Vanishing tumor of the tectal plate – stressing the need for a stringent radiological surveillance: A review of possible pathogenetic mechanisms

Sir,

A spontaneously regressing intracranial tumor, in the absence of any intervention known to cause its regression, is called a 'vanishing tumor' or a 'ghost tumor.' It is reported in 1:60,000 to 1:100,000 cases of malignant tumors.[1-5] The commonest pathology in these spontaneously resolving lesions is usually presumed to be a primary CNS lymphoma[PCNSL].[1-7]

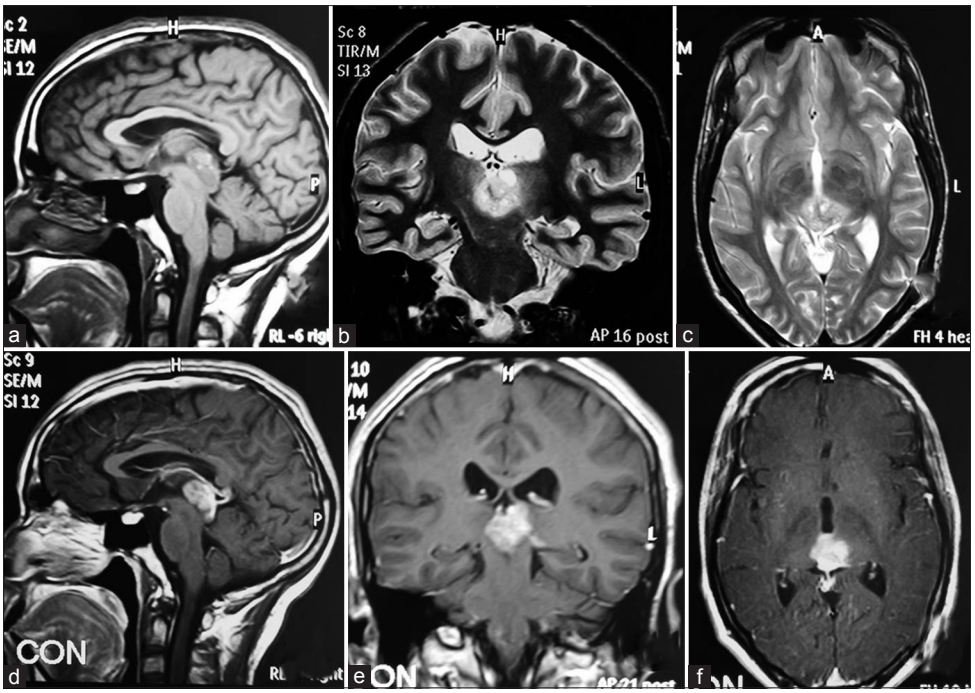

Figure 1: Initial MRI study of the patient. The top panel depicts the plain images: (a) T1-weighted (W) sagittal, (b) T2W coronal, and (c) T2W axial. The tumor is seen as a well-defined lesion in the pineal region iso- to hyperintense to the gray matter. A few cysts are visualized in b and c. There was no significant hydrocephalus. The bottom panel demonstrates corresponding MRI sections after gadolinium administration revealing a signifi cant postcontrast enhancement. (d) Sagittal, (e) Coronal and (f) Axial images define the relationship of the lesion to the deep venous system

