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EXTRA SKELETAL PANCREATIC EWING SARCOMA CASE REPORT

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INTRODUCTION

Pancreatic extra-skeletal Ewing sarcoma (EES) is an aggressive mesenchymal soft-tissue tumor that accounts for 1% of all soft-tissue sarcomas and 2% to 5% of malignant bone neoplasms. Ewing sarcoma (ESB) was initially characterized by James Ewing in 1921.[1] It can be identified when the soft-tissue tumor has a sarcomatous appearance with an osteoid or cartilage matrix but no underlying bony connection.[2] With a median age of 66 years, this uncommon tumor affects men and women equally in the fifth and sixth decades of life, in contrast to osteosarcoma of the bone, which primarily occurs in the first two decades.[3-5]

Radiation treatment and trauma are well-known risk factors for the development of around 10% of EES patients.[6] Roughly 30% of all Ewing Sarcoma Family Tumors (ESFTs) are extraosseous; they often appear in the soft tissues of the trunk or extremities, but they can also occur in the biliary system, stomach, esophagus, and oral cavity. There are just 27 examples of the pancreas being documented globally, making it a very rare extraosseous site.[7,8] We report an instance of this illness.

CASE DESCRIPTION

A 13-year-old male football player presented with epigastric pain, vomiting, mild fever and tender abdomen in March 2019, complete workup and imaging done for him, Hb 10g/L, WBC 13 × 10^9/L, Normal renal function test (RFT), liver function test (LFT), carcinoembryonic antigen (CEA).

A contrast-enhanced CT scan revealed a mass with central necrosis connected to the stomach raising suspicion of a gastrointestinal stromal tumor (GIST) causing gastric outlet obstruction (GOO). Exploratory laparotomy was performed, during which a cystic mass was found in the body and tail of the pancreas containing infected fluid and adhering to the posterior wall of the stomach. The intraoperative diagnosis was pancreatic pseudocyst. Fluid was aspirated and sent for cytology, culture and sensitivity testing. A cystogastrostomy was performed after obtaining a biopsy from the cyst wall.

Postoperatively, the patient showed clinical improvement. Histopathological examination of the cyst wall confirmed a pancreatic pseudocyst, and cytology of the aspirated fluid showed inflammatory changes without malignant cells. Further investigations conducted to determine the underlying cause of the pseudocyst and pancreatitis. Based on history, clinical signs and laboratory findings, a diagnosis of traumatic pancreatitis was established.

During the follow up, the pseudocyst gradually decreased in size. At five months postoperatively, a transabdominal ultrasound (TAUS) complete resolution of the cyst. However, at seven months' post-surgery, the patient experienced recurrence of mild epigastric pain unrelated to meals, a transabdominal ultrasound (TAUS) revealed a recurrent 4 cm pancreatic cyst. A subsequent CT scan showed well defined non enhancing pancreatic cyst with internal debris **(figure 1).** Over the following two months, serial imaging showed a gradual increase in cyst size. At nine-month post-surgery, TAUS revealed multiloculated hypoechoic collection about 69 x 69 x 57 mm at the pancreatic tail, with concomitant gallstones for the first time. magnetic resonance cholangiopancreatography MRCP confirmed no connection between the main pancreatic duct (MPD) the cyst, ruling out a pseudocyst secondary to pancreatic ductal disruption **(figure 2)**. He was referred for endoscopic ultrasound (EUS) –guided evaluation, but he refused the procedure. Due to the Covid-19 pandemic,

follow up was interrupted and he returned back 1year post-surgery with recurrence of abdominal pain, a new TAUS revealed gall stone and pancreatic pseudocyst with no solid components, similar in size to previous findings, OGD done for him and showed that cystogastrostomy site was closed, EUS requested but because of COVID19 pandemic and unavailable EUS in our city, we decided second operation for him which includes Cholecystectomy, Reux en y cystojejunostomy and biopsy from the cyst wall for histopathology, this procedure done without complete resection because there was no intraoperative frozen section facility due to the pandemic. Postoperative result of the cystic fluid aspiration showed high CEA, LDH and amylase. HPE of the cyst wall revealed features of poorly differentiated carcinoma most possibly neuroendocrine tumor (NET), and staging CT scan **(figure 3)** showed thick irregular inner wall cystic mass measuring (5.3 x 5.9)cm involving the anterior aspect of the distal pancreas at the region of the tail and distal body, showing enhancing nodular solid component mainly involving the anterior wall between 10-3 o'clock in clockwise direction, associated with mild degree MPD dilation in the distal pancreas, but normal pancreas proximally and normal proximal MPD, no distal metastasis, oncologic consultation was done recommend to proceed with surgery then adjuvant chemotherapy, third operation was planned a week later to achieve oncologic resection, the procedure involved distal pancreatectomy with partial gastrectomy and splenectomy **(Figure 4)** and R0 resection margin was achieved.

