ing stress on the aortic wall and may prevent or slow progression of the dissection.* In the usual case of proximal aortic dissection, medical therapy is used only as a temporizing measure until the patient can be referred for surgical repair of the dissection. In this patient's chronic proximal dissection, blood pressure control, along with conservative management of her recurrent episodes of cardiac tamponade, provided time for the local healing that allowed her to survive more than four years without surgery. To our knowledge, such long-term survival following nonsurgical management of aortic dissection complicated by cardiac tamponade has not been previously reported.

REFERENCES

Primary Malignant Fibrous Histiocytoma of the Aorta Associated with Aortic Dissection*

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We report a unique case of primary aortic malignant fibrous histiocytoma presenting clinically as aortic dissection. To our knowledge, this occurrence is the first ever reported. The magnetic resonance imaging technique may provide superiority in the differential diagnosis between tumor and hematoma of aortic dissection. Aortic tumor, although rare, should be included among the causes of aortic dissection. (Chest 1991; 99:1049-50)

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Primary tumors of the aorta are extremely rare, and less than 30 cases have been published in the literature.1 4 Clinically, for the patient with a primary aortic tumor, the most common presenting symptom is pain, either from local tissue destruction or ischemia due to invasion of branch vessels or arterial tumor embolization. Other symptoms and signs include hypertension, decreased peripheral pulse, claudication, or nonspecific manifestations of malignancy such as fever, malaise, anorexia, and weakness. Herein we report a unique case of primary malignant fibrous histiocytoma of the proximal descending aorta associated with DeBakey type 3 aortic dissection. To the best of our knowledge, a primary aortic tumor presenting clinically as aortic dissection has not yet been reported.

CASE REPORT

A 55-year-old man was admitted because of intermittent chest pain with radiation to the back for two weeks. He had had hypertension for five years. The case was first diagnosed as DeBakey type 3 aortic dissection by means of digital subtraction angiography and CT (Fig 1) seven months ago. Thereafter, the patient received medical treatment, with an uneventful course.

The recurrence of symptoms during the past two weeks made him seek help in our hospital on Aug 1, 1989. Physical examination revealed a well-oriented man in acute distress. His blood pressure was 140/84 mm Hg. All pulses in the neck and extremities were symmetric. A grade 2 systolic ejection murmur was audible over the left sternal border. Other findings from physical examinations were unremarkable. A chest x-ray film showed cardiomegaly and a widened mediastinum. Another CT study to define the evidence of progression of aortic dissection failed due to an allergy to the contrast medium. Instead, MRI was performed and showed aortic dissection from the distal aortic arch to the abdominal aorta. A mass shadow, located at the aortic arch, was interpreted as a hematoma in the false lumen due to retrograde extension of aortic dissection (Fig 2). The patient was referred for surgery. During surgery a solid mass measuring 7 × 3.5 × 3 cm located at the proximal descending aorta was noted. An aortic dissection with intimal tear and inlet of a false lumen just under the tumor was also noted. The tumor was growing out from the external aspect of the false lumen.

The excised tumor was sent for pathologic examination. Grossly, it was elastic and firm, with a brownish mucoid area separated by fibrous trabeculae. Microscopically, the tumor was composed of various cellularity in different areas. In the cellular area, spindle-shaped tumor cells arranged in a storiform or whorled pattern with pleomorphism were noted (Fig 3). The immunohistochemical study was negative for markers of muscular or nervous origin. Malignant fibrous histiocytoma was the final pathologic diagnosis. After operation, the patient recovered uneventfully.

FIGURE 1. Thoracic CT scan showing aortic dissection with thrombus formation in false lumen. "Mass" (arrow) over aortic arch was misinterpreted as hematoma.
FIGURE 2. Magnetic resonance imaging (coronal section). Tumor mass (arrow) was noted over aortic arch, which pushed trachea to right side. Thoracic aorta was divided into false and true lumen by intimal flap.

**DISCUSSION**

Wright et al reviewed 22 cases of primary aortic tumor previously reported in the literature. They suggested that aortic tumors primarily involving the intima present with symptoms referable to thromboembolic metastasis or luminal obstruction, whereas tumors invading the media or adventitia have a locally aggressive mass lesion with limited metastasis. In our unique case the aortic tumor involved the mural layer, which demonstrated an intramural growth pattern and presented clinically as aortic dissection.

Because of their rarity, primary aortic tumors are seldom suspected and diagnosed in clinical practice. Clinicians should be highly alert to list primary aortic tumor in the differential diagnosis. Radiology plays an important role in the diagnosis of aortic tumors. A characteristic angiographic finding shows an irregular filling defect within the aortic lumen, with the remainder of the aorta appearing relatively smooth and of normal caliber; however, it is difficult to use aortography to demonstrate extra-aortic extension. Computed tomography is helpful in defining the extra-aortic extent; however, when the tumor primarily involved adventitia, differentiation from mediasinal or retroperitoneal malignancy is usually difficult. In addition, CT does not differentiate intraluminal tumor from thrombus. In the present case, since the tumor was located outside the false lumen, it was impossible to diagnose by angiography. The concomitant existence of the aortic dissection also caused the tumor to be misinterpreted as hematoma or extravasation on the CT study.

Magnetic resonance imaging is an innovative and powerful diagnostic tool for the evaluation of aortic disease, especially for aortic dissection. It effectively displays aortic wall and periaortic abnormalities. In the evaluation of suspected thoracic aortic dissection, MRI provides a sensitivity and specificity higher than 90 percent. Tissue characterization on the basis of magnetic relaxation time is another potential superiority of MRI over CT or angiography. The MRI of the present case demonstrated aortic dissection of the thoracic aorta and a mass shadow over the aortic arch. If we are highly alert for the possibility of primary aortic tumor, it is possible before surgery to diagnose with certainty by the advanced MRI technique.

It is of particular interest to associate the primary aortic tumor with the pathogenesis of aortic dissection. As we know, degeneration or weakness of the aortic media is the prerequisite for the development of aortic dissection. In the present case, weakness of aortic media related to mural aortic sarcoma, combined with the shearing force on the aorta imposed by hypertension, might be responsible for the aortic dissection. The dissection inlet just under the tumor and the tumor located just outside the false lumen lend strong support to our presumption.

**REFERENCES**