary cancer hospital in India, there were 17 patients of pinealoblastoama with a median age of 14 years.[4] After a median follow-up of 30 months following surgery and adjuvant chemoradiotherapy, disease recurrence and death were noted in 7 (41%) and 3 (17%) patients, respectively, with estimated median recurrence-free survival of 5.4 years. There were no standard guidelines to manage this malignancy in adults.

This case depicts a rare malignancy of the central nervous system which presented in an uncommon age group of the fifth decade. Although there are no standard guidelines for treatment of pinealoblastoama in an adult, surgical resection followed by craniospinal irradiation with boost to tumor bed along with chemotherapy gives a good response. Clinical trials with a larger number of patients are needed to look for the best treatment modality.

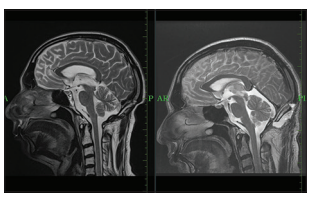

Figure 1: Right: Presurgery: A circumscribed lesion with well-defined margins is seen involving the pineal gland, which has mixed intermediate and high signal on sagittal T2-weighted images. Left: Postsurgery/postchemotherapy/radiotherapy: Evidence of craniotomy and postoperative defect is noted in occipital bone. No altered signal intensity lesion is appreciated

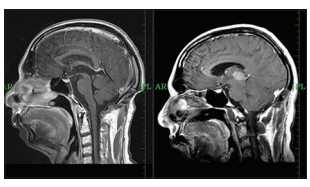

Figure 2: Right: Presurgery: A well circumscribed tumor arising from the pineal gland shows contrast enhancement on sagittal T1- weighted image. Left: Postsurgery/postchemotherapy/radiotherapy: Evidence of craniotomy and postoperative defect is noted in occipital bone. No altered signal intensity lesion is appreciated

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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