Cerebellar Medulloblastoma With Postoperative Extracranial Spread
Report of a Case

SHIZU MIYAKE, M.D., MITSUO TOYAMA, M.D., BIN ETANI, M.D., AND SEISHI FUKUMA, M.D.
Department of Pathology, Central Clinical Laboratory and 1st Department of Surgery,
Kyoto Prefectural Medical College, Kyoto, Japan

Until recently there has been skepticism concerning extracranial metastases of intracranial neoplasms, whereas their spread throughout the central nervous system is well known. Case reports of extracranial dissemination, however, are rare and many of them have been questionable because of incomplete description of pathologic findings. The following case has been studied recently in our laboratory and substantiates the finding of distant metastases of a primary cerebellar medulloblastoma.

Case Report

History. A 12-year-old, right-handed boy was referred to the hospital on April 30, 1962. He was well until the beginning of March 1962, when he experienced difficulty in writing and handling chopsticks. In April he started to have headaches, nausea and vomiting, accompanied by weakness of the right leg. He complained of excreting pain behind the eyes and in the occipital region.

Examination. The patient was alert, cooperative and well oriented but was markedly dehydrated. He showed signs of a right cerebellar lesion. His speech was somewhat slow and he staggered slightly to the right side. Romberg's sign was slightly positive with falling toward the right side. Intracranial pressure was moderately elevated. There was no papilloedema. On May 4, 1962 angiography, including vertebral counterpart, showed nothing remarkable except for slight hydrocephalus.

Operation. On May 8, 1962 (231 days before his death) he was subjected to suboccipital craniectomy following pneumoventriculography. There was evidence of tumor of the cerebellar hemispheres and pontine angle. On the 206th day before death, Torkildsen's ventriculocisternal shunt was performed because a contrast ventriculogram disclosed obstruction of the aqueduct of Sylvius. Bilateral pontine syndrome, predominantly on the right side, developed gradually and he became comatose. From Oct. 5, 1962 (101 days before death) he remained in a coma. Tumorous spread was observed at the following locations: On the 81st day before death, right exophthalmus. On the 67th day before death, multiple tumors on frontal to parietal convexity of scalp. On the 82nd day before death, symmetrical swellings in all the junctions of the ribs. The patient died on Jan 18, 1963. Complete autopsy was performed 8 hrs. later.

Pathological Data. Gross. The outer surface of the skull was occupied by numerous nodules (Fig. 1) which were greyish-pink in color, rubbery in consistency, and varied in size. The largest one was located adjacent to the defect in the suboccipital bone, covered by soft tissues that were firmly adherent to the leptomeninges over the inferior vermis and underlying cerebellar surface. The dura mater of the convexity was smooth, except for a few scattered epidural nodular tumors. Externally, the brain was swollen and congested, and weighed 1,900 gm. after fixation. Leptomeninges of the convexity was not thickened but the basal part was edematous and cloudy. Coronal section of the brain after the method of Fischer revealed dilatation of both lateral and 3rd ventricles. The ependymal surface was almost smooth. There was no tumor found in either cerebral hemisphere. The 4th coronal section showed the first evidence of tumor at the level of the inferior colliculus, where the cerebellum was distorted. Most of the cerebellum was replaced by the tumor which was semi-transparent, friable, and moderately demarcated from cerebellar tissue. Both denticulate nuclei and white matter were obscured (Fig. 2).

No superficial cervical, axillary or inguinal lymph nodes were enlarged, but paratracheal and deep inguinal nodes were swollen by metastases. Tumor nodules of various sizes were found extensively along the vertebral column (Fig. 3). These were suggestive of the tumor arising from both sympathetic trunks and were diagnosed as neuroblastoma. But both adrenals were normal and the pachymeninx was free from metastases except for microscopical deposits in the cervical posterior spinal root. Deposition of tumor along the vertebral column was located rather more laterally. These find-
ings were enough to exclude the peripheral origin of the tumor. Furthermore, the tumor involved both clavicles, sternum, right humerus including bone marrow, both orbital cavities and parenchyma of liver.

Microscopic. The specimens from most parts of the cerebellum revealed compact nuclei, which were generally uniform and arranged in irregular patterns. There was some formation of rosettes (Fig. 4). The tumor had undergone degeneration (calcification), scattered in many places. The tumor cells had a tendency to concentrate around the small vessels without proliferation of endothelium. Cytoplasm of the neoplastic cells was scanty and neither neuroglial nor reticular fibrils were demonstrated. In metastatic areas, for instance in skull or lymph nodes, there were abundantly proliferated fibrous tissues, by which tumor cells were arranged singly or in groups.

Comment

It has been believed for many years that intracranial tumors, gliomas in particular, are virtually incapable of remote metastases. Willis\(^8\) said that strong precautions should control the interpretation of possible cases because of the danger of confusing them with examples in which a small and latent primary carcinoma is overlooked. A review of the literature revealed a few cases of cerebellar medulloblastoma with extracranial spread.\(^5\)\(^-\)\(^7\)\(^,\)\(^10\)\(^,\)\(^11\)

In order to be recognized as a verified case as outlined by Weiss,\(^8\) it is necessary that the characteristic tumor be found histologically in the central nervous system, the initial symptoms prove to be the result of the intrinsic tumor, and a complete autopsy reveal no other neoplasm. Only the cases of Nelson,\(^3\) Rubinstein,\(^4\) Paterson,\(^5\) and Oberman \textit{et al.},\(^1\) that satisfy these criteria, are well documented examples.

Summary

Complete autopsy of a 12-year-old boy with a medulloblastoma and postoperative extracranial spread has been described. The initial symptoms had begun as a result of a cerebellar tumor. At autopsy, the dissemination of tumor was found extensively on the outer surface of skull, in paratracheal and inguinal lymph nodes, both clavicles, sternum, ribs, right humerus, parenchyma of liver, and along the spinal column. Previous craniectomy would have paved the way of dissemination from suboccipital decompression to remote various places.

References


\begin{figure}[h]
\centering
\includegraphics[width=0.5\textwidth]{medulloblastoma_with_extracranial_spread.png}
\caption{Primary medulloblastoma occupying the cerebellum in 6th section (Fischer).}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=0.5\textwidth]{rosette_like_structure.png}
\caption{Rosette-like structure of primary lesion. (Hematoxylin and eosin, X800)}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=0.5\textwidth]{metastatic_tumor.png}
\caption{Metastatic tumor along the spinal column.}
\end{figure}