

Mixed Germ Cell Tumour of the Suprasellar Region

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Abstract

Background and Importance: Intracranial germ cell tumour is a rare entity and encompasses germinomatous and non germinomatous neoplasms. These usually affect the pineal and hypothalamic regions and manifest with characteristic clinical features based on their location. A combination of mature cystic teratoma and germinoma is rare.

Keywords: Germinoma; Teratoma; Suprasellar; Intracranial; Germ cell tumour.

Clinical Presentation

We present a case of mixed germ cell tumour in the suprasellar region with the clinical triad of visual disturbances, diabetes insipidus and hypopituitarism along with radiological and histopathological details.

Introduction

Primary germ cell tumours (GCT) of the central nervous system are uncommon accounting for 0.5% of all tumours

arising in this location. These tumours usually affect paediatric population and comprise 2-3% of intracranial tumours [1,2]. GCTs are categorized as germinomatous and nongerminomatous tumours, the latter comprising of teratomas, choriocarcinomas, embryonal carcinoma and mixed tumours. The most common location of these tumours is the pineal region and the suprasellar region. Tumours affecting the suprasellar region cause dysfunction of the hypothalamic pituitary axis and are also referred to as neurohypophyseal tumours [3,4].

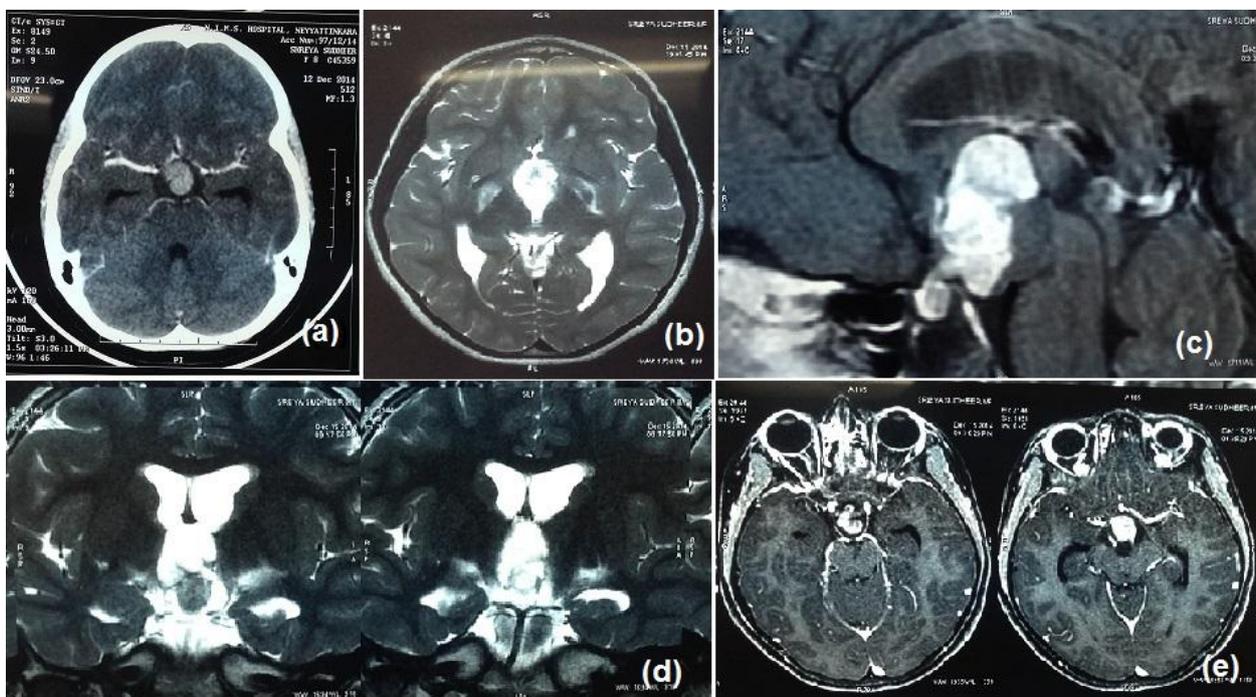
Germinoma, frequently affects suprasellar regions, however, mature cystic teratoma is extremely rare [5]. We report a case of mixed germ cell tumour containing mature cystic teratoma and germinoma of the suprasellar region.

