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Pancreatic Extraskeletal Ewing Sarcoma/Primitive Neuroectodermal Tumor

Jasmine L Martin

Charles E Schmidt College of Medicine, Florida Atlantic University, Boca Raton, Florida, United States

Abstract

Ewing sarcoma, characterized by an 11:22 translocation, commonly occurs in bone and soft tissue sites. We present a patient with a history of both chest wall desmoid tumor and a subsequent adenosquamous pancreatic cancer treated with distal pancreatectomy, adjuvant chemotherapy, and radiation. Two years later the patient developed lymphadenopathy, which was biopsied and sent for the tissue of origin testing. Results revealed a sarcoma with 90% certainty, primitive neuroectodermal tumor subtype. The original pancreatic tumor was sent for next-generation sequencing, which revealed an EWSR1/FLI1 fusion as the sole genetic mutation suggestive of an underlying Ewing sarcoma. Additionally, the same fusion gene was detected on the supraclavicular lymph node biopsy. Our case affirms a change in diagnosis that was not suspected clinically with a resultant change in therapy. Though the pancreas is an unusual site of extraskeletal Ewing sarcoma (EES), emerging technologies such as next-generation sequencing can aid in management.

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Jasmine L Martin Charles E Schmidt College of Medicine, Florida Atlantic University, 777 Glades Rd, Boca Raton, Florida, 33431, USA