Editorial Commentary

More on Pulmonary Artery Sarcoma

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The following single-case reports on pulmonary artery sarcoma (PAS)[1,2]—2 of them in number—emphasize its rarity and underscore its characteristic propensity to masquerade as pulmonary thromboembolism (PTE). Correct diagnosis and complete resection are the key to survival. In both patients, the clinical diagnosis was PTE—acute in one,[1] subacute-to-chronic in the other.[2] Nevertheless, in each case, pulmonary embolectomy uncovered PAS, not PTE. One of the patients died on the 10th postoperative day.[2] The other patient was alive and well with no evident disease 36 months after surgical resection, chemotherapy, and radiation therapy. These 2 reports end with essentially the same advice: consider PAS whenever you suspect PTE, especially in patients who have no evidence of deep vein thrombosis and who respond poorly to anticoagulant or thrombolytic therapy.

At first blush, that advice seems sound. On reflection, however, I find that it should be amended. To begin with, PAS characteristically arises close to the pulmonary valve and grows distally.[3] This means that the tumor typically occupies at least the pulmonary artery trunk or one or both of the main branches. Consequently, imaging studies that show filling defects confined to the lobar arteries or beyond—findings common in PTE—essentially eliminate PAS as a diagnostic possibility. So in that light, one need not consider PAS every time one considers PTE.

Second, in my extensive clinical experience with PTE,[4] patients who have this disease frequently do not have evidence of deep vein thrombosis. Therefore, the absence of such evidence is not a special reason to consider PAS, particularly when PTE is far more likely than PAS in the first place.

In conclusion, I thank the authors of both reports for making me and our readers more aware of PAS. As a result, I now view PAS as PAS—problematic and sinister.

References

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