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SELLAR/SUPRASELLAR GERMINOMA MASQUERADING AS PITUITARY ADENOMA

Pathology

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ABSTRACT

We report a case of sellar/suprasellar germinoma in a 17-year-old female which was misdiagnosed as pituitary adenoma both clinically and on imaging. Patient presented with severe headache of short duration and diminished visual acuity in left eye. Endocrinological work up revealed hyperprolactinemia and markedly reduced blood cortisol. MRI revealed homogenously enhanced intrasellar mass with suprasellar extension showing characteristic dumbbell configuration (snowman sign). Provisional diagnosis of pituitary adenoma was made and transsphenoidal resection of tumor was performed which on histology and immunohistochemistry proved to be germinoma.

Sellar/suprasellar germinoma without diabetes insipidus is considered to be very difficult to diagnose preoperatively and the differentiation from pituitary adenoma based on MRI findings was not possible in this case. However it is suggested that in a child with visual impairment, endocrine dysfunction and sellar/suprasellar mass a possibility of germ cell tumor should always be considered in differential diagnosis and a frozen section has to be performed to avoid excessive dissection.

KEYWORDS

Sellar/ Suprasellar; Germinoma; Pituitary adenoma

INTRODUCTION:

CNS germ cell tumors principally affect children and adolescents and seem to be more prevalent in eastern Asia than in the USA and European countries. It represents approximately 3% of primary pediatric brain tumors in the west, whereas in Asia the incidence from 8-15% was observed in various series.[1,2] CNS germ cell tumor preferentially affect the midline; 80% or more arise in structures around the third ventricle and pineal gland being most common site of origin. The second most common site is suprasellar region and in particular the neurohypophyseal/infundibular stalk. Diabetes insipidus is the most common presentation and has been reported in almost all patients of suprasellar germinomas.[3] Herein, we present a case of sellar and suprasellar germinoma without clinical or biochemical evidence of diabetes insipidus. Other clinical findings, hormonal profile and imaging of the case were also mimic pituitary adenoma.

Case Report:

A 17 year old female presented with severe headache since three days and blurring of vision in left eye since 20 days. She had a history of off and on headache for last one month. On neurological examination she was normal except diminished left visual acuity. She was well oriented to time, place and person and no abnormality was detected on cardiovascular, abdominal and respiratory examination. Investigation revealed normal haemogram, kidney function tests, serum electrolyte and coagulation profile. Viral markers were negative. Urine osmolality was found within normal range. Hormonal examination revealed hyperprolactinemia, PRL- 782.4 uIU/ml (normal range 44.5-390 u IU/ml) and markedly reduced blood cortisol, <1.50 nmol/L. Thyroid profile was normal. Sagittal T1 weighted post contrast MR images revealed sellar mass with suprasellar extension showing characteristic Dumbbell configuration “Snowman sign” (Fig. 1A). Axial T1 weighted post contrast MR imaging revealed homogenously enhancing suprasellar mass reaching up to anterior third ventricle encasing left middle cerebral artery (Fig. 1B). Diffusion weighted images showing restricted diffusion suggests dense cellularity. No evidence of cystic changes, hemorrhage or calcification was noted. Based on above findings possibility of pituitary adenoma was suggested and thereafter transsphenoidal resection of tumor was performed. Histology revealed sheets of cells with round, vesicular, centrally located nuclei, irregular macronucleoli, discrete cell membrane and abundant clear cytoplasm with intervening delicate fibrous septa which was infiltrated by small lymphocytes (Fig. 2A, B). Tumor cells were seen infiltrating the pituitary tissue. On IHC tumor cells were showing strong cell membranous labeling for CD117 (Fig. 2C) and nuclear reactivity for OCT4. Synaptophysin was positive in pituitary tissue and KI-67 labeling index was up to 60% (Fig. 2D).Diagnosis of sellar/suprasellar germinoma was made. During postoperative period patient developed diabetes insipidus probably due to hypothalamic injury which was managed and further management was planned.

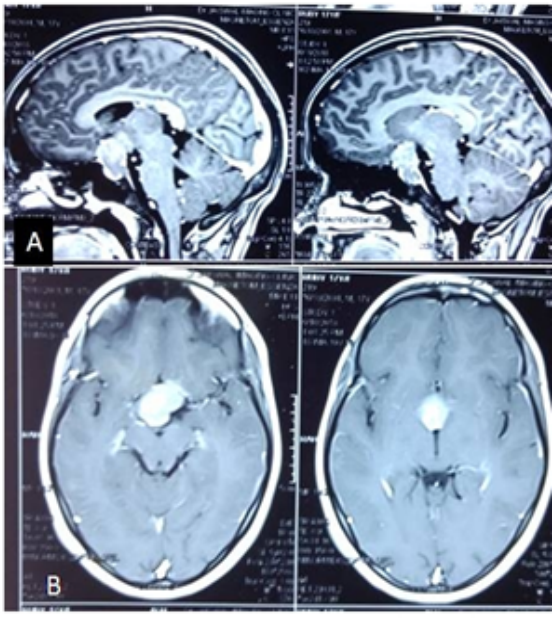


Figure 1. A: Sagittal MR T1-weighted image showing dumbbell configuration.B: Axial MR T1-weighted post contrast image showing homogenously enhanced suprasellar mass reaching anterior third ventricle encasing left middle cerebral artery.

