Clear Cell Sarcoma of Tendon and Aponeuroses (CCTA), also known as Melanoma of soft tissue is a rare and highly malignant soft tissue neoplasm which mostly occurs in young adults. A 26-year-old female presented with pain and thickened soft tissue in the popliteal fossa at the posterior aspect of the left knee. Ultrasonography demonstrated a cystic mass with irregular wall and internal debris representing a complex popliteal cyst. Further work-up with MRI demonstrated a cystic mass lateral to the semimembranosus muscle tendon. Excisional biopsy of the mass was consistent with malignant neoplasm with plasmacytoid features. Immunoperoxidase and cytogenic studies supported the diagnosis for Clear Cell Sarcoma arising from the tendon sheath. A Positron emission tomography showed central tumor necrosis and metastasis to hemi-pelvis lymph nodes. During the late stage of chemo-radiation, the patient was hospitalized due to increased abdominal distention and pain. A computerized tomography of the abdomen demonstrated extensive intra-abdominal and retroperitoneal metastatics, peritoneal carcinomatosis, omental caking and extensive ascites. Patient opted comfort care and died later.

This case emphasizes the importance of promptly establishing a correct diagnosis in patients with persistent leg pain. It also illustrates the aggressive nature of CCTA. Despite extensive surgical excision and adjunctive chemo-radiation, adequate control of local disease was not achieved and tumor metastasized to the abdomen. CCTA is a diagnostic challenge due to its benign presentation, histopathological resemblance with malignant melanoma and a therapeutic dilemma because of its poor prognosis secondary to an aggressive nature and early metastasis.