Virchows Archiv A Path. Anat. and Histol. 379, 351–358 (1978)

Virchows Archiv A

Case Report

Primary Fibroxanthosarcoma of the Thoracic Aorta

Christopher P. Crum [1], Philip S. Feldman [1], and Stanton P. Nolan [2]

[1] Department of Pathology, University of Virginia Hospital, Charlottesville, Virginia, USA
[2] Department of Surgery, University of Virginia Hospital, Charlottesville, Virginia, USA

Summary. A case is presented of a 58-year-old female with a primary malignant tumor of the aorta. The lesion was diagnosed histologically as a fibroxanthosarcoma and represents the first primary neoplasm of the aorta so classified. The gross and microscopic features of the lesion are described and the morphologic characteristics of previously reported aortic sarcomas are briefly reviewed and examined in light of the current histologic criteria for soft tissue tumors.

Key words: Fibroxanthosarcoma – Malignant fibrous histiocytoma – Aorta – Sarcoma.

Introduction

Primary tumors of the aorta and systemic arteries are rarely seen. From his own experience and a review of domestic and foreign literature, Salm described only thirteen well documented cases (Salm, 1972). One additional case has since been reported (Steffelaar et al., 1975).

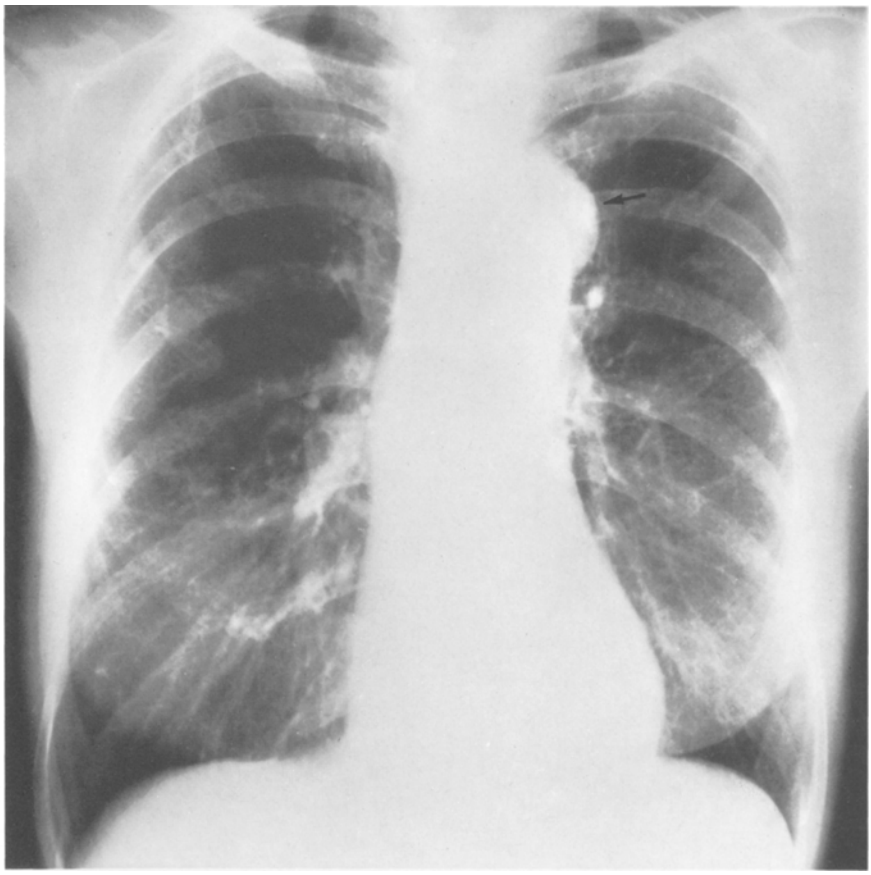
The present report illustrates a case of an adventitial sarcoma of the thoracic aorta which was classified histologically as a fibroxanthosarcoma. The gross and histologic features of this uncommon neoplasm are described.

