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Case report

Bifocal primary intracranial germinoma in a child. Case report

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Background. *Bifocal primary intracranial germinal tumors are rare. Only 5-10% of all germ cell tumors are found both in the suprasellar and pineal region.*

Case report. *In presented patient we found two primary intracranial germinomas in pituitary and pineal gland that were successfully operated. Radiological properties of germinomas and differential diagnosis are discussed.*

Conclusions. *Although the definite histological diagnosis cannot be achieved by computer tomography and/or magnetic resonance images alone, a detailed analysis of neuroradiological images is useful for predicting the histological diagnosis.*

Key words: computed tomography; magnetic resonance; germinoma, intracranial

Introduction

Germ cell tumors located in the central nervous system (CNS) represent less than 4% of the intracranial tumors and affect primarily children and young adults. These tumors frequently arise in the suprasellar and pineal region and in the midline structures around the third ventricle. Germ cell tumors can be divided as germinoma and nongerminomatous tumors regarding the histology. Only 5-10% of all germ cell tumors are found both in the suprasellar and pineal region[1] mostly as germinomas.

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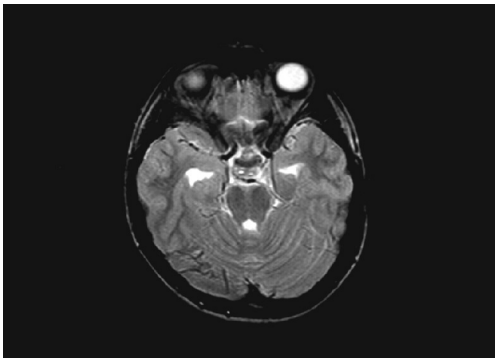
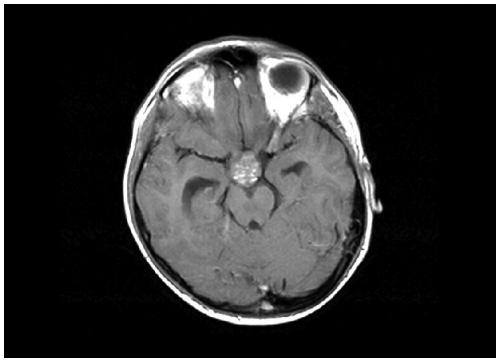
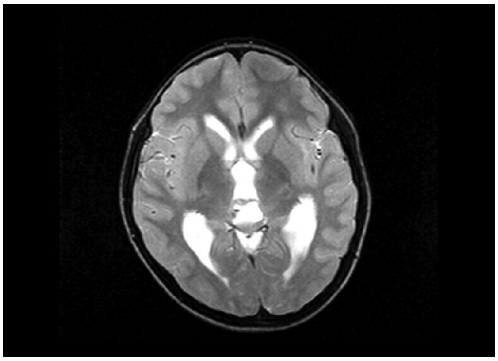
We present a case of isolated primary bifocal germinoma of CNS in a child.

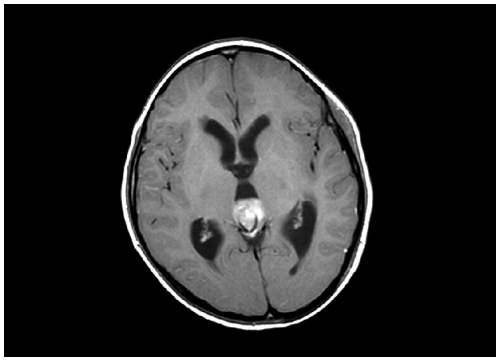
Case report

An eleven-year-old boy with nocturnal disuria was admitted to our hospital because of vomitting and frontal and retro orbital headache, which was depended on the position of the head. There were no vertigo or visual disturbances. We found signs of premature puberty, bilateral papillary edema and raised serum concentrations of PRL. The computed tomographic (CT) scan showed homogenous, well-delineated suprasellar tumor with obstructive hydrocephalus. An additional pineal region tumor was found on magnetic resonance images (MRI) of the brain. Both tumors had similar neuroradiological characteristics (Figures 1-4). We didn't









Figures 1,2. MRI (SE T2WI) in axial plane. Well-delineated oval lesions in sella turcica and pineal gland. Peripheral solid part is isointense, central cystic part is hyper intense. Hydrocephalus.

Figures 3,4. MRI (SE T1WI) in axial plane. Solid part of tumors is isointense with cortex.