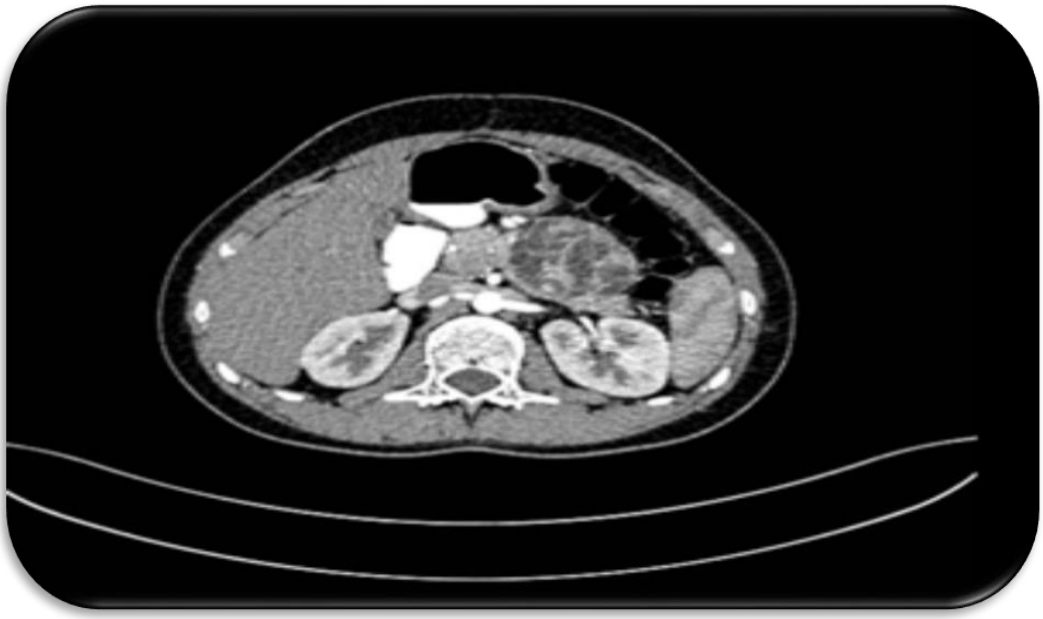

Figure 1: Contrast-enhanced CT showed a large well defined non enhancing pancreatic cyst with internal debris.

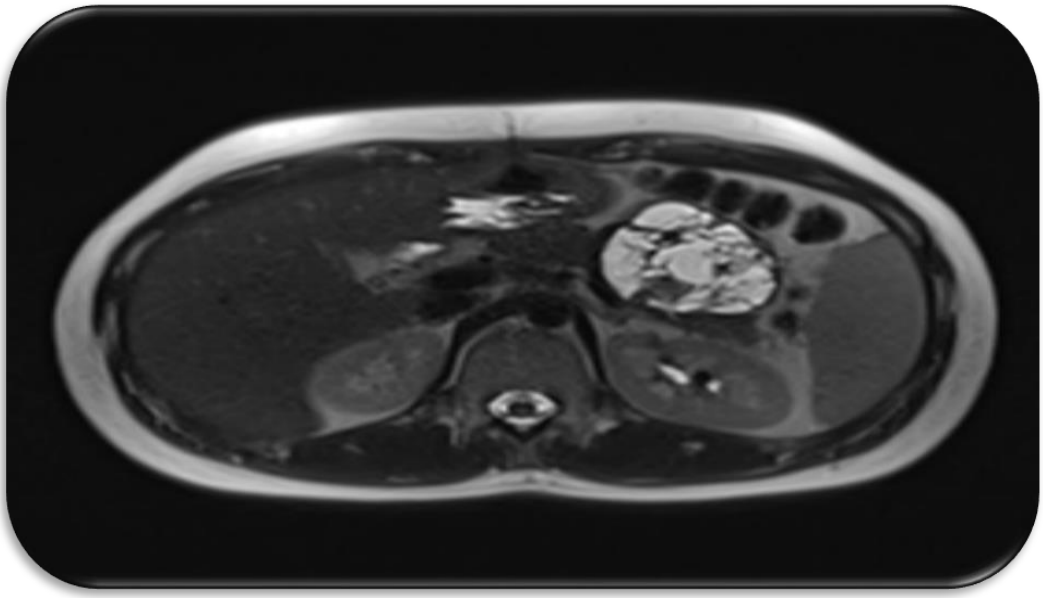

Figure 2: MRI with MRCP showed a multiloculated cyst with no connection to the main pancreatic duct.