Case Report

An eight-year old girl presented with a history of headache, blurring of vision, hyperphagia and polydipsia since one and a half years. On physical examination she was moderately built and nourished and had not attained puberty. Neurological and ophthalmic examination was normal.

Hormonal assays revealed evidence of deficiency of pituitary functions with hypothyroidism, hypocortisolism and diabetes insipidus. Biochemical markers including serum alpha fetoprotein (AFP) and beta human chorionic gonadotropin (β -HCG) and Carcino-embryonic antigen (CEA) levels were normal. Computed tomography (CT) and Magnetic resonance imaging (MRI) of the brain revealed a well-defined lobulated contrast enhancing solid space occupying lesion measuring 3.2x2.4x2.3cm with low and high signal areas, filling and expanding suprasellar cistern (Figure 1).

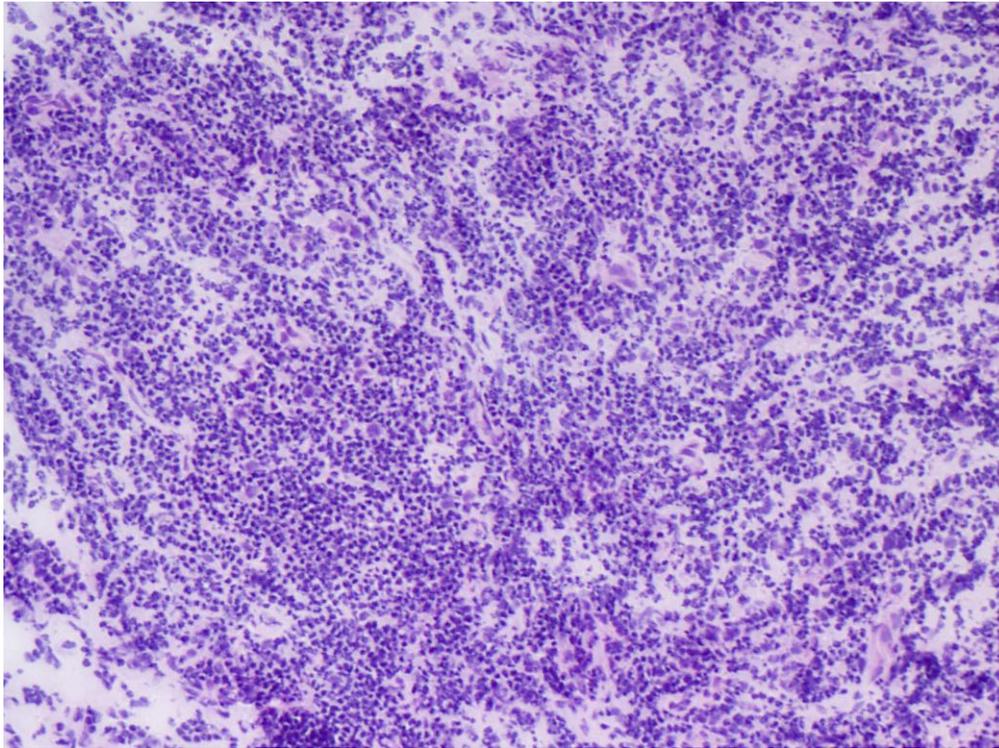
Figure 1: (a) Contrast CT Brain, axial cut showing a well circumscribed, contrast enhancing lesion in the suprasellar region. The lesion seems to be hyperintense on T2W MRI (axial and coronal cuts) (b) (d). Sagittal MRI image, contrast series showing a contrast enhancing suprasellar lesion extending into the hypothalamus with pituitary seen separately (c). The lesion has taken up gadolinium as seen in the axial MRI T1 contrast series (e).



Clinical diagnosis of low grade hypothalamic/chiasmal glioma was made. Decompression of the lesion was done by right pterional craniotomy with orbitozygomatic extension. Intra operatively, greyish white predominantly solid lesion with mixed consistency was seen in the suprasellar area. The lesion was seen merging with the

hypothalamus and invading the third ventricle. A piece of tissue was sent for frozen section. Frozen section imprints of the specimen were paucicellular. Sections showed numerous small round cells with scant cytoplasm with few interspersed larger cells suggesting a possibility of small round cell tumour (Figure 2).