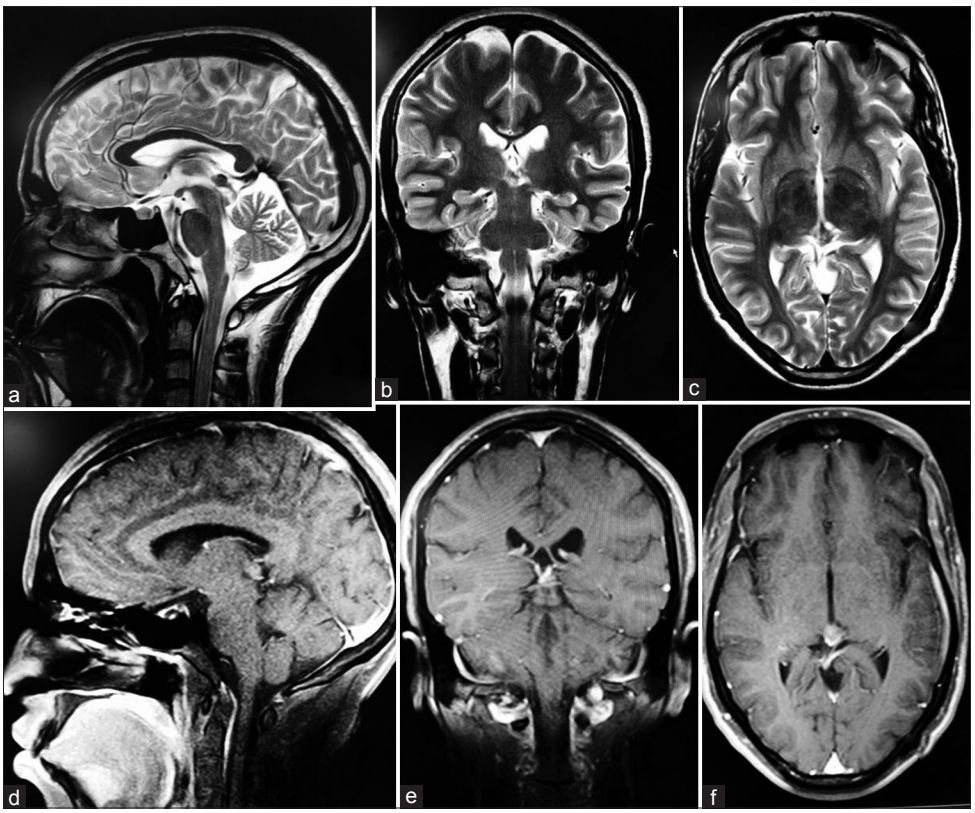

Figure 2: Follow-up MRI study of the patient. The top panel depicts the plain images: (a) T2W sagittal, (b) T2W coronal, and (c) T2W axial. The lesion demonstrates more than 90% resolution. (d-f) Postcontrast images, with a small tumor seen in the axial section

Table 1: Review of literature of spontaneous regression of germinomas in proximity to the pineal region

Year	Authors	Age (years)/ sex	Location	Intervention	Pathology	Serum/CSF markers of germinomas	Use of steroids	Duration to F/up MRI or Sx	Number of CT scans performed	Outcome	F/up
1997	Ide *et al*.[3]	21/M	Third ventricle	VP shunt, excision of suprasellar component	NA (germinoma for suprasellar tumor)	−ve	Yes	NA	2	Good	1 year
1999	Fujimaki *et al*.[6]	39/M	Pineal, cerebellar vermis	Excision of the vermian lesion	NA (germinoma for vermian lesion)	−ve	Yes	20 days	2 + angiography	Good	2 years
2000	Murai *et al*.[4]	17/M	Pineal	VP shunt	Germinoma	NA	No	2 months	5	Good	3 years
2011	Ono *et al*.[5]	15/M	Pineal	Endo Bx	Germinoma	−ve	No	9 days	NA	Good	NA
2012	Okita *et al*.[1]	36/M	Thalamus, midbrain	Bx	Germinoma	NA	Yes	1.5 months	NA	Good; recurrence 6 months	5 years
2015	Dadlani *et al*. (present case)	22/M	Pineal	Nil	NA	−ve	Yes	9 months	0	Good	-

M - Male, F/up - Follow-up, Sx - Surgery, NA - Not available, GP - Globus pallidus, −ve - Negative, Endo Bx - Endoscopic biopsy, Bx - Biopsy

A 22-year-old male student from Kenya presented with a 9-month history of diplopia and headaches. His initial magnetic resonance imaging (MRI) revealed a pineal lesion [Figure 1], for which he received a tapering course of oral steroids only. He had partial remission of the clinical symptoms and was planned for a surgical excision based on the original MRI when he was seen at our institute. However, a repeat MRI [Figure 2] was recommended, which revealed the near-complete resolution of the tumor. He was managed non-surgically and has been advised surveillance MRI scan in 6 months. Cerebrospinal fluid (CSF) test results and blood markers such as human chorionic gonadotropin (hCG), β-subunit of hCG, alpha fetoprotein (AFP), and carcinoembryonic antigen (CEA) were within normal range.

