

Littoral cell Angioma (LCA) is a rare, potentially malignant tumor which originates from the red pulp sinuses of the spleen. While typically benign, the rate of malignant transformation is unclear, and malignancy cannot be determined until final pathology. Because it arises from the vasculature of splenic tissue, LCA has the potential to affect accessory spleens (splenules), which may exist in multiple locations within the abdomen, most commonly the splenic hilum. Few cases in the literature have reported LCA of splenules, and mainly describe splenules within the hilum. We present a unique case of a patient with LCA affecting both the spleen and an intrapancreatic splenule within the pancreatic tail. In this case the patient was found to have numerous non-specific splenic masses noted incidentally on computed tomography (CT) of the chest during a work-up for dyspnea. Magnetic resonance imaging (MRI) revealed numerous splenic lesions as well as a lesion of the pancreatic tail concerning for LCA, which was confirmed via percutaneous splenic biopsy and a EUS guided biopsy of the pancreas. As no guidelines exist for management of LCA, the options of surveillance and surgical resection were presented, and the patient elected to proceed with resection via laparoscopic distal pancreatectomy and splenectomy. The patient tolerated the procedure well and was discharged on post-operative day two. Final pathology confirmed the diagnosis and ruled out malignant transformation. The patient has no evidence of persistent or recurrent disease on surveillance imaging 2 years post-operatively and has had no post-operative complications. There have only been four reported cases of LCA in an accessory spleen, and there are no official guidelines for treatment or surveillance of LCA. The options of surveillance and resection should be discussed thoroughly with patients presenting with LCA to help guide a patient-centered management approach. This case describes the effective, minimally-invasive management of LCA which should be considered as a treatment option when presented with this rare diagnosis.