Report of a Case

A 58-year-old white female was admitted to the University of Virginia Medical Center on March 18, 1976, with a ten day history of right pleuritic chest pain and four days of left pleuritic chest pain. A chest x-ray had revealed an abnormal density in the area of the aortic knob (Fig. 1A). The patient had apparently been in good health all of her life with the exception of mild hypertension which was well controlled. On physical examination the blood pressure was 150/90 mm of mercury

For offprints contact: Christopher P. Crum, M.D., Box 214, University of Virginia Hospital, Charlottesville, Virginia 22908, USA

0340-1227/78/0379/0351/$01.60



A

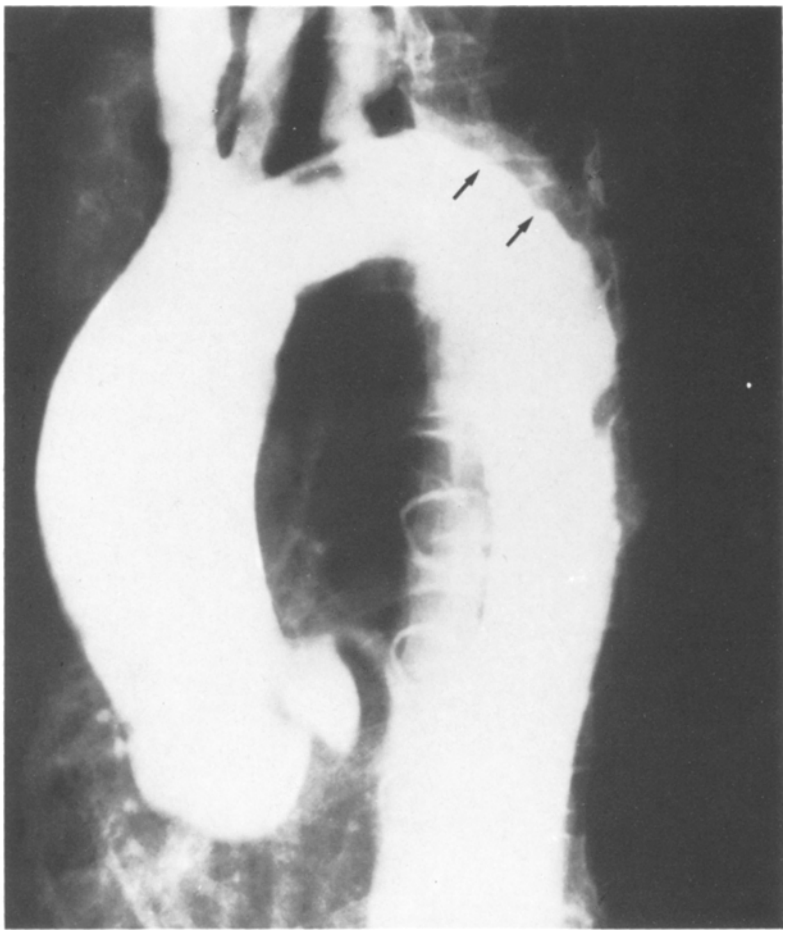
Fig. 1. A An admission chest film revealed a soft tissue density in the region of the aortic knob (*arrow*). **B** An aortogram illustrated irregularities in the intima associated with the tumor (*arrows*)

and a grade 2 systolic ejection murmur was heard along the left sternal border. An intermittent thrill could be palpated in the second left intercostal space.

Laboratory data included a hematocrit of 36% and a white blood cell count of 7200/mm^3. The only laboratory abnormality was a serum alkaline phosphatase elevated to 120 I.U. which remained elevated on all subsequent determinations. Fluoroscopic examination confirmed that the density was physically associated with the first portion of the descending aorta. In addition, a 4 by 5 cm soft tissue mass was present at the junction of the aortic arch and the descending aorta but failed to fill with contrast medium (Fig. 1B). These findings were thought to be compatible either with a tumor mass in the region of the aortic isthmus or a thoracic aortic aneurysm with luminal obliteration.

On March 25, 1976, a left exploratory thoracotomy was performed. A 4.5 cm diameter spherical mass arose from the posterior and lateral portions of the descending thoracic aorta, 3 cm distal to the left subclavian artery. The mass was firm and yellow in color. Palpation of the aortic wall demonstrated marked thickening extending proximally and distally for approximately 6 cm. There were no palpable atherosclerotic plaques. A biopsy of the tumor mass was performed and a frozen section diagnosis of sarcoma was made. The chest was then closed.

