

## CASE REPORT

# Post-Radiation, Primary Pancreatic Sarcoma Diagnosed by EUS-FNA

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### ABSTRACT

We report a case of post-irradiation pancreatic sarcoma occurring in a 53 year-old woman, forty-nine years after radical nephrectomy for Wilms tumor. Sarcomas comprise less than 1% of pancreatic tumors. They are aggressive neoplasms with poor prognosis. Second malignancies are an issue following treatment of childhood tumors. This is the first reported case of pancreatic sarcoma following radiotherapy for Wilms tumor. The article also highlights the increasing role of EUS as a primary modality for diagnosing unusual malignancies.

### INTRODUCTION

This is the first reported case of pancreatic sarcoma following radiotherapy for Wilms tumor. The article also highlights the increasing role of EUS as a primary modality for diagnosing unusual malignancies.

### CASE REPORT

A fifty-three-year-old woman presented with a 12 month history of dyspepsia and unexplained lumbar pain, with weight loss of 20 pounds over 3 months and new onset diabetes mellitus. Past medical history was significant for Wilms tumour diagnosed at the age of four, treated with right nephrectomy and chemo-radiation, discharged from oncologic follow up.

Her physical examination was unremarkable with no icterus, lymphadenopathy, hepatosplenomegaly or ascites. Blood tests including liver function tests and tumor markers CA 19-9 and CEA were normal. Contrast enhanced CT showed a large 9x6cm lobulated necrotic retroperitoneal mass within the right nephrectomy bed with patent celiac axis, circumferential invasion of superior mesenteric artery and superior mesenteric vein inferior to the HOP showing possible tumor thrombus. MRI had shown the tumor in addition to be extending into duodenal wall and distal bile duct without occlusion, with multiple

small hepatic nodules suggestive of metastasis. CT and MRI demonstrating the pancreatic mass is illustrated in **Figure 1(a-c)**. EUS demonstrated a 7x7 cm lobulated necrotic mass in the head of pancreas with vascular invasion (**Figure 1d**). Rapid on-site assessment of EUS-FNA showed malignant epithelioid cells in groups (**Figure 2a**). The cell block showed spindle cell morphology (**Figure 2b**). An extensive immunohistochemical analysis was performed. The tumor was immunoreactive for vimentin, desmin and smooth muscle actin and negative for various other markers, thus ruling out an epithelial malignancy or other spindle cell lesions (S100, CD56, AE1/3, CK7, CK19, CD117, DOG1, myogenin and MyoD1). The cytomorphology and immunohistochemistry was supportive of a diagnosis of sarcoma, NOS (not otherwise specified) (**Figure 2 cd**).

The tumor was determined to be unresectable. The patient elected for palliative chemotherapy and was treated with Gemcitabine and Docetaxel.

### DISCUSSION

Wilms tumour is a common pediatric malignancy, with a dramatic improvement in the survival rate with advances in chemoradiotherapy [2]. However, the latter has led to an increase in radiation induced malignancy (RIM), with the cumulative risk increasing steadily with time [3]. The condition is also described as 'secondary' malignancy, implying a cancer triggered by the initial therapy. The overall incidence of post radiation sarcoma in patients surviving longer than 5 years is 0.1%, with the prognosis being worse than sporadic tumors [4]. The latter is attributed to the delay in diagnosis often making them unresectable, older age at presentation, with a latency upto five decades and lack of effective adjuvant treatment [4, 5].

Radiation threshold for RIM is believed to be around 5 Gy. It is presumed to be due to genetic alterations caused by the ionizing radiation, with a higher risk in young smokers

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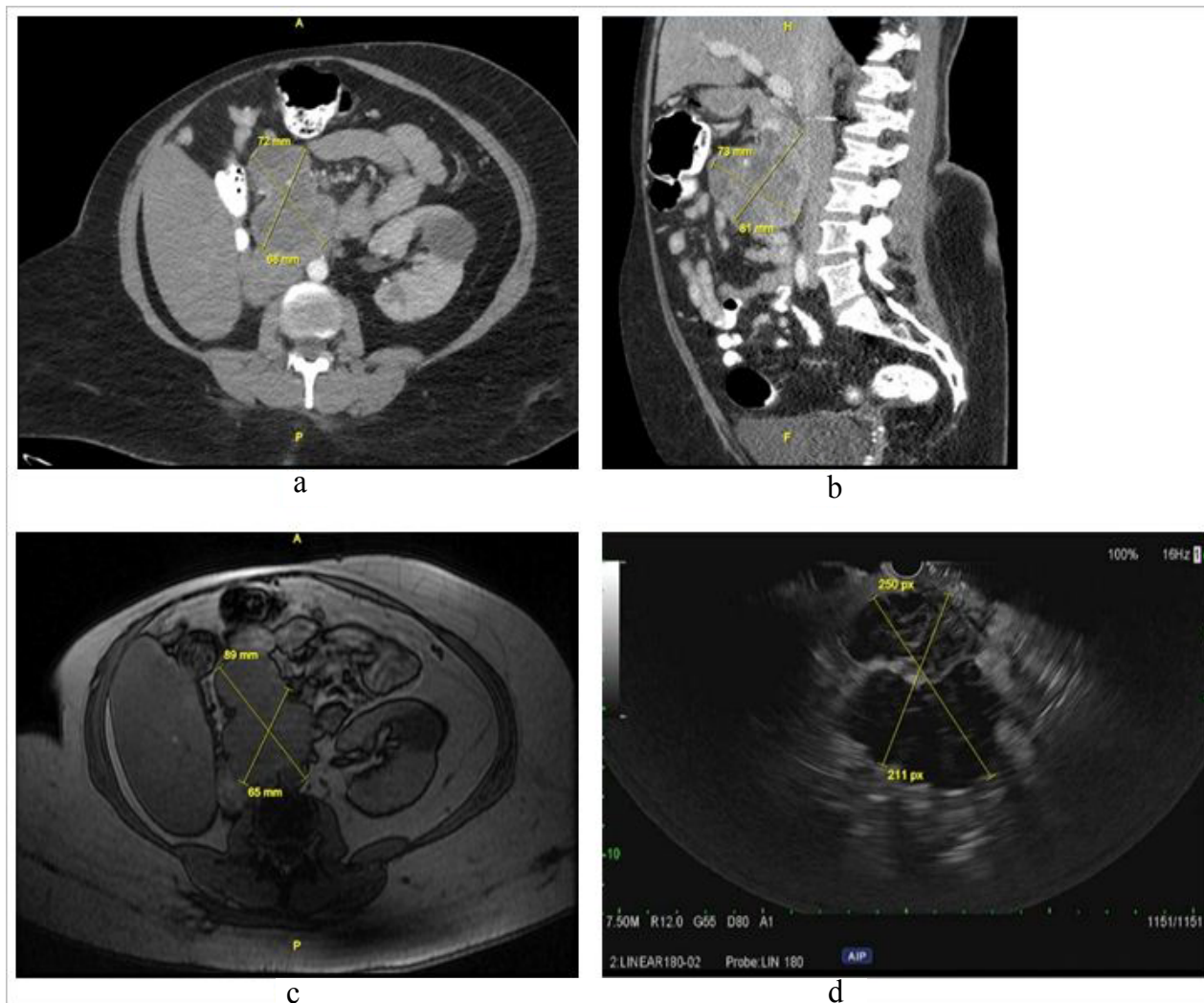


Figure 1. Radiologic Findings. a, b- CT, c-MRI and d- EUS.

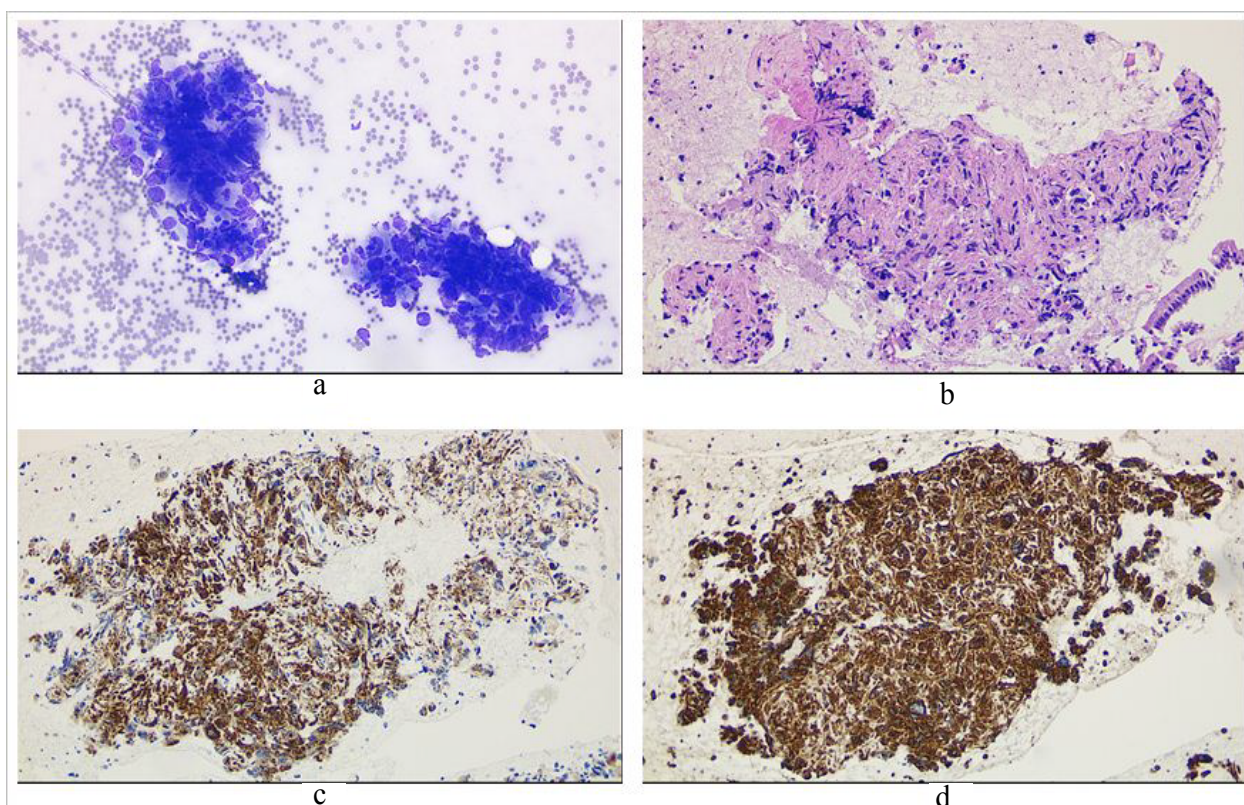


Figure 2. Cytopathology and Immunohistochemistry.

and with the use of radiosensitizing chemotherapeutic agents [6, 7]. The diagnosis of RIM was made in accordance with the original criteria laid out by Cahan et al in 1948 [5]. This includes 1) different histologies for the primary and secondary cancer, 2) secondary cancer within the irradiated area and 3) latency of over 5 years for the secondary cancer. The search for the signature mutation of RIM is ongoing [8]. Precision targeting has minimized the risk of RIM.

Sarcomas arise from the mesenchymal tissue and are often slow growing and hence large at the time of diagnosis. These were the first solid tumors to be linked to ionizing radiation, bone and soft tissue being most susceptible. There seems to be a linear dose relationship with high-dose fractionated radiation exposure over 10Gy at least till 40Gy [8]. Genetic conditions like Nijmegen Breakage Syndrome, Li-Fraumeni syndrome, Retinoblastoma and Costello syndrome are extremely sensitive to the oncogenic effects of radiation.

Although sarcoma involving the gastrointestinal system following radiotherapy for Wilms has been reported [7, 9, 10], this is the first report of a pancreatic sarcoma occurring in this context. They do not have any pathognomonic radiologic characteristic and have to be differentiated from metastatic sarcomas and invasion from nearby organs. Diagnosis relies heavily upon cytology and immunohistochemistry. The exact nature of the stromal malignancy was confirmed by EUS-FNA cytology, avoiding the need for increasingly morbid, invasive tissue acquisition procedures in a patient with advanced disease [9]. This case report adds to the limited scientific information available today on this specific entity.

At present, there are no accepted guidelines for the surveillance of long term survivors of radiotherapy.

Ongoing conscious effort is needed to minimize radiation exposure, given the high fatality rates associated with RIM.

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## Conflicting Interest

The authors had no conflicts of interest

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