CEREBRAL METASTASIS FROM A SILENT CARDIAC SARCOMA

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Sarcomas of the heart rarely may present a clinical picture in which distant metastases produce the dominant symptoms and signs. In a case of rhabdomyosarcoma of the heart, which was reported recently, the complaint presented was of bilateral masses of the breast. Blanes considered the possibility of distant metastases causing the initial symptoms in cases of cardiac tumors. Three cases of sarcoma are listed in Whorton’s review of tumors of the heart, in which cerebral metastases occurred in association with other distant sites of secondary tumor.

The patient in the following case had a fibrosarcoma of the mitral valve with a solitary metastasis which was in the brain. The clinical picture is unique because the symptoms were solely those of an expanding intracranial lesion.

**CLINICAL SUMMARY**

The patient, a 25-year-old civilian glaciologist, working on an ice floe in northern Canada, had right-sided Jacksonian seizures on Aug. 3, 1959. During the succeeding 4 days, a total of ten seizures were recorded. These were characterized by right-sided clonic and tonic contractions of the face and extremities, without loss of consciousness, lasting 1 to 3 minutes, each time with full recovery of motor function. On Aug. 8, 1959, he noted transient weakness of his right arm and leg with gradual progression to hemiparesis by 1 week after onset of symptoms. He complained of a dull left frontal headache aggravated by movement.

**Examination.** He was seen by a physician of the U. S. Air Force after evacuation to Thule, Greenland. Function of the cranial nerves was intact and sensory findings were normal. There was marked weakness with asterognosis and apraxia of the right arm and leg. Hyperreflexia of the right leg was noted, and a right Babinski’s sign was elicited.

He was evacuated by air to the U. S. Navy Hospital at San Diego, California. On arrival, Aug. 14, 1959, there was right hemiplegia, including right central facial paresis. Right homonymous hemianopia, complete right hemihypesthesia, and marked dysphasia also were noted. There was early bilateral papilledema. He continued to have severe left frontal headache and had vomited several times. Routine roentgenograms of the skull were normal.

On August 19, carotid angiography showed a marked shift of the left anterior cerebral artery to the right.

**Operation.** A left craniotomy was performed on Aug. 20, 1959. The left frontoparietal cortex appeared normal. With a ventricular needle, a small, black hematoma, 5 cm. in diameter, was found underlying the arm area of the motor cortex. After cortical incision the cavity of the hematoma was evacuated and the surrounding necrotic white matter was removed by suction.

**Biopsy** of the surrounding tissue revealed only gliosis. Gradually, the dysphasia improved, but the total right hemiplegia and sensory changes persisted. The wound healed well, the headache decreased, and he was eating a good diet postoperatively. He had two grand mal seizures September 22.

He was transferred for further neurological care to the Huntington Memorial Hospital, Pasadena, on Sept. 28, 1959. Physical examination revealed no cardiac murmurs, and no enlargement of the heart, liver, or spleen. Radiographically the heart was normal in size and configuration, and there was no tumor in the pulmonary fields.

Repeated left carotid angiography on Oct. 6, 1959 showed much less shift of the anterior cerebral artery. The posterior temporal arterial branches to the Huntington Memonal Hospital, Pasadena, on Oct. 6, 1959. Physical examination revealed no cardiac murmurs, and no enlargement of the heart, liver, or spleen. Radiographically the heart was normal in size and configuration, and there was no tumor in the pulmonary fields.

**Autopsy.** The brain weighed 1600 gm. On the left, in the anterior parietal region, a tumor presented on the surface. There was a shift of the midline structures to the right, with a marked tentorial pressure cone, and secondary hemorrhages of the midbrain. Coronal sections revealed a firm, red, discrete tumor, 8X6X7 cm. in size, above the left lateral ventricle, extending from 2 cm. behind the frontal pole to the occipital lobe. It was only loosely adherent to the surrounding brain (Fig. 1).

The heart weighed 360 gm., and there was borderline right ventricular hypertrophy. The epicardium was smooth and glistening. There was a firm nodular tumor, measuring 2.3X2.8X1.0 cm., in the anterior leaflet of the mitral valve. It extended from the ring of the valve to the free edge of the leaflet (Figs. 2 and 3), and slightly occluded the orifice. There were granular endocardial excrescences over portions of the tumor and over thickened attached portions of several chordae tendineae. The endocardium of the left atrium was thickened.
No other gross neoplasm was found. There was generalized hyperemia with hemorrhage throughout the lower lobes of both lungs.

**Microscopic Examination.** The tumor of the heart (Figs. 4 and 5) formed a nonencapsulated mass in the mitral valve and extended beneath the endocardium up into the atrial wall, up to the aortic valve and down into the chordae tendineae. In the mitral valve it had broken through the endocardium. Stain for reticulin and Mallory's phosphotungstic acid hematoxylin stain demonstrated fibers of reticulin and collagen, in some areas forming a delicate network about individual cells of the tumor, while in other areas collecting in dense bands with occasional hyalinization, widely separating neoplastic cells. Capillaries were abundant in some areas. The neoplastic cells were predominantly fat and spindle-shaped, but varied markedly in size and shape. Occasional multinucleated cells and rare mitotic figures were seen. There was marked clumping of chromatin, and considerable variation in the size and shape of the nuclei was noted. No intracellular longitudinal or cross striations were found in sections stained by phosphotungstic acid hematoxylin and Gümöri's trichrome stain.

The brain tumor (Fig. 6) was similar but had a more delicate network of collagen and was more vascular and cellular. Anaplastic cells, multinucleated cells, and mitoses were seen more frequently. There were necrotic foci. No pseudopalisading of nuclei or endothelial proliferation occurred. Neuroglia could not be demonstrated. Multiple sections of the brain adjacent to the tumor showed no invasion.

No tumor was encountered microscopically in any of the thoracic or abdominal viscera except the heart. Generalized acute passive hyperemia was present throughout the organs. There was hemorrhage in some sections of the lungs and bronchi, and multiple variably sized foci of bronchopneumonia, usually accompanied by hemorrhage, were present.

**DISCUSSION**

Primary malignant tumors of the heart were reviewed in 1949 by Whorton, who added a case to the 99 reported previously. Brucker and Glassy in 1955, brought the total reported cases to 148. In these reviews, there were 4 tumors arising in cardiac valves, and 2 other cases were found in a survey of the English literature to the present date. One of these was a fibrosarcoma of the mitral valve with a solitary vertebral metastasis.

The present case is of especial interest as several examiners found no clinical evidence suggesting a cardiac lesion, and roentgenogram of the chest was normal. The solitary cerebral lesion was the only metastasis found on autopsy. The vascularity of the cerebral metastasis, as shown by Fig. 6, perhaps explains the rapid progression of symptoms at the onset of illness and the intra-