Malignant melanoma of the choroid plexus epithelium

Case report

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A patient with a malignant melanoma of the choroid plexus epithelium is described. The embryological implications of the epithelial rather than stromal location are emphasized. This case appears to be unique in the literature.

KEY WORDS: choroid plexus · epithelium · melanoma · brain tumor

A wide variety of tumors arising from the vascular core or stroma of the choroid plexus has been described. These include meningioma, sarcoma, angioma, chondroma, lipoma, teratoma, simple cyst, and xanthoma. However, tumors arising from the choroid plexus epithelium are decidedly rare, and have included papilloma, carcinoma, and recently, acinar adenoma. The present report describes a patient with a malignant melanoma arising from the epithelium of the choroid plexus in the lateral ventricle.

Case Report

One month before admission this 8-year-old boy developed headaches, lethargy, and poor concentration.

Examination. The patient was alert and cooperative. General physical examination was unremarkable; there was no evidence of pigmented lesions on the skin or in the fundi. Nystagmus was present at extremes of gaze laterally and vertically. Low-grade papilledema was present but there were no localizing signs. Skull films were normal. An electroencephalogram demonstrated a slow wave focus in the left temporal area while technetium 99m brain scan confirmed the presence of a lesion deep in the left temporoparietal area. A ventriculogram on September 3, 1968, showed the intraventricular location of the mass.

Operation. Transcortical removal of the tumor was performed on September 3. The tumor appeared to originate from the choroid plexus of the atrium; macroscopically it was not invading the cerebral substance around the ventricle. Removal of the tumor was thought to have been complete.

Postoperative Course. The patient progressed well and demonstrated only a right homonymous hemianopsia. He was given irradiation therapy with cobalt-60 over a 40-day period and received a total dose of 5459 R to the midplane of the tumor.

Pathological Examination. The tumor measured 3.8 × 2.5 × 2.8 cm. On sectioning, the center was hemorrhagic and was surrounded by small cysts. No pigment was observed grossly. The microscopic appearance consisted of rather normal-appearing choroid plexus adjacent to areas where the choroid plexus epithelium had proliferated.
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Some papillary-like structures were preserved in the ill-defined cellular areas of tumor. Stromal proliferation appeared minimal. There were occasional true rosettes and pseudo-rosettes, and a few areas of necrosis surrounded by pseudo-palisading cells. Pleomorphism was not prominent although a few mitotic figures were seen. The most striking finding was the presence, after staining with hematoxylin and eosin, of brown pigmented granules lying within the tumor cells (Fig. 1). Some pigment appeared to be lying in the stroma and within phagocytes. The pigment was not seen on potassium permanganate bleached sections.

Second Examination. One month before readmission on March 23, 1969, the patient developed bilateral foot drop. On examination Babinski signs were present bilaterally and abdominal reflexes were absent. There was spotty hypesthesia to pinprick over the toes of both feet and a sensory change at T-10. A myelogram showed irregular defects in the thecal sac and multiple defects on the right from T-3 to T-6. The Queckenstedt maneuver was normal; the spinal fluid protein was 68 mg%.

Second Operation. On March 25, 1969, a decompressive laminectomy of T-2 to T-8 was performed. The spinal cord appeared peppered with pigmented tumor implants.

Second Postoperative Course. The patient was given another course of radiation over 4 weeks, this time 3500 R to the entire spinal axis. His course was steadily downhill, characterized by hoarseness, difficulty in swallowing, and incontinence. He died on October 12, 1969.

Postmortem Examination. The general examination was unremarkable except for bronchopneumonia. No obvious primary melanoma was found. Unfortunately, the parents did not permit examination of the eyes.

Grossly the surface of the brain, spinal cord, and cauda equina were studded with black melanoma metastases (Fig. 2 left). Sections of the brain showed multiple metastases lining the ventricles and lying within the cerebral substance (Fig. 2 right). There also was tumor in the region of the original surgery but it appeared to lie within the cerebral parenchyma. It was impossible to deter-
Robert A. Beatty

Fig. 3. Photomicrograph of the cerebral metastasis adjacent to the normal brain in upper right. Papillary structures are reproduced; melanin pigment is located intracellularly. H & E, × 155.

mine whether this was a metastatic implant or regrowth and invasion by the primary tumor. Microscopic sections of the spinal cord and cerebral metastases again confirmed the melanin-containing epithelial cells in papillary-like arrangements (Fig. 3).

Discussion

The epithelial location of the pigment was unexpected in view of the embryology of the choroid plexus. This structure is formed by the pia arachnoid invaginating into the lateral ventricle and pushing a layer of ependyma ahead of it. The epithelium of the choroid plexus is thus modified ependyma while the stroma is connective tissue and pia-arachnoid remnants. Meningiomas of the choroid plexus have been explained embryologically by the proliferation of these remnants.

Another tumor that originates from the meninges is primary meningeal melanosis. This tumor apparently is a malignant transformation of the melanocytes, of neural crest origin, which normally lie in the meninges of the ventral medulla, spinal cord, and cerebellum. Although some cases of intracerebral or intraspinal cord melanoma have been attributed to melanocytes lying in the pia within the Virchow-Robin spaces, this mechanism would seem not to apply here.

On embryological grounds, therefore, one would not be surprised to find pigment in the stroma of the choroid plexus. Our patient, however, had pigment predominantly in the epithelium. Melanin apparently has not been described in this location, but Bargmann has described intra-cytoplasmic pigment granules that stained with Turnbull blue.

The possibility that this tumor represents a metastasis should be considered. Although the eyes were not examined at autopsy, funduscopic examination was repeatedly normal. Moreover, it seems unlikely that a metastasis would appear first in the choroid plexus. In fact, in children, metastatic carcinoma to the choroid plexus has not been reported.

Our case seems unique in the medical literature. However, Fowler and Simpson described a 2-year-old boy with a malignant melanoma arising from the roof of the fourth ventricle. There were many metastatic implants along the spinal cord and cauda equina, some of which had a papillary appearance. The pigment was most prominent in the epithelial cells. They raised the possibility of choroid plexus origin but felt that the tumor probably was a variety of medulloblastoma because of the age of the patient and the midline location. Their conclusion that the tumor was a combination of the normal capability of neural blastoma to produce melanin pigment and papillary structures does not apply to our patient in whom the tumor clearly arose from the choroid plexus. However, it is conceivable that their tumor also developed from the choroid plexus.

References

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