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 astereognosis 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 hemihypoglossia, 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.** Course. 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 neurosurgical 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 artery could be seen but other branches of the middle cerebral artery were demonstrated incompletely, suggesting a lesion in the left frontotemporal area. His clinical course, especially motor power and dysphasia, continued to improve gradually and progressively, and in view of the angiographic improvement, surgery was deferred. He was discharged home Oct. 14, 1959.

On November 1, 2 weeks later, he became progressively somnolent and confused. The headaches on the left became intense and were accompanied by vomiting. He was re-admitted for further treatment on Nov. 6, 1959. Papilledema had increased to 3 diopters, and his hemiplegia again was complete. He rapidly became comatose, and died.

**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 2.3X2.3X1.0 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 ex crescences over portions of the tumor and over thickened attached portions of several chordae tendineae. The endocardium of the left atrium was thickened.

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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 Gomori'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-
cerebral hematoma found at operation. Postoperatively, the neurologic and angiographic improvement were confusing features.

We feel the cerebral tumor was secondary to the cardiac tumor for the following reasons: The lungs were not involved. Metastases to the cardiac valves are rare,¹⁰ and when they do occur, unlike the present tumor, they resemble vegetative endocarditis. The tumors were similar histologically. The presence of greater anaplasia in the metastasis than in the primary tumor is said by Willis⁸ to be frequent.

Fig. 4. Photomicrograph of tumor of the valve showing invasion of the endocardium. (Hematoxylin and eosin, low power.)

Fig. 5. Photomicrograph of cellular portion of tumor of mitral valve. (Hematoxylin and eosin, high power.)
We classify this neoplasm as a poorly differentiated fibrosarcoma, rather than as a tumor of muscle or perineurial sheath, because of its abundant collagen and predominantly spindle-shaped cells, which show no nuclear palisading, which do not stain like muscle, and in which neither cross striations nor myofibrils can be identified.

SUMMARY

A unique case of fibrosarcoma of the mitral valve with a cerebral metastasis is described in which the presenting symptoms were those of a primary brain tumor. The primary cardiac tumor was not discovered until autopsy, at which time the brain proved to be the only site of metastasis. A brief survey of the literature of primary malignant cardiac tumors is given. Of these cases, in only 6 were the primary tumors in the valves. The absence of cardiac symptoms in the clinical picture of cardiac tumor is a rare feature of this case.

We wish to thank Dr. William C. Manion and other members of the staff of the Armed Forces Institute of Pathology for examining histologic sections from this case and for reporting to us their diagnosis of primary sarcoma arising in the leaflet of the mitral valve with metastasis to the brain.

REFERENCES