The patient recovered satisfactorily from the operation. A subsequent bone scan revealed multiple areas of increased uptake in the right tibia, left pelvis and left kidney. A liver scan was performed



B

and demonstrated a defect in the right lobe of the liver. It was felt that radiotherapy would not be indicated because of the risk of tumor necrosis and aortic rupture. Therefore, plans were made for chemotherapy which would be instituted at the time that the patient developed further symptoms. The patient returned home and apparently did well until May 17, 1976, when she was admitted through the Emergency Room in status epilepticus with focal motor clonic seizures involving the right face, neck, arm and leg. In the hospital she was treated with dexamethasone and diphenylhydantoin, but expired on the third day with cardiorespiratory arrest.

Autopsy

At autopsy a well circumscribed, yellow, fibrous mass measuring $6 \times 4 \times 4$ cm was found to be attached to the posterior lateral aspect of the descending aorta just distal to the left subclavian artery (Fig. 2). The tumor was adherent, but not fixed to the visceral pleura of the left posterior aspect of the lung and infiltrated focally into the posterior mediastinal soft tissues. The underlying aortic wall was indurated beneath the tumor, and sectioning revealed the intimal

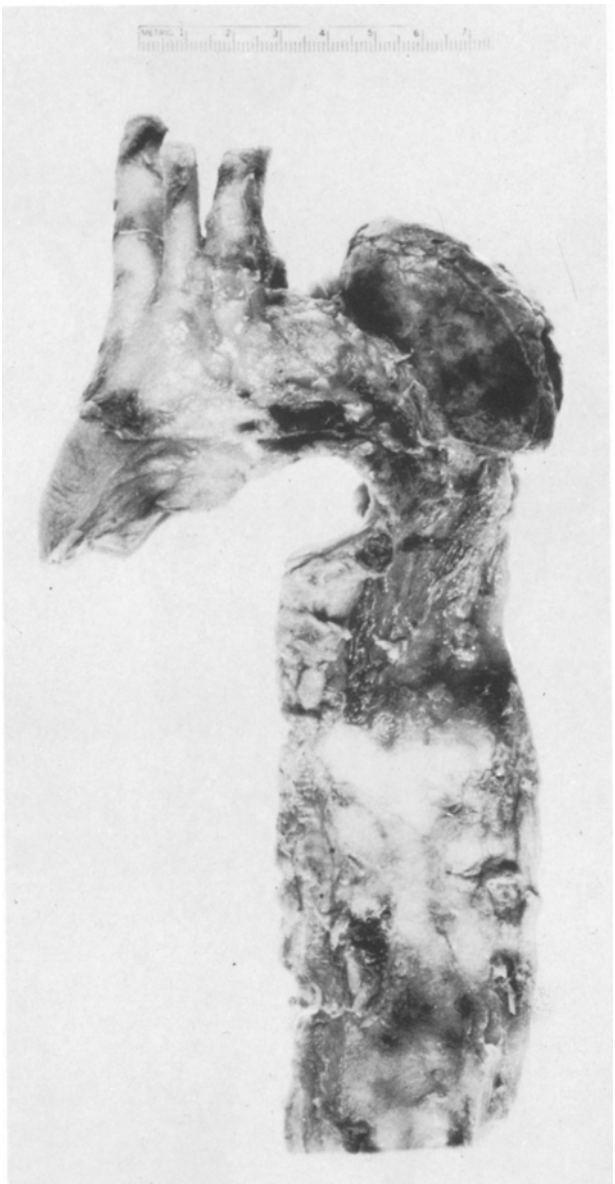


Fig. 2. The aorta at autopsy contained a broad based tumor projecting outward from the adventitial surface

surface to be covered with masses of soft, pale thrombus (Fig. 3). The thrombus was loosely adherent to the intima and extended proximally to occlude the orifices of the vessels of the aortic arch. A normal intimal surface resumed three centimeters distal to the lesion and the remainder of the aorta showed mild atherosclerosis. Disruption of the media by the tumor was not seen grossly.

