Embryonal carcinoma in the cerebellum

Case report

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A cerebellar embryonal carcinoma in a 5-year-old child is reported. The tumor was well encapsulated and completely removed. Postoperative radiation was given. The patient is alive and healthy.

KEY WORDS - embryonal carcinoma 
ectopic pinealoma

It is generally accepted that the “pinealoma” that occurs in the pineal regions and the “ectopic pinealoma” occurring outside the pineal gland belong to the group of atypical teratomas or germinomas. The two tumors display a variety of elements representative of germinal layers, such as cartilage, muscle, tubules of mucous-secreting epithelium, and sheets of neoplastic cells resembling seminoma of the testis or dysgerminoma of the ovary. There is yet another group of neoplasms that supposedly arise from the pineal parenchymal cells, the so-called pineocytoma and pineoblastoma. Recently Borit reported a case similar in appearance to embryonal carcinoma of the testis and proposed that the term “embryonal carcinoma” be used for this type of germinal neoplasm. His classification was similar to that of Dixon and Moore. The ectopic pinealomas reported have usually been in the tuber cinereum, third ventricle, infundibular region, optic chiasma, and suprasella and intrasellar regions.

We are reporting an example of an ectopic pinealoma in the cerebellum that resembles embryonal carcinoma of the gonads.

Case Report

A 5-year-old Thai boy was admitted to Ramathibodi Hospital complaining of increasing headache and vomiting for about 1 month.

Examination. There was hypotonicity of all the extremities; no other cerebellar signs were found. There was no papilledema. Skull films showed separation of the sutures and ballooning of the posterior fossa. Right brachial angiography showed a left cerebellar mass displacing the fourth ventricle to the right. A ventriculogram showed large ventricles and complete obstruction of the fourth ventricle due to a mass in the left cerebellar hemisphere. The suprapineal recess was enlarged, but showed no filling defect.

Operation. Posterior fossa exploration

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was done on July 13, 1972. There was considerable tonsillar herniation, greater on the left than the right. The left cerebellar hemisphere was tapped, and at a depth of 1 cm, a solid tumor mass was found. The tumor was well encapsulated, situated entirely in the cerebellar hemisphere, and had no immediate relationship with the fourth ventricle or pineal region. It was totally removed.

Postoperative Course. Recovery was uneventful, and the patient was discharged on August 4, 1972. He was given a course of radiation of 4000 rads within 31 days. Follow-up brain scans were done on three occasions, the last on May 29, 1973; all showed no recurrence. The boy is healthy and shows no neurological abnormalities.

Histological Examination. The surgical
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The specimen consisted of an oval, encapsulated, grayish brown, firm mass, measuring 4 x 3 x 2 cm. The cut surface was whitish brown, mottled with yellow (Fig. 1). Microscopically, the tumor was surrounded by thick fibrous tissue (Fig. 2 left). The neoplasm itself showed two main features at low magnification, namely, solid areas and areas of cystic formation, interspersed by broad bands of fibrovascular stroma. In the solid areas, the tumor cells were loosely arranged, without definite pattern. The cytoplasmic borders of the individual cells were indistinguishable. The nuclei were large, irregular, and hyperchromatic, with prominent basophilic nucleoli. The cytoplasm was abundant, containing fine acidophilic and amphophilic granules, and sometimes vacuolated (Fig. 2 right). Mitotic figures were frequent.

In areas of cystic formation, the tumor cells were arranged in tubular or glandular patterns, which varied in size and shape. The tumor cells lining these tubules or glands were cuboidal to columnar, with nuclei similar to those found in the solid area, but with more vacuolation in the cytoplasm (Fig. 3 left). Occasionally, structures suggestive of “embryoid bodies” were found (Fig. 3 right). In some areas trophoblastic differentiation and even structures resembling placental villi were found (Fig. 4).

In the interspersed fibrous stroma, there were groups of foamy histiocytes and occasional smooth muscle cells (Fig. 5). Hemorrhagic areas were also present. Mucicarmine stain showed the occasional presence of mucin within the glandular lumina, as well as in the cytoplasm. The periodic
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Fig. 4. Photomicrograph of embryonal carcinoma showing trophoblastic differentiation and structures resembling placental villi. H & E, × 320.

acid-Schiff reaction demonstrated large numbers of PAS-positive granules, intracellularly and extracellularly. These PAS-positive granules disappeared almost completely after digestion with diastase. A moderate number of reticulin fibers were present within the dense stroma and around the blood vessels. The phosphotungstic-acid-hematoxylin stain failed to demonstrate any neurological fibrils.

The microscopic picture of the neoplasm was that of an embryonal carcinoma associated with epithelial differentiation.

Discussion

Tumors with this appearance have been described and documented by various authors. According to Russell and Rubinstein, carcinomas of the choroid plexus usually occur in the young age group, so extension from a papillary carcinoma of the choroid plexus in the fourth ventricle had to be considered in this case, because of the presence of glandular structures in the tumor. However, as mentioned previously, the tumor was situated entirely in the cerebellar hemisphere, had no immediate relationship with the fourth ventricle, and the choroid plexus appeared normal at surgery. Papillary carcinoma of the choroid plexus was therefore excluded.

Since the testicles of this boy were normal and the chest films showed no abnormalities, metastasis from either of these two areas was unlikely. Extension or metastasis from the pineal gland or pineal region is a possibility in spite of the negative skull film and brain scan. It has been stated that pinealoma can be diagnosed radiographically by the characteristic type of calcification. Our own experience is entirely in agreement with this fact, particularly for Thai children. That a small lesion in the pineal gland may be associated with a large extended lesion has been reported but was excluded in this case by ventriculography.

The presence of a primary germ cell tumor in the cerebellum can be explained from the embryological standpoint in that the germ cells migrate through certain tissues before arriving at the gonadal region. Rorke, et al., in their study of cerebellar heterotopia in infancy, commented that cell rests in the cerebellum may result whenever the following abnormalities occur in the development of the cerebellum: 1) fusion of several cell layers; 2) arrest in the migration of germinal elements from the germinal epithelium lining the cavity of the fourth ventricle; and 3) an alteration or derangement of the normal process of differentiation of germinal cells. Thus, a logical basis for the existence of an extragenital germinal tumor in the cerebellum is suggested.

Embryonal carcinoma of the central nervous system was first reported by Nishiyama, et al., and subsequently Borit reported another case with full microscopic description. To the best of our knowledge, the present case is the third of embryonal carcinoma of the central nervous system and represents the first instance in which the tumor occurred in the cerebellum.
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Fig. 5. Left: Photomicrograph showing areas of tubular or glandular structures interspersed by a dense fibrous stroma. There are groups of foamy histiocytes. H & E, × 80. Right: Occasional smooth muscle cells are seen in the stroma. Masson, × 320.

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References


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