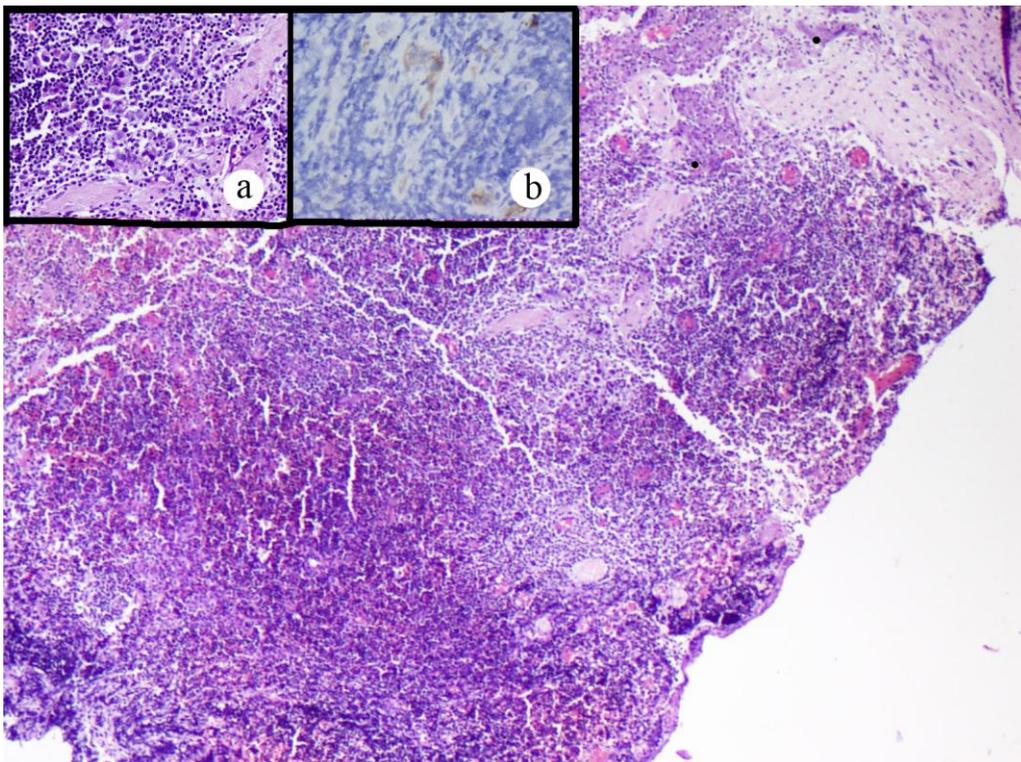
Figure 2: Photomicrograph showing frozen section with numerous dispersed small round cells (10X)



Further, more tumour tissue was sampled and sent for histopathological examination. The paraffin sections of the frozen section sample revealed a tumour composed of nests, cords and single large polygonal cells with moderate amount of eosinophilic cytoplasm, round nuclei with dispersed

chromatin and prominent nucleoli surrounded by dense lymphocytic infiltrate. The large polygonal cells demonstrated immunoreactivity to CD117 and stained negative with AFP (Figure 3).