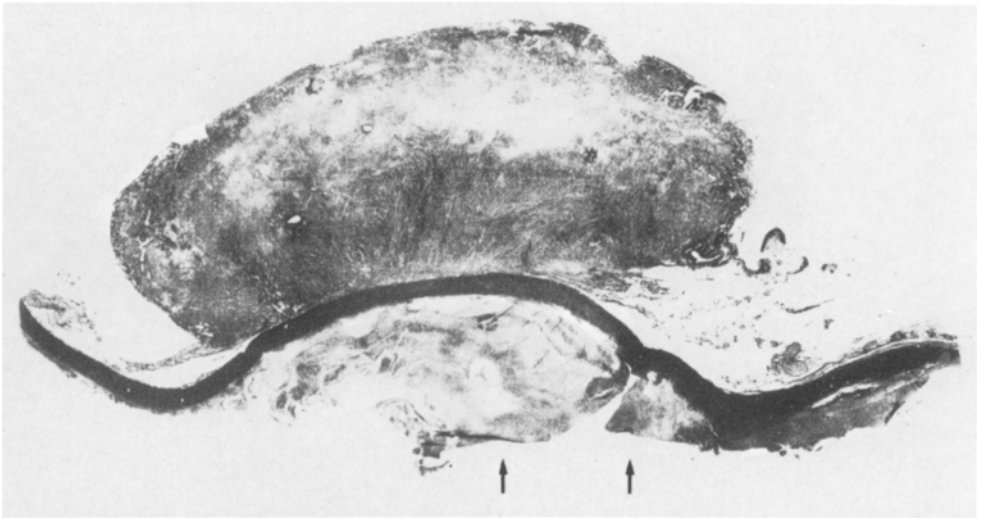


Fig. 3. A whole mount section of the aortic tumor. Note the uniform appearance of the media beneath which there was extensive secondary intimal replacement by tumor thrombus (*arrows*) (X2 H & E)

The remainder of the gross examination disclosed pulmonary congestion and focal infarcts in the kidneys, spleen, small bowel and brain. Grossly visible thrombi were observed in the vessels supplying all of these organs. There was no gross evidence of metastatic disease in the thoracic or abdominal organs and sections of the vertebral and costal bone marrow likewise appeared normal.

Histologic examination of the biopsy and necropsy material from the tumor disclosed a poorly differentiated sarcoma with marked architectural and cellular pleomorphism. At low magnification some areas demonstrated a myxoid pattern whereas others were densely cellular with a storiform arrangement (Fig. 4). Foci of necrosis with hemorrhage and acute inflammation were abundant. On examination at a high power a variety of cell populations were observed. The predominant cell type consisted of poorly differentiated mesenchymal cells which resembled fibroblasts and contained spindle shaped to ovoid nuclei. These cells were numerous throughout the lesion but predominated in the storiform areas. They were admixed with a second cell type characterized by round, irregular, pleomorphic nuclei, prominent nucleoli and cytoplasm which varied from pale to eosinophillic. Numerous bizarre, multinucleated tumor giant cells were present and contained dense, eosinophilic pale cytoplasm which was sometimes vacuolated (Fig. 5). Many mitotic figures were noted, some of which were bizarre. Small clusters of xanthoma cells were also observed. A Masson trichome stain revealed a diffuse and extensive deposition of collagen which was more abundant in the storiform areas. On the basis of these microscopic features the diagnosis of fibroxanthosarcoma was made and confirmed by Richard L. Kempson, MD, of Stanford University Medical Center and David C. White, MD, of The Armed Forces Institute of Pathology.

