Primary reticulum-cell sarcoma (microglioma) of the brain with massive cardiac metastasis

Case report

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A patient is presented with reticulum-cell sarcoma of the brain and a large metastasis to the heart.

**KEY WORDS** - reticulum-cell sarcoma - microglioma - metastatic tumor

Tumors of the lymphoreticular system may involve the central nervous system (CNS) as primary tumors, or may arise elsewhere and secondarily reach the nervous system. Primary lymphoreticular tumors of the CNS are the more common variety, and are presently the subject of much interest. We report a case of reticulum-cell sarcoma arising in the brain and having a large metastasis to the heart and microscopic deposits of tumor in the lung. No previous case report of reticulum-cell sarcoma of the brain with visceral metastases of this kind has been found.

**Case Report**

This 56-year-old woman was admitted for evaluation of aphasia and right hemiparesis. She had been well until 5 months earlier, when she began experiencing gait disturbance with loss of balance and falling to the right side. One month previously, she had an episode of urinary incontinence, and 2 weeks later developed slurred speech. In the last few days before admission, she had noted leg weakness, more marked on the right.

**Examination.** The patient manifested aphasia and paraparesis, more marked on the right. Computed tomography demonstrated areas of increased density in the thalamus bilaterally, which were considered to be glioblastoma multiforme. Chest x-ray films showed a mass in the right hilum and prominence of the right atrium and ventricle of the heart. She was started on radiation therapy to the brain and received 2600 rads. During her therapy she developed staphylococcal pneumonia and died.

**Postmortem Examination.** There was severe staphylococcal pneumonia with microabscesses bilaterally. The heart weighed 560 gm, and contained a lobulated soft tumor mass in the right atrium and ventricle. The tumor mass measured 2 × 3 cm, and had a gray gelatinous consistency. Tumor nodules 2 to 3 mm in diameter were scattered over the endothelial surface of several papillary muscles of the right ventricle.

The brain contained necrotic pale yellow-brown tumor throughout both basal ganglia, from the level of the anterior limb of the internal capsule posteriorly into the thalamus and inferiorly into the midbrain (Fig. 1). The lateral ventricles were mildly dilated.

Microscopically, the tumor in the heart was a reticulum-cell sarcoma and was quite cellular, with dense aggregates of tumor cells infiltrating the myocardial fibers. The tumor cells were moderately pleomorphic, varying from cells that resembled lymphocytes, with deeply chromatic nuclei and scant cytoplasm, to larger histiocytic-appearing cells, with vesicular nuclei, prominent nuclear membranes, distinct nucleoli, and more abundant cytoplasm. Mitotic figures were frequent in the latter cells (Fig. 2). The arrangement of tumor cells around blood vessels was very prominent, and reticulin fibers were abundant in a ring-like pattern around the vessels.

Foci of tumor cells identical to those in the heart...
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were found microscopically in the upper and lower lobes of the right lung.

Histologically, the brain did not contain any viable tumor mass. The grossly softened areas consisted of reactive tissue, mainly made up of macrophages and cellular debris within a loosened framework (Fig. 3). This softened zone merged with a marked astrocytosis that extended far beyond the apparent limits of the gross tumor and involved almost all of the white matter in both hemispheres and midbrain. Blood vessels at the margins of the softened zone showed prominent perivascular cuffing with tumor cells and lymphocytes, and contained abundant concentric strands of reticulin (Fig. 4).

Discussion

The histogenesis of the cell or origin of intracranial reticulum-cell sarcoma remains unclear. It is generally accepted that it originates from an undifferentiated mesenchymal element, and the same primitive cells appear to give rise to primary malignant reticulum-cell lesions in either CNS and extra-CNS sites. Slight variations in the morphology of the final mature forms may depend on the primary site of the tumor; however, the lesions are lymphoreticular tumors.

Reticulum-cell sarcoma of the CNS is a rapidly growing tumor that most commonly arises in the
cerebrum of the adult. It is usually a pale gray granular tumor with poorly defined margins. Histologically, the tumor is a dense cellular mass of non-uniform cells that range from small lymphocytic elements to larger cells having scant cytoplasm, prominent round or oval nuclei, and nuclear membranes, and may have a large nucleolus. Phagocytosed material may be evident in the cytoplasm of the large histiocytic elements. At the periphery of the lesion, the tumor mass gradually diminishes to perivascular collections of tumor cells and lymphocytes. Still more peripherally, the number of tumor cells around the vessels decreases until only a lymphocytic collection remains. Two characteristic histological features of this tumor relate to the vascular relationships of the tumor cells. First, the tumor cells do not merely occupy the perivascular spaces, but are intimately associated with the actual vessel walls, which may appear quite thickened. Second, there is an increase in the perivascular content of reticulin that occurs in a concentric laminated pattern.

Reticulum-cell sarcoma of the CNS occurs most frequently in cases associated with immunosuppression and possibly with immune deficiency. The risk of developing a lymphoreticular malignancy in recipients of renal transplants with immunosuppression has been reported to be as high as 35 times that of the general population. The majority of such malignancies are reticulum-cell sarcomas, and these occur in the transplant patient population 350 times more commonly than in the general population. There is also a remarkable tendency for reticulum-cell sarcoma in the transplant patient to occur as a primary CNS lesion. Reticulum-cell sarcomas of the CNS account for 50% to 73% of lymphoreticular tumors in transplant patients. This figure contrasts with the relatively low incidence of CNS reticulum-cell sarcomas in nontransplant patients, less than 1% of brain tumors.

Reticulum-cell sarcomas of the CNS may occasionally have extracerebral deposits of tumor in the lymph nodes, bone marrow, lung, and spleen. The rather brief survival periods of these patients may influence the failure of many of these tumors to extend outside the CNS. We have found no previous report of a primary reticulum-cell sarcoma metastasizing to the heart. Metastases to the heart generally are late manifestations of malignancy. The vast majority of tumors that secondarily involve the heart are intrathoracic malignancies, either primary or metastatic, and, of these primary tumors of the lung and breast, leukemia-lymphoma and melanoma, are the most common. Metastases to the heart from primary tumors of cutaneous or visceral origin are likely to be gross deposits, while metastases to the heart from the leukemia-lymphoma group are most often microscopic deposits. This incidence is in contrast to the cardiac metastasis in our case, which was a large mass quite evident on gross inspection. As demonstrated by our case, the heart may be rather extensively involved by metastases and still be asymptomatic.

Although the tumor in the brain was undergoing radiation-induced degeneration, the characteristic pattern of the perivascular reticulin strands, as well as the perivascular cuffing with tumor cells, still allow a diagnosis of reticulum-cell sarcoma to be made. The gross and microscopic features of the tumor in the basal ganglia are those of a primary tumor rather than a metastatic lesion. Reticulum-cell sarcoma of the brain occurs much more frequently as a primary tumor in the brain than as a metastasis from an extracerebral site to the brain.

References


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