Intimal Sarcoma of the Descending Aorta

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Primary intimal angiosarcoma of the aorta (i.e., mostly intraluminal sarcomas with evidence of endothelial differentiation) is extraordinarily rare. We report a case in which the diagnosis was accurately made using immunohistochemistry in an embolecotmy specimen. The patient was a 78-year-old man with a two-month history of bilateral claudication. Doppler ultrasound proved an embolus in both popliteal arteries, which was removed. The highly atypical cells comprising these emboli were positive immunohistochemically for CD68, vimentin, and CD31. Magnetic resonance imaging also showed an irregular tumor (invasion to the left main bronchus). This case emphasizes the need for a wide panel of immunohistochemical studies in tumor emboli of unknown origin.

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peripheral emboli or grow along the lumen. The mural type originates from media or adventitia and extends to paraaortic tissues. In most cases, the intimal subtype originates from the descending thoracic or abdominal aorta. Most cases are diagnosed at autopsy. The symptomatic patients usually have signs of tumor embolus in lower extremities or mesenteric arteries. The disease is more prevalent in men with a mean age of 60 years. The work-up of a patient presenting with peripheral emboli, begins with echocardiography. Once a cardiac source has been excluded, magnetic resonance angiography (MRA) of the aorta is the most sensitive imaging modality for detection of the tumor. MRI study can differentiate tumor from atheromatous plaque by enhancement and reveals the extension of tumor to adjacent structures. Compared with conventional angiography, there is no risk of catheter-induced embolization or contrast-induced nephrotoxicity with MR study. When malignant tumor of aorta is suspected in MR, bone scintigraphy should be carried out, since the rate of bone metastasis is high. If bone metastases are confirmed, major surgical intervention is not indicated. Immunohistochemistry is important for making a definite diagnosis of intravascular malignancy. Positivity for CD31, von-Willebrand factor, and Ulex Europeaus with negative result for CD34 are in favor of intimal angiosarcoma. In patients without bone metastasis, treatment consists of en-bloc resection with chemo-radiotherapy, but prognosis is still poor with a life expectancy at diagnosis of almost 14 – 27 months.

References