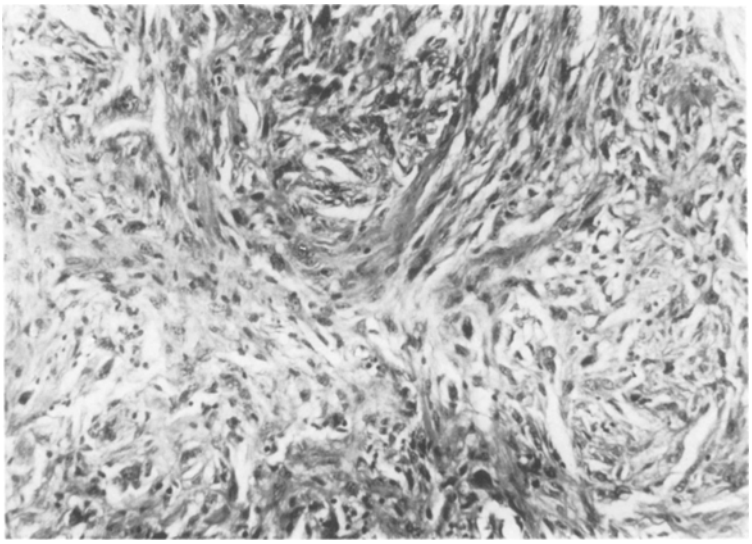


Fig. 4. Fibroxanthosarcoma of the aorta. This microphotograph illustrates the characteristic storiform arrangement of fibroblasts within a collagen stroma (X80 H & E)

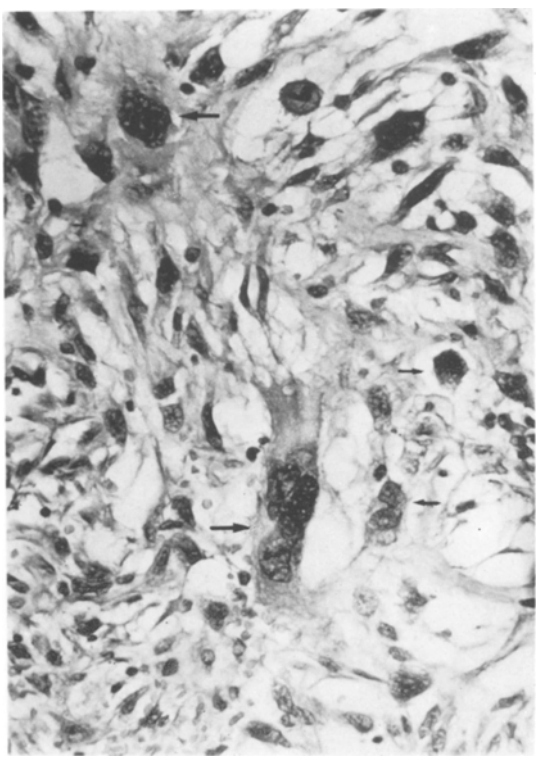


Fig. 5. Fibroxanthosarcoma. At higher magnification a biphasic population of spindle shaped fibroblasts and histiocyte-like cells (*small arrows*) are observed. Multinucleated tumor giant cells (*large arrows*) are also present (× 500 H & E)

Although the media showed no evidence of tumor invasion microscopically, the intima was replaced by amorphous fibrinoid material in which numerous neoplastic cells were present in a myxoid matrix. In addition, tumor cells were arranged in parallel rows along the luminal surface of the thrombus where they were covered by a thin layer of fresh thrombin.

Histologic sections from the other organs, including liver, lung, brain and bone marrow failed to show metastatic sarcoma. Numerous sections from the vascular thrombi revealed tumor in the renal thrombi only. The cerebral/vasculature appeared patent despite the presence of numerous, superficial, cerebral cortical and cerebellar infarcts. The cause of these infarcts was presumably that of generalized cerebral and cerebellar anoxia secondary to obstruction of the common carotid arteries at their origin by sarcoma.

Comment

This report represents, to the best of our knowledge, the first description of a primary fibroxanthosarcoma of the aorta. The histologic findings of a storiform pattern with fibroblasts, histiocyte-like cells, multinucleated tumor giant cells and xanthoma cells have been previously established as pathognomonic for this subtype of malignant fibrous histiocytoma (Kempson and Kyriakos, 1972; Merkow et al., 1971). The primary nature of the tumor is demonstrated by the observation at operation that it was strictly confined to the aorta and to the lack of other organ involvement at autopsy. The features of intimal invasion, systemic embolization and rapid clinical deterioration are reminiscent of cases previously described (Salm, 1972).