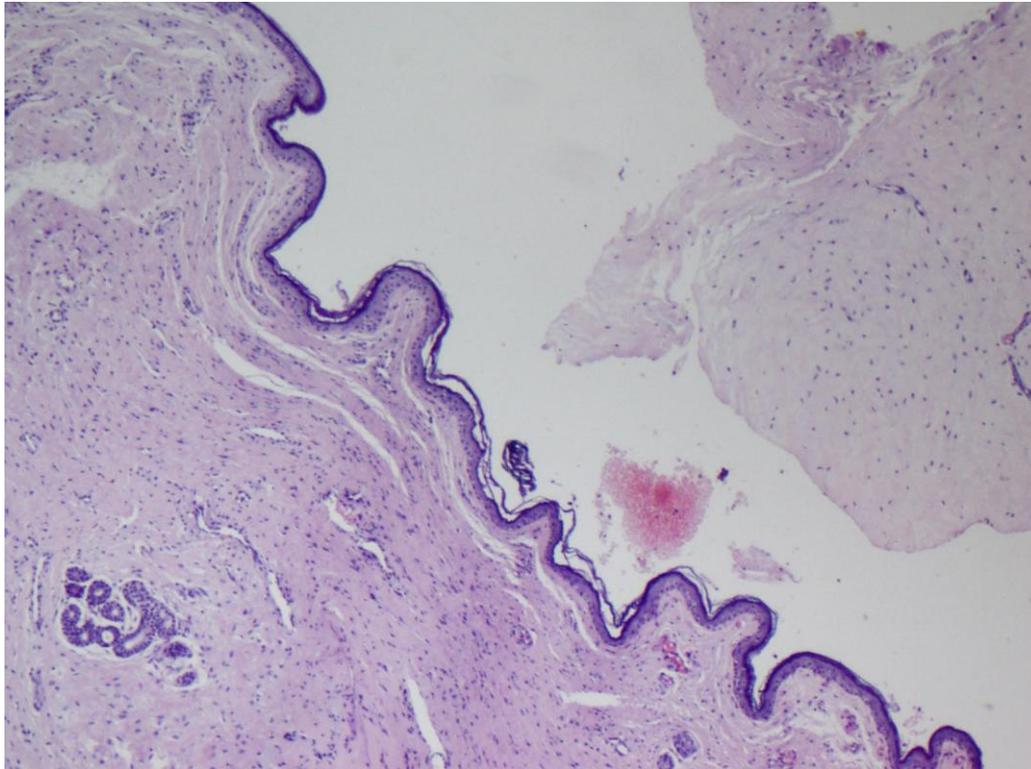
Figure 3: Photomicrograph showing germinoma with numerous lymphocytes and intervening clusters of large germinoma cells. (H&E, 20X) Inset a. H&E, 40X b. CD117, 40X.



The morphology of the tumour was suggestive of a germinoma. Microscopy of the sample sent later, in addition, also revealed a mature cystic teratoma composed of polypoid structures lined by stratified squamous and respiratory epithelium and luminal keratin flakes. The

underlying stroma showed mature glial tissue, eccrine and sebaceous glands, smooth muscle fibres and congested vessels (Figure 4). Final diagnosis of a mixed germ cell tumour, with mature cystic teratoma and germinoma, was rendered.

Figure 4: Photomicrograph showing mature teratoma with keratinized stratified squamous epithelium overlain on stroma with adnexal structures (H&E, 4X)



Discussion

Primary intracranial GCT account for 0.5-11% of all intracranial tumours and usually affect the midline, commonly involving the pineal region (50%) and suprasellar (40%). Other locations include ventricles, basal ganglia, cerebrum and posterior fossa. These tumours show a male preponderance and affect individuals in the first and second decade of life. Suprasellar lesions in paediatric age group should always raise a suspicion of germ cell tumour. Clinically, suprasellar lesions present with classic triad of visual disturbances, diabetes insipidus and features of pituitary dysfunction. They may also present with cranial nerve palsies or mimic pituitary apoplexy [1, 6,7]. Our patient also presented with the classic triad associated with suprasellar lesions.

On clinical presentation, suprasellar GCT mimic craniopharyngioma and pituitary adenoma. However, the presence of classic symptomatic triad supported by neuroimaging, biochemical markers and histopathology, helps in making the correct diagnosis [3, 7]. Neuroimaging gives a clue to the type of GCT by analyzing the cystic and solid components along with the infiltrating nature, necrosis

and haemorrhage within the tumour; the latter features being more common in high grade GCTs [7]. Elevated serum and cerebrospinal fluid levels of AFP and HCG are helpful in identifying intracranial GCTs. However, radiology and tumour markers, both are not very sensitive in subclassifying them. In a study done by Matsutani et al, [4] none of the patients diagnosed with mature teratoma or germinoma had elevated serum AFP or HCG levels, as seen in our case [4].

Histopathologically, intracranial GCTs resemble their gonadal counterparts. They are categorized as germinomas, choriocarcinomas, embryonal cell carcinomas, yolk sac tumours, teratomas and mixed germ cell tumours. In the suprasellar region, germinoma is the most common GCT. It is often accompanied by dense lymphocytic infiltrate and may be associated with elevated serum pituitary antibodies, making it difficult to differentiate it from hypophysitis. Identification of the germinoma cells in the biopsy clinches the diagnosis. Immunoreactivity with CD117 helps in confirming the diagnosis as seen in the present case. Other markers that aid in diagnosing germinomas include OCT4 [8 - 10].

Mature teratoma in this region is rare with few cases reported in literature. It is characterized by the presence of derivative of endoderm, mesoderm and ectoderm [5]. It is also essential to identify all components of germ cell tumour in a mixed type like yolk sac tumour and embryonal carcinoma, which were excluded on morphology and immunohistochemistry in our patient. The present case showed predominantly lymphoid cells in the frozen section with few scattered germinoma cells. On examination of the entire specimen, a diagnosis of mixed germ cell tumour with germinoma and mature cystic teratoma was made.

The pathogenesis of intracranial tumour is unclear. Germinomas are hypothesized to arise from primordial, undifferentiated germ cells while nongerminomatous tumours are the result of embryonal cell nests, at different stages of development, enfolded into neuraxis during formation of the neural tube [11].

Treatment modalities include surgical resection, radiation therapy and chemotherapy. Mature teratomas can be treated by surgical resection. Germinomas respond well

to radiation therapy following surgical resection. However, combination of radiotherapy and chemotherapy is essential for treatment of non germinomatous tumours [2, 4, 10].

Pure germinomas and mature teratomas have good prognosis while mixed GCTs with germinoma, mature teratoma, immature teratomas or teratomas with malignant transformation have intermediate prognosis. All other non germinomatous tumours have poor prognosis [4].

Conclusion

Suprasellar mixed germ cell tumour comprising teratoma and germinoma is a rare occurrence. It is important to identify all components of the disease as it significantly affects the treatment protocol and clinical outcome. This emphasizes the need for correlation of clinical details, neuroimaging, biochemical markers along with vigilant histological examination of the entire lesion.

References

1. Smith AA, Weng E, Handler M, Foreman NK. Intracranial germ cell tumors: a single institution experience and review of the literature. *J Neurooncol* 2004; 68: 153-9.
2. Li Y, Zhang Y, Xu J, Chen N. Successful surgical treatment of mature teratoma arising from the sella. *J Clin Med Res.* 2015; 7:122-5.
3. Wildenberg LE, Vieira Neto L, Taboada GF, Moraes AB, Marcondes J, Conceição FL, et al. Sellar and suprasellar mixed germ cell tumour mimicking a pituitary adenoma. *Pituitary.* 2011;14(4):345-350.
4. Matsutani M, Sano K, Takakura K, Fujimaki T, Nakamura O, Funata N, Seto T. Primary intracranial germ cell tumors: a clinical analysis of 153 histologically verified cases. *J Neurosurg.* 1997;86(3):446-455.
5. Sweiss RB, Shweikeh F, Sweiss FB, Zyck S, Dalvin L, Siddiqi J. Suprasellar mature cystic teratoma: an unusual location for an uncommon tumor. *Case Rep Neurol Med.* 2013;180497.
6. Furtado SV, Thakar S, Ghosal N, Hegde AS. Atypical presentation of pediatric mixed germ cell tumors in the sellar-suprasellar region. *Neurol India.* 2012;60(1): 90-93.
7. Nada R, Mohan H, Dhir SP, Mukherjee KK, Kak VK. Suprasellar malignant mixed germ cell tumour presenting as craniopharyngioma. *Neurol India* 48(4):381-384,2000
8. Gutenberg A, Bell JJ, Lupi I, Tzou SC, Landek-Salgado MA, Kimura H, Su J, Karaviti LP, Salvatori R, Caturegli P. Pituitary and systemic autoimmunity in a case of intrasellar germinoma. *Pituitary.* 2011;14(4):388-394.
9. Kishore M, Monappa V, Rao L, Kudva R. Mixed malignant germ cell tumour of third ventricle with hydrocephalus: a rare case with recurrence. *J Clin Diagn Res.* 2014;8(11):FD03-5.
10. Yuping Gao, Jiyao Jiang, Qiang Liu. Clinicopathological and immunohistochemical features of primary central nervous system germ cell tumors: a 24-year experience. *Int J Clin Exp Pathol.* 2014;7(10):6965-6972.
11. Sano K. Pathogenesis of intracranial germ cell tumors reconsidered. *J Neurosurg.* 1999;90:258-264.

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