Recently, an attempt was made at classifying 'ghost tumors' into tumor-like lesions spontaneously regressing on follow-up ('vanishing tumors'), space-occupying lesions that are non-neoplastic ('tumor-like lesions'), and variations in normal anatomy mimicking a tumor ('false tumors').[2]

However, a 'vanishing' brain tumor has generally been defined as a tumor that undergoes a spontaneous reduction in size of more than 70% without any definitive treatment (other than steroids).[1,3-6]

A review of literature was undertaken in three databases, *viz*., MEDLINE, Scopus, and EMBASE, and only five articles were identified that fit the search criteria [Table 1].

Approximately, 60%–70% of all pineal region tumors are known to be germinomas, with 90% occurring in the young adult male population.[7] Although the present case lacked a histological diagnosis, the patient profile and radiological findings strongly favored the possibility of a germinoma.

There have been several theories proposed for the spontaneous regression of tumors, most notably, immunological responses, spontaneous apoptosis, possible effect of diagnostic X-rays (CT scans), effects of CSF drainage, and corticosteroid therapy.[5] Infection and fever have also been implicated as possible stimuli for regression.[3]

Surgical intervention, most often a biopsy, associated with tumor regression in approximately 40% of tumors, is another proposed hypothesis.[3,4]

The etiopathogenesis of the vanishing tumor remains conjectural. Although histological diagnosis is not always possible, as illustrated in this case and in the literature reviewed, stringent radiological surveillance of patients with tumors in the pineal region is recommended, especially just prior to biopsy or surgical excision, to avoid intraoperative surprises.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Ravi Dadlani[1,2], L. Vijay Kumar[3]
[1]Department of Neurosurgery, Apollo Specialty Hospital, Nellore, Andhra Pradesh, Departments of [2]Neurosurgery and [3]Radiology, MaxCure Hospitals, Hyderabad, Telangana, India
E-mail: ravi.dadlani@gmail.com

References

1. Okita Y, Narita Y, Miyakita Y, Ohno M, Fukushima S, Maeshima A, *et al*. Long-term follow-up of vanishing tumors in the brain: How should a lesion mimicking primary CNS lymphoma be managed? Clin Neurol Neurosurg 2012;114:1217-21.
2. Frassanito P, Tamburrini G, Massimi L, Caldarelli M, Di Rocco C. Ghost tumors of the central nervous system: Definition, clinical implications and proposal of classification. World Neurosurg 2015;84:663-70.
3. Ide M, Jimbo M, Yamamoto M, Hagiwara S, Aiba M, Kubo O. Spontaneous regression of primary intracranial germinoma. A case report. Cancer 1997;79:558-63.
4. Murai Y, Kobayashi S, Mizunari T, Ohaki Y, Adachi K, Teramoto A. Spontaneous regression of a germinoma in the pineal body after placement of a ventriculoperitoneal shunt. J Neurosurg 2000;93:884-6.
5. Ono H, Shin M, Takai K, Oya S, Mukasa A, Saito N. Spontaneous regression of germinoma in the pineal region before endoscopic surgery: A pitfall of modern strategy for pineal germ cell tumors. J Neurooncol 2011;103:755-8.
6. Fujimaki T, Mishima K, Asai A, Suzuki I, Kirino T. Spontaneous regression of a residual pineal tumor after resection of a cerebellar vermian germinoma. J Neurooncol 1999;41:65-70.
7. Fang AS, Meyers SP. Magnetic resonance imaging of pineal region tumours. Insights Imaging 2013;4:369-82.

Access this article online	
Website: www.neurologyindia.com	**Quick Response Code**
DOI: 10.4103/0028-3886.177614	

How to cite this article: Dadlani R, Kumar LV. Vanishing tumor of the tectal plate – stressing the need for a stringent radiological surveillance: A review of possible pathogenetic mechanisms. Neurol India 2016;64:351-4.