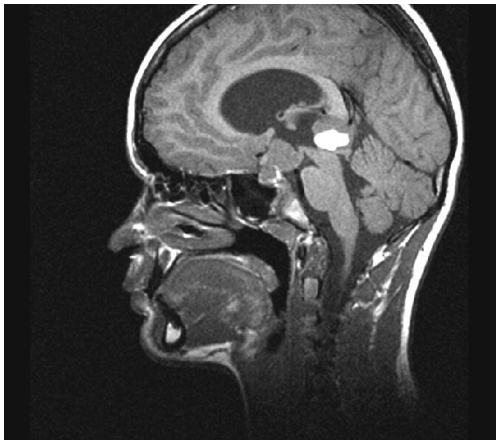
find similar lesions in testicles or elsewhere in the body.

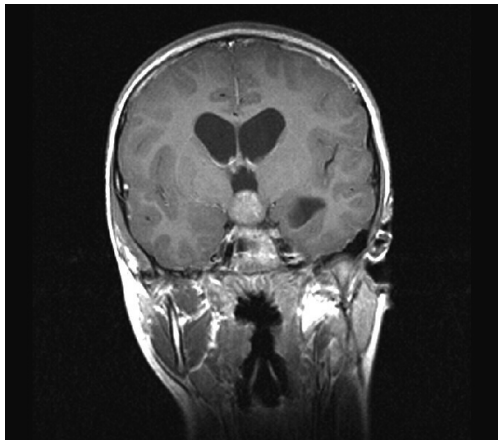
The patient had two operations. Surgeons resected first pituitary lesion. The macroscopically gray tumor had a peripheral solid vascular part and a central soft avascular one. The tumor compressed both optical nerves and the inferior part of optical chiasm and grew into infundibulum and through sellar diaphragm into sella turcica (Figure 5). Hypophysis was completely destroyed. The pineal tumor was resected by the second operation (Figure 6). This tumor grew from the third ventricular wall posteriorly in the pineal region to both basal veins and was macroscopically similar to the previously reseceted suprasellar tumor (Figures 7,8). The histological examination of both specimens showed mature germinoma with the positive reaction to alkaline phosphatase. All other tumor markers were negative.

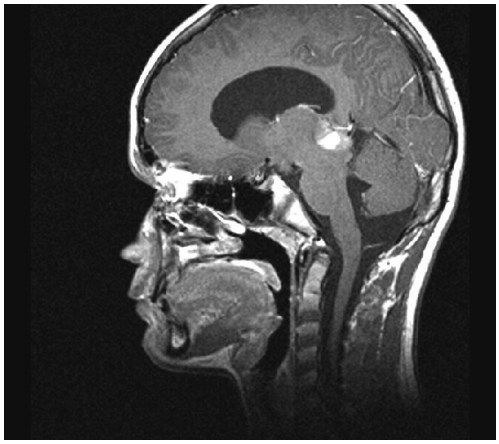
The patient had upper gaze failure and diabetes insipidus postoperatively. Cerebrospinal liquor for malignant cells was negative. No tumor was found on postoperative MRI (Figures 9,10). The additional radiotherapy of the third ventricular region with 4000 cGy was performed. The boy was treated with a substitute hormonal therapy and antiepileptic prophylaxis. The child is in complete remission 15 months after the diagnosis.

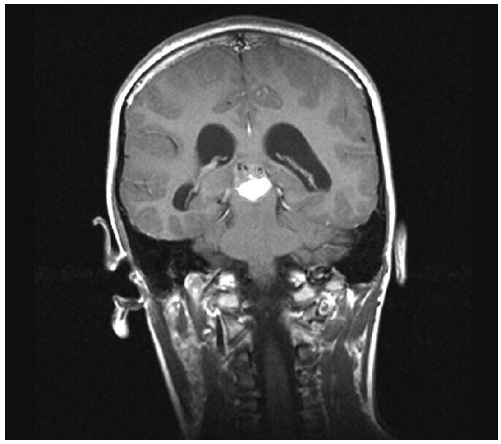
Discussion

Intracranial germ cell tumors are a heterogeneous group of lesions that occur in children and young adults. Within the classification of









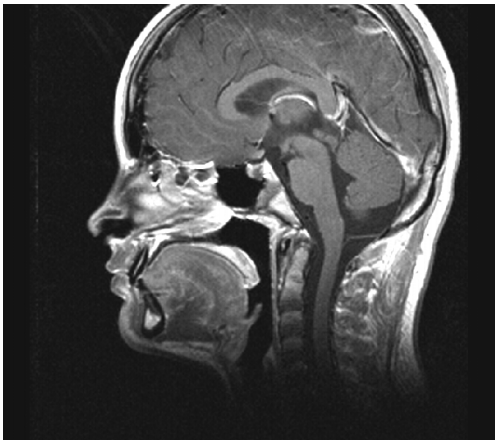
Figures 5,6. MRI (SE T1WI) in sagital plane before and after administration of gadolinium contrast media (Gd CM). Solid part of tumors enhance homogeneously after administration of Gd CM. Note compression of surrounding structures. No communication between lesions is seen.