The majority of primary tumors of the aorta reviewed by Salm were classified as fibrosarcomas or fibromyxosarcomas (Salm, 1972). The diagnosis was generally based on the observance of a spindle cell sarcoma with a variable degree of myxoid change. Of interest is the mention of cellular pleomorphism and/or multinucleated tumor giant cells in some of the cases (Salm, 1972). The latter histologic findings are not currently accepted as characteristic of fibrosarcomas (Pritchard et al., 1974; Stout and Lattes, 1967). This discrepancy is, in part, understandable when viewed in the context of the confusion that has previously surrounded the histologic classification of soft tissue tumors (Fu et al., 1975; Pritchard et al., 1974; Soule and Enriquez, 1972; Stembridge et al., 1964; Stout and Lattes, 1967). Furthermore, many of the known cases of primary aortic sarcoma were published prior to the advent of electron microscopy (Salm, 1972). The subsequent expansion and diversification of the nomenclature for the soft tissue tumors has led to refinements in histologic classification which have resulted in the renaming of tumors heretofore diagnosed as fibrosarcomas (Fu et al., 1975; Pritchard et al., 1974; Soule and Enriquez, 1972; Stembridge et al., 1964; Stout and Lattes, 1967).

The category of malignant fibrous histiocytoma is one of many into which selected fibrosarcomas have been reclassified (Pritchard et al., 1974; Stout and Lattes, 1967). Inasmuch as the features of myxoid changes, cellular pleomorphism and multinucleated giant cells have been observed in malignant fibrous

histiocytomas, it is conceivable that some of the aortic tumors previously reported might warrant this diagnosis when examined in the light of current histologic criteria (Weiss and Enzinger, 1977).

References

Fu, Y-S, Gabbiani, G., Kaye, G.I., Lattes, R.: Malignant soft tissue tumors of probable histiocytic origin (malignant fibrous histiocytomas): General considerations and electron microscopic and tissue culture studies. Cancer **35**, 176–185 (1975)

Kempson, R.L., Kyriakos, M.: Fibroxanthosarcoma of the soft tissues. Cancer **29**, 961–976 (1972)

Merkow, L.P., Frich, J.C., Stifkin, M., Kyreages, C.G., Pardo, M.: Ultrastructure of fibroxanthosarcoma (malignant fibroxanthoma). Cancer **28**, 372–383 (1971)

Pritchard, D.J., Soule, E.H., Taylor, W.F., Ivins, J.C.: Fibrosarcoma—A clinicopathologic and statistical study of 199 tumors of the soft tissues of the extremities and trunk. Cancer **33**, 888–897 (1974)

Salm, R.: Primary fibrosarcoma of the aorta. Cancer **29**, 73–83 (1972)

Soule, E.H., Enriquez, P.: Atypical fibrous histiocytoma, malignant fibrous histiocytoma, malignant histiocytoma, and epithelioid sarcoma. Cancer **30**, 128–142 (1972)

Steffelaar, V.W., van der Heul, R.O., Blackstone, E., Vos, A.: Primary sarcoma of the aorta. Arch. Path. **99**, 139–142 (1975)

Stembridge, V.A., Luibel, F.J., Ashworth, C.T.: Soft tissue sarcomas: Electron microscopic approach to histogenetic classification. South Med. J. **57**, 772–779 (1964)

Stout, A.P., Lattes, R.: Tumors of the soft tissues. Atlas of tumor pathology, fasc 1. pp 101–115. AFIP, 1967

Weiss, S.W., Enzinger, F.M.: Myxoid variant of malignant fibrous histiocytoma. Cancer **39**, 1672–1685 (1977)

Received February 14, 1978

Anmerkung der Schriftleitung

In der Sammlung des Pathologischen Institutes der Universität Heidelberg befindet sich das Präparat eines „emboliformen Sarkomes" der Arteria pulmonalis. Der Fall wurde von Prof. C. Froboese vor fünfzig Jahren sehr genau untersucht *und* geklärt. Froboese erörterte bei dem auf der Pulmonalisgabel reitenden, 5:3:3 cm messenden Gebilde das Vorliegen einer etwaigen sarkomatösen Organisation eines Embolus, kam aber aus guten Gründen zu der Überzeugung, daß es sich um ein primäres, vorwiegend spindelzelliges Sarkom der Intima gehandelt hatte. (Zbl. Path. **44**, 148–153, 1928).

Wilhelm Doerr (Heidelberg)