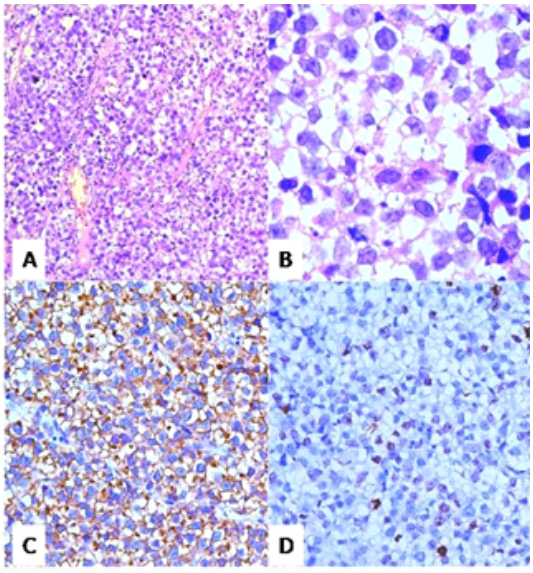


Figure 2. A: H & E staining showing sheets of tumor cells separated by fibrovascular stroma with lymphocytic infiltration (H and E, x100). B: The cells are pleomorphic with vesicular nuclei and frequent mitosis (H and E, x200). C: Immunohistochemistry showing strong cell membrane labeling for CD117. D: and high KI-67 labelling index.

DISCUSSION:

Pediatric suprasellar tumors are different from their counterparts in adults where pituitary adenoma is the most common tumor. The most common suprasellar tumor in children is craniopharyngiomas followed by chiasmatic/hypothalamic low-grade gliomas (optic nerve glioma) and germinomas. Among germ cell tumors of CNS; suprasellar location is second most common location and accounts 22.9% in incidence.[3] Germinomas are the prevalent tumor type in the suprasellar compartment and basal ganglionic/thalamic region, while non-germinomatous germ cell tumors dominating at other sites. An interesting observation is that while the majority of pineal germ cell tumors are encountered in boys, most of the suprasellar germ cell tumors are seen in girls.[3] McCathy et al. after analyzing large tumor registries from Japan and United states found male preponderance in the pineal region tumors with incidence-rate ratio (IRR) male-to female was 11.5:1, compared to IRR of 2.1:1 for tumors located in non pineal region.[1] Similar observation is noted by Borde et al. in an Indian scenario where female constituted 72.7% of GCTs in suprasellar region.[4]

Sellar/suprasellar Germinoma characteristically presents with a classical clinical triad of diabetes insipidus, pan-hypopituitarism and visual complaints.[5] Clinical features of Craniopharyngioma are non-specific essentially presents with features of visual disturbances, endocrine deficiencies, and increased intracranial pressure. Incidence of diabetes insipidus is seen in up to 17% of children only [6]and hyperprolactinemia is found in few due to the *stalk effect*. Optic nerve glioma; histologically majority of them are pilocytic astrocytomas often produce visual symptoms, proptosis, visual loss, endocrine/hypothalamic disturbances and obstructive hydyocephalus. Sellar/suprasellar germinoma on CT and MRI are usually solid masses that are iso- or hyperdense relative to grey matter and prominent contrast enhancing.[6] With T1-weighted images they are hypointense/isointense and isointense/hyperintense on T2-wighted images. They are not prone to calcification or cystic changes which are common in thalamic and basal ganglion germinoma and other germ cell tumors. Craniopharyngiomas can be accurately diagnosed on CT and MR with contrast enhancement of solid portion and cyst capsule as well as typical calcification. Variable radiological pictures are noticed in Optic nerve glioma which are often iso to hypointense on T1-weighted images with variable contrast enhancement. On T2-weighted images they are hyperintense centrally with low signal intensity at the periphery representing the dura. A consistent finding of hypointensity in relation to orbital fat on imaging along with predominance of visual symptoms often visual loss and/ or proptosis help to distinguish optic nerve gliomas from suprasellar germinomas.

Pituitary adenomas comprise 2.7 % of suprasellar tumors in childhood, and Prolactinoma is the commonest pediatric pituitary adenoma seen in adolescents.[7] Pubertal delay/failure, menstrual abnormality, galactorrhea, headaches/visual disturbances are the common presentation. On T1 MR, pituitary adenomas are generally hypointense relative to surrounding pituitary with delayed enhancement. T2- weighted images are variable, but often a little hyperintense. Macroadenomas generally show a heterogeneous enhancement pattern, in contrast to intense enhancement of germinoma and this finding should be closely observed. [8]

Our report adds to unusual clinical presentation of sellar/suprasellar germinoma without diabetes insipidus and with hyperprolactenemia along with homogenously enhanced intra- and suprasellar dumbbell tumor. Based on clinical findings and imaging the case could not be differentiated from pituitary adenoma preoperatively. [9]

CONCLUSION:

The present report emphasize a diagnostic dilemma, but more important is a reminder to the clinicians that in case of sellar/suprasellar lesion in paediatric population a possibility of germinoma should always be considered even if no classical symptoms and laboratory and MR findings are found. A per-operative frozen section is to be performed in such cases to avoid excessive dissection.

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