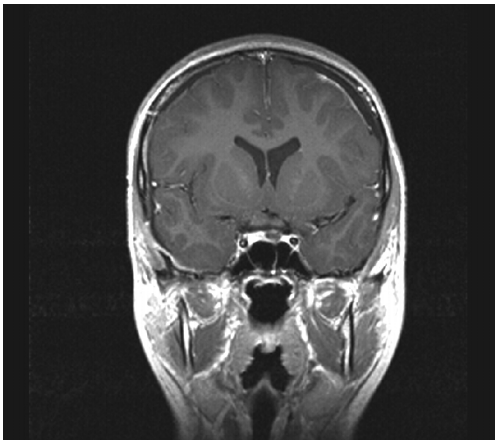
Figures 7,8. MRI (SE T1WI) in coronal plane after administration of Gd CM. Midline position of tumors with impression of third ventricular floor.

intracranial germ cell tumors, there are a variety of different tumor types which carry different prognoses. The most recent World Health Organization classification of germ cell tumors is as noted in Table 1. The most frequent histological type is germinoma (65%) and it holds the best prognosis[2] with over 90% 5 year survival rate.[3-5] They are located usually in the pineal or the suprasellar region.[6] Lesions in infundibulum alone are also described.[7] Rarely they appear in thalamus and basal ganglia or other intracranial loca-

Table 1. WHO classification of intracranial germ cell tumors

5.0	Germ cell tumors
5.1	Germinomas
5.2	Embryonal carcinoma
5.3	Yolk sac tumor
5.4	Choriocarcinoma
5.5	Teratoma
5.5.1	Immature
5.5.2	Mature
5.5.3	Teratoma with malignant transformation
5.6	Mixed germ cell





Figures 9,10. Ppostoperative control MRI (SE T1WI) in sagital and coronal plane after administration of Gd CM. There is no residual tumour.

tions. Pineal germinomas have strong male predominance in contrast to suprasellar germinomas that are more frequent in females. Bifocal lesions in these regions are found in 5-10%.[1] It is unclear whether they represent actual spread of the tumor or the simultaneous development of tumor in two sites.

Clinical presentations of germinoma are dependent on the localization of lesions. Tumors in pineal area can present with hydrocephalus, visual symptoms, obtundation, pyramidal tract signs and ataxia. Suprasellar tumors often produce diabetes insipidus and a pituitary hormonal dysfunction.

Germinomas are macroscopically solid, quite homogenous tumors with a possible soft or partly cystic central part. They can seed by cerebrospinal liquor to other part of brain or meningeal surface.[8,9] Germinomas are composed of more than one cellular type in 10%,[10] their unspecific histological tumor marker is placental alkaline phosphatase.[11]

The neuroimaging characteristics of germinomas and nongerminomatous germ cell tumors are similar enough to limit diagnostic certainty, and either tissue confirmation or measurement of specific tumor markers are needed for the diagnosis. In addition, germ cell tumors in the pineal region cannot be definitively separated on the basis of neuroimaging characteristics from other tumors such as pineoblastomas, pineocytomas or gliomas. In the suprasellar region germinomas may be difficult to separate from other lesions which infiltrate the surrounding brain mimicking gliomas and histiocytomas.

Germinomas are radiologicaly well delineated, oval or lobular and expansive or partly infiltrative tumors.[6] The proportion of water to tumor cells determines their radiological morphology. The tumor can be isodense to hyperdense on CT and isointense to hyperintense on T1WI and T2WI on MRI. The solid part of the tumor shows isointense signal to cortex and intense opacification after the application of gadolinium contrast media Gd CM[3] (Figures 1-8). Germinomas are quite homogenous and uncapsular with no calcinations and other inclusions or important cystic and/or hemorrhagic areas.[12]

The treatment for germ cell tumors has become somewhat divergent as recommendations for the treatment differ between pure germinomas and other forms of germ cell tumors.[13] The craniospinal radiation of germinomas together with local doses of 4000 cGy showed excellent results.

Although a definite histological diagnosis cannot be achieved by CT and/or MRI alone, the detailed analysis of neuroradiological im-

ages is useful for predicting the histological diagnosis.

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