Intracranial chondrosarcoma with extracranial metastases

Case report

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This report describes a case of primary intracranial meningeal chondrosarcoma arising from the falx with extracranial metastases to the heart, liver, pancreas, kidneys, and adrenals.

KEY WORDS: intracranial chondrosarcoma · falx · extracranial metastases

Intracranial meningeal chondromas are rare. There have been a few instances of malignant intracranial chondroma of meningeal origin recorded, but none with extracranial metastases.

Case Report

A 51-year-old woman had had weakness of both legs for 2 months.

Examination. The patient was alert, well oriented, and cooperative. General examination was normal. There was spastic paraparesis, more marked on the left, with bilateral hyperreflexia of the legs and bilateral extensor plantar responses. Papilledema was noted bilaterally. Skull films showed a destructive lesion in the right parietal region. Right carotid angiograms demonstrated a tumor stain in the right frontoparietal region corresponding with the bone destruction. The chest film was normal, as were laboratory examinations.

Operation. The morning after admission the patient was found semicomatose. Immediate craniotomy was performed. A large, chiefly right and partly left, frontoparietal tumor was practically totally removed. Although not encapsulated, the tumor was entirely solid and well demarcated from the surrounding brain. The upper portion of the tumor adhered firmly to the overlying dura which it penetrated to erode the adjacent skull. The tumor had completely surrounded and invaded the falx, and the middle third of both superior and inferior sagittal sinuses.

Postoperative Course. Recovery was very slow; there was residual paraparesis and mental dullness. Chest and skeletal films revealed no pathological changes. Approximately 4900 R of cobalt-60 were given, but 2 months after operation both arms became paretic, and angiograms showed recurrence of the tumor. Reoperation was recommended but the patient refused. One month later a few nodules were biopsied in the craniotomy site; the nodules continued to grow and increase in number. Frequent chest films and electrocardiograms revealed no abnormality. As the nodules of the scalp reached a grotesque shape and a huge size, the patient's general condition deteriorated. Jaundice developed about 10 months after craniotomy.
Intracranial chondrosarcoma with extracranial metastases and 12 months after the onset of symptoms. The patient died 1 month later.

Postmortem Examination. A complete autopsy was performed but only pertinent findings will be described. The external examination of the head showed that there were many sessile nodules of varying size, both inside and outside, and along the line of the craniotomy scar. Coronal section of the brain including the skull and scalp (Fig. 1 upper) revealed a large, relatively demarcated but not encapsulated recurrent tumor lying between the two cerebral hemispheres, adherent to the remaining falx. The tumor was firm but contained many areas of necrosis, softening, and small hemorrhagic foci. The tumor had invaded the dura, destroying the overlying skull, and continued into the scalp. In the subdural space near the main tumor there were several small nodules. A subdural nodule was found in the left anterior fossa and another in the posterior fossa. The other parts of the brain and entire spinal cord contained no tumor.

The heart contained a whitish-gray nodule of firm consistency, measuring 2 × 2 × 1.5 cm, in the right atrium near the atrial septum (Fig. 1 lower). There were no enlarged lymph nodes and no nodules in the lungs. The liver contained five nodules, the pancreas two, the kidneys one, and the adrenal one. Every nodule was pale whitish-gray in color, firm in consistency, and sharply demarcated from the surrounding tissue.

Histological Findings. The tumor from the surgical specimen contained many cartilaginous areas (Fig. 2 left). The tumor cells in the most cellular portion were polymorphous nuclei of various shapes and sizes (Fig. 2 right). Mitotic figures were scattered, and no giant cells were found. The neighboring dura and the superior and inferior sagittal sinuses were invaded by the tumor. Silver impregnation revealed an abundant reticulin network between the tumor cells. Some fibers coiled around the tumor cells. Neither calcification nor new bone formation was found. Transition from the cellular fibromatous tissue to cartilage was indistinct. The biopsy specimen was morphologically the same as the surgical one. Postmortem histological examination of the tumors from the brain, subdural space, scalp, heart, liver, pancreas (Fig. 3 left), kidneys (Fig. 3 right), and adrenals revealed the same type of tumor, which was diagnosed as chondrosarcoma.

Discussion
Most of the meningeal chondromas reported have been attached to the dura of the convexity or the superior sagittal sinus, the falx, or the choroid plexus. Histologically, these tumors contained fibromatous as well as cartilaginous components. Lichtenstein and Bernstein distinguished "mesenchymal chondrosarcoma" from other chondroid tumors. Since then, intracranial solitary mesenchymal chondrosarcomas have been reported sporadically. Whether our case belongs to this category is not deter-

Fig. 1. Upper: Coronal section of the brain including the skull and scalp showing extensive tumor invasion. Lower: Metastatic tumor in the heart (arrow).
Fig. 2. Left: Photomicrograph of the chondrosarcoma removed at operation showing cartilaginous portion. H & E, ×100. Right: Photomicrograph of the tumor showing noncartilagenous, highly cellular portion. H & E, ×100.

Fig. 3. Left: Photomicrograph of metastatic tumor in the pancreas. H & E, ×100. Right: Photomicrograph of metastatic tumor in the kidney. H & E, ×100.
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mined. In several instances, the meningeal cartilaginous tumors have been regarded as malignant\(^1,2,5,12\) but no documented extracranial metastases have been reported.

Meningeal tumors frequently invade the dura and the dural sinuses with or without histological malignancy, but reports of extracranial metastasis have been very rare.\(^6\) There have been about 40 meningeal tumors reported, both malignant and benign, with extracranial metastases in which no cartilaginous tumor has been found. Many followed craniotomy or repeated craniotomies. However, unoperated malignant tumors and even unoperated benign ones developed extracranial metastases. Most of the extracranial metastases were found for the first time at autopsy. Some metastatic tumors were found by x-ray examination and biopsies. The most frequent location of the extracranial metastases have been the lungs, pleura, liver, and lymph nodes. Heart metastases have been very rare.\(^7,14,15\) Metastases to the thyroid and breast have been reported.\(^14\)

Many of the cases of extracranial metastasis reported had certain features in common: 1) at least one previous operation (often multiple); 2) direct invasion of the dura, bone, or scalp at or near the site of operation; 3) relatively long survival; and 4) radiation therapy. Operation may favor lympho- genic or blood-borne spread of tumor by bringing intracranial tumor in contact with extracranial channels. The extracranial metastasis of meningeal tumors is rare compared to the common occurrence of venous invasion. Terminal emaciation may also be a factor. The significance of immunological factors in the development of extracranial metastases from a meningeal tumor is unknown.

According to Potter, et al.,\(^11,12\) the following conditions are necessary for neoplastic cells to gain access to the bloodstream: decrease of cohesiveness of tumor cells, increase of cell motility, and capability of transendothelial migration. Transendothelial migration of meningeal tumor cells or increase in cell motility have not been observed, and plication or interdigitation between cells has been observed in meningotheliomatous meningiomas. In human malignant gliomas, Kung, et al.,\(^9\) observed that the tumor cells were present in the vascular basement membrane, between the endothelial cells and even inside the lumen of blood vessels. They conclude that, although the presence of glioma cells in the bloodstream is probably a common phenomenon, implantation and viability of the dislodged tumor cells in a new site is most unlikely, possibly due to a metabolically hostile environment or immune rejection by the prospective host tissue. On the other hand, Tani and Ametani\(^16\) observed that a glioblastoma cell penetrates capillary walls through the dilated junction between the endothelial cells and that, within the lumen of the capillary, the tumor cell forms a new junction with the endothelial lining. They consider that the presence of glioma cells free in the bloodstream is uncommon because of formation of this new junction between tumor cells and the endothelial lining, and that this phenomenon may be one of the reasons why extracranial metastases are so rare. Tumor cells have not been observed in the capillary lumen of meningeal tumors.

We believe that this is the first case reported of primary intracranial meningeal chondrosarcoma with extracranial metastases.

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