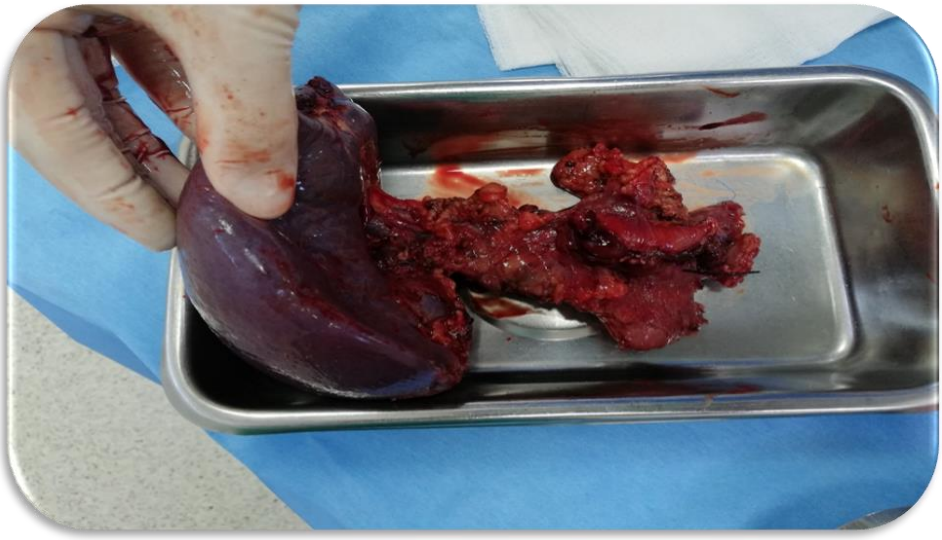

Figure 3: Post-operative staging CT scan showed no distant metastases apart from thick irregular inner cystic distal pancreatic mass.

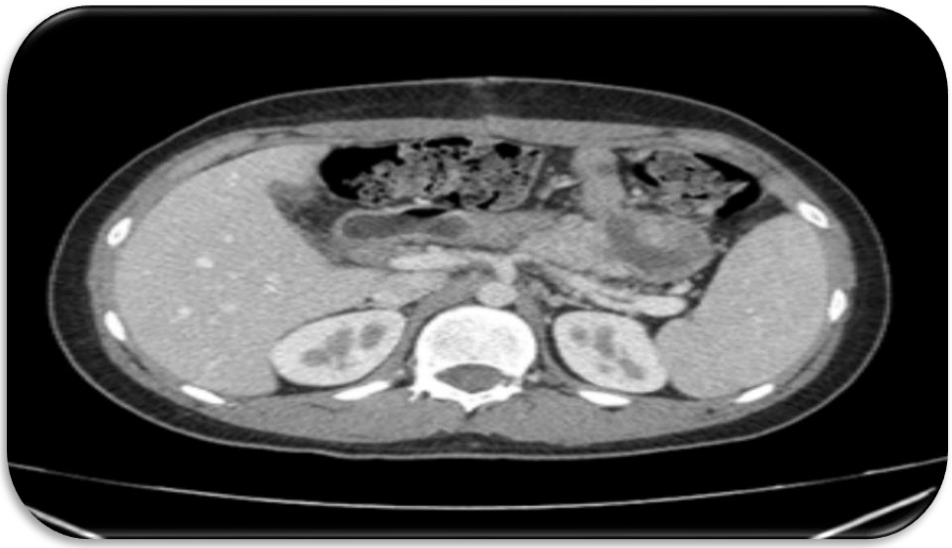

Figure 4: Distal pancreatectomy with partial gastrectomy and splenectomy.

The microscopic examination revealed about 5-10% of the tumor cells showing granular cytoplasmic staining for Synaptophysin & CD56 while about 10-15% showed nuclear staining features of poorly differentiated NET, we did an MDT discussion and reviewed the slides of biopsy and immunohistochemically with another histopathologist the report showed high-grade malignant small blue cell tumor involving the stroma of the pancreas with scarring and preservation of the intervening epithelial structures, the following stains were negative, CD34. BCL-2, PALP, OCT3/4, CD45, Ber-Ep4, CDX-2, β-Catenin. ER, PR, CD56, Chromogranin, Synaptophysin, CD10, CK7, CEA and NSE, this immunoprofile conclusively excludes neuroendocrine carcinoma, the conclusion was high-grade malignant pancreatic neoplasm, with morphological and immunohistochemical features of Sarcoma, consistent with extra skeletal Ewing sarcoma **(figure 5, 6 and 7)** this changed the whole scenario, we were obliged to take another histopathologist opinion which was again the same diagnosis of pancreatic Ewing sarcoma, the patient was referred to oncologist and he received chemotherapeutic agents VAC (Vincristine, Adriamycin and Cyclophosphamide: Ifosfamide and Etoposide) for 8 cycles instead of 17 because of upper gastrointestinal bleeding and hemoptysis, at after resuscitation and treating duodenal ulcer, a PET scan was done showed nodular lesion close relation to the distal aspect of pancreatic remnants along the surgical sutures that indicating recurrence of the tumor (6 month post definitive surgery), the patient refused more chemotherