Posterior fossa teratoma

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ABSTRACT

تسبب أورام الخلية الجرثومية ما يقارب من 2-5% من جميع أورام الدماغ في مرحلة الطفولة. تنشأ هذه الأورام غالباً في المنطقة الصنوبرية وفوق السرج ولكنها قد تحدث في جميع أنحاء الدماغ. تنقسم الأورام المسخية إلى الأورام التناسلية والأورام خارج الجهاز التناسلي. إن حدوث الأورام المسخية في الحفرة الخلفية هو أمر نادر الحصول. إن محور هذا التقرير هو حدوث ورم مسخي في طفل عمره 5 أعوام والذي تم علاجه بنجاح باستخدام الطرق الطبية والجراحية. نحن ايضاً نستعرض الصور التشخيصية والنسيجية.

Germ cell tumors comprise approximately 2-5% of all childhood brain tumors. They arise predominantly in the pineal and suprasellar region, but may occur throughout the brain. Teratomas are generally divided into gonadal and extragonadal types. A posterior fossa teratoma is a rare occurrence. The focus of this discussion is a 5-year-old boy with posterior fossa teratoma who recovered completely after medical and surgical intervention. We also present his interesting imaging and pathological findings.

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Teratomas are tumors that contain tissue representative of all 3 germ cell layers of the embryo (ectoderm, mesoderm, and endoderm). They represent approximately 2% of pediatric intracranial tumors. Although teratoma of the posterior fossa is a rare occurrence, we present the case of a 5-year-old boy with a midline posterior fossa teratoma. Our objectives in presenting this particular case are to include teratoma in the differential diagnosis of posterior fossa tumors in children, and to share interesting radiology, intraoperative, and pathology images.

Case Report. A 5-year-old boy presented a year ago to the local hospital with 3 month's history of headache, vomiting, and irritability. The course was progressive, and a month prior to seeking medical advice he became obtunded and stopped responding to questions and verbal commands. A CT scan was carried out in the local hospital and revealed a cystic posterior fossa lesion (**Figure 1**), and he was urgently referred to our hospital. He was a product of a full term pregnancy and uncomplicated vaginal delivery. His Apgar score, head circumference, weight, and fontanels at birth were all normal. His milestones were normal until the time of presentation. His medical, surgical, and family history were unremarkable and vaccinations were up-to-date.

His vital signs were normal on examination in our emergency department. He was obtunded but opened his eyes spontaneously and obeyed simple commands. His pupils were equal and reactive and extra ocular muscle and gaze examination revealed paralysis of upward gaze. The remaining cranial nerves, motor functions, and coordination were normal. An emergency extra-ventricular drain was inserted, dexamethasone therapy was started, and he was admitted to the pediatric intensive care unit. He developed a generalized tonic clonic seizure on the second postoperative day and was started

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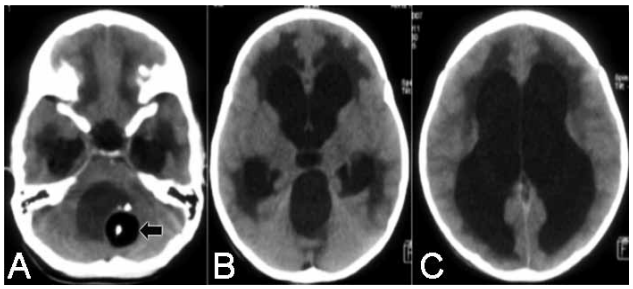

Figure 1 - Patient CT of head without contrast showing: **A, B, C)** massive communicating hydrocephalus, and **A)** a cystic posterior fossa lesion with a hyperdense round area (arrow) most likely representing a tooth.

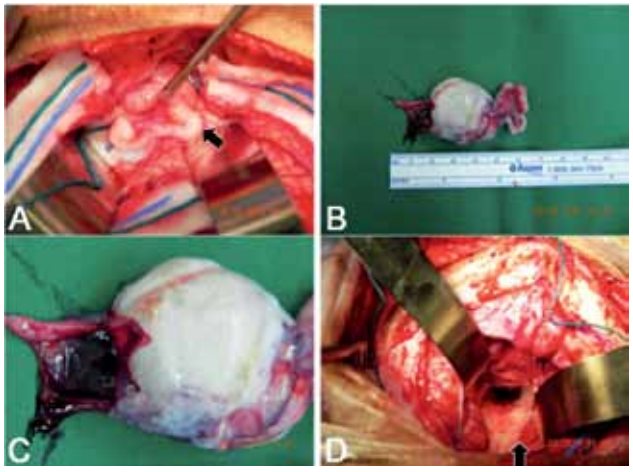

Figure 2 - Intraoperative photos showing: **A)** Operative view showing a large intestine as part of the tumor. **B & C)** The tumor measuring 6 cm containing hair, intestine, and omentum. **D)** Operative view at the end of surgery showing complete resection of the tumor.

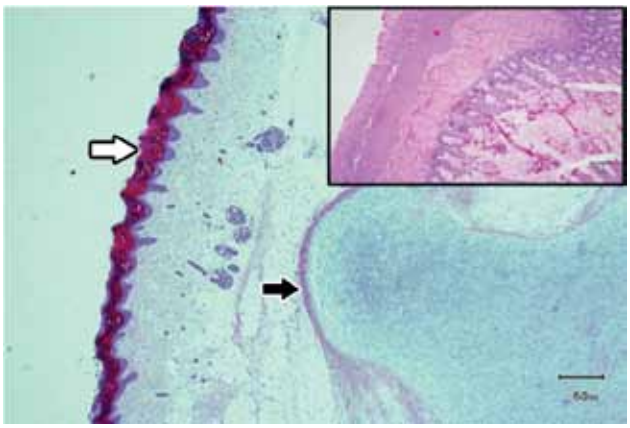

Figure 3 - The pathology shows different components of mature teratoma (Hematoxylin & Eosin X50). The white arrow shows keratinized squamous epithelium resembling normal skin and representing derivation of the embryonal ectoderm. The black arrow shows mature cartilage, representing the embryonal mesoderm. The inset shows a Hematoxylin & Eosin stained section (X200) of a different portion of the specimen containing mucosa and muscularis propria, similar to normal colon, and representing the embryonal endoderm.

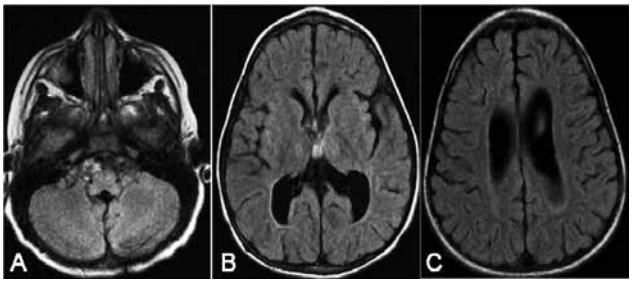

Figure 4 - Follow-up patient MRI one year after surgery showing no evidence of recurrence with resolution of hydrocephalus.

on lorazepam and phenytoin therapy. Three days later, a suboccipital craniotomy and resection of the posterior fossa tumor were carried out with no complications. Intraoperatively, the tumor was composed of a large loop of intestine with teeth and hairlike structures, and it was removed completely (**Figure 2**). The pathological diagnosis was posterior fossa mature teratoma (**Figure 3**). He required a ventriculoperitoneal shunt due to the hydrocephalus and high intracranial pressure on monitoring. This was carried out 8 days later with no complications. The postoperative course was uneventful and 6 weeks later, in the outpatient department, his parents reported no significant complaints and the neurological examination was normal. The parents expressed their satisfaction regarding his progress and management plan. One year later, an MRI of the brain showed significant improvement of the communicating hydrocephalus with no evidence of tumor recurrence (**Figure 4**).

Discussion. Although relatively rare, pediatric malignancies are considered the second most common cause of death in children less than 19 years of age (after trauma). They constitute up to 20% of all malignancies in this age group. Clinical presentation depends on several factors, including age of the child, growth rate of the tumor, and its localization in the CNS. Although the etiology of most childhood CNS tumors is unknown, 2% of these tumors are caused by or associated with a hereditary syndrome. Some of these associations are extremely strong, for example type-1 neurofibromatosis has a higher incidence of low-grade glioma and other

types of tumors. Reaching the diagnosis of a childhood CNS malignancy is often challenging, especially in the early stages of disease. Studies that evaluated the delay in diagnosis of childhood cancer demonstrated that the mean time of delay for diagnosis of brain tumors was as long as 29.3 weeks.[1] Sufferers usually present with morning headaches, nausea, and vomiting. However, in infants the presentation is atypical, with paresis of upward gaze due to tectal pressure resulting in downward deviation of the eyes, sometimes referred to as, "the setting-sun sign."

Germ cell tumors (GCTs) commonly occur in children and adolescents with males being affected more often than females. The incidence of intracranial GCTs varies significantly according to geography. In Western countries, they account for 0.4-3.4% of all pediatric CNS tumors, while series from Japan and other Asian countries have reported that these growths account for up to 11% of all pediatric brain tumors.[2] **Table 1** illustrates the general classification of GCTs. More than 90% of the affected patients are younger than 25-years-old, and most cases occur in the second decade, with their peak coinciding with the onset of puberty.[2] Although they may arise throughout the neuroaxis, such tumors are preferentially situated in the midline region, and in or around the pineal gland, third ventricular region, and suprasellar region. The GCTs are thought to arise from misplaced germ cell nests during embryogenesis. Arrested migration is presumed to account for the extragonadal locations in the normal path of the primordial germ cells (retro-peritoneum), whereas aberrant migration results in cells at other extragonadal sites (ie, pineal and sacrococcygeal).

The presenting clinical features of teratoma are related to the size and the localization of the lesion. Localization in the pineal region leads to the lesions compressing and obstructing the cerebral aqueduct and CSF flow, resulting in progressive non-communicating hydrocephalus and intracranial hypertension. In addition, pineal region lesions make the tectal plate vulnerable, which when involved produce a characteristic paralysis of upward gaze and convergence known as Parinaud's syndrome.[3] In patients with teratoma, assay of serum and CSF for α-fetoprotein (AFP; normally synthesized by yolk sac endoderm, fetal hepatocytes, and intestinal epithelium) and beta human chorionic gonadotropin (β-HCG; normally secreted by syncytiotrophoblast) is now routine in the presurgical assessment of suspected GCTs. Elevations of either oncoprotein constitute compelling evidence of germ cell neoplasia, the pattern of marker elevation being somewhat predictive of tumor histology. The diagnosis of teratoma in our case was suspected on imaging, and thus the diagnostic yield of serum CSF was expected to be low. Intracranial teratomas present with symptoms of space-occupying lesions.

Table 1 - World Health Organization classification of intracranial germ cell tumors.[2]

Germinomas
Nongerminomatous germ cell tumors
Embryonal carcinoma
Yolk sac tumor (endodermal sinus tumor)
Choriocarcinoma
Teratoma (benign teratoma, immature teratoma, teratoma with "malignant transformation")
Mixed germ cell tumors

The neuroradiological features of CNS GCTs are generally non-specific. However, on CT or MRI a diagnosis of teratoma should be suspected when a lesion demonstrates intratumoral cysts admixed with calcified regions, heterogeneous density, and foci of low-attenuation similar to fat.[3] Mature teratomas are composed exclusively of highly differentiated, variably organized "adult-type" tissue elements, which represent the 3 germ cell layers of the embryo, with low or absent mitotic activity. The more common ectodermal components present in such tumors includes skin, brain, and choroid plexus. Mesodermal tissue types include cartilage, muscle, bone, and fat. Cysts lined by epithelium of respiratory or gastrointestinal type are the usual endodermal representatives, with some also containing pancreatic or hepatic tissue. The phenomenon of "fetus-in-fetu" results from advanced organogenesis and somatic organization through incorporation of a dizygotic twin via epithelial or neural tube defects. This disrupts the amniotic septum and has also been suggested to account for some cases of this pathologic description.

Approximately one-half of all childhood brain tumors arise in the posterior fossa.[4] The 5 most common tumor types that arise sub-tentorially are: medulloblastoma, cerebellar pilocytic astrocytoma, brain stem glioma, ependymoma, and atypical teratoid/rhabdoid tumor. The first report of a teratoma located in the midline posterior fossa was in 1912.[5] Since then, 22 cases of midline posterior fossa teratomas have been reported,[6] in which females predominate, in contrast to male predominance in pineal and suprasellar locations. Fourteen cases were mature teratomas and 7 cases were immature, with one case reported as neither mature nor immature.[6]

The prognosis of CNS GCTs depends on histological type with 5-year survival rates of mature teratomas as high as 93%. Mature teratomas are thought to be radioresistant, and therefore the treatment of choice is total surgical resection.[7] Immature teratomas carry somewhat poorer prognosis and require total resection followed by local brain field radiation.[8] They advocate only aggressive surgical resection for low-grade immature teratomas, and reserve adjuvant chemotherapy and radiotherapy for the high-grade type.[9] Teratomas with malignant components need craniospinal radiation therapy to prevent spinal metastases.[8] Immature teratomas and those with secondary malignant components have been shown to have 75% 5-year survival rates.[10] The prognosis of intracranial congenital teratomas is dismal with mortality rates of >90%. Death usually occurs before or shortly after birth due to massive replacement of brain tissue by the tumor.

In conclusion, we report the case of a 5-year-old child with deterioration of consciousness and cerebellar mass. The radiological features are consistent with mature teratoma, and the final histopathological evaluation confirmed this diagnosis. Although intracranial GCTs are more frequent in the pediatric population, the occurrence of a mature teratoma in the cerebellum is a distinctly rare event. Our case is unique with regard to the patient's progressive neurological deterioration over a relatively short period of time, the size of the lesion, and the presence of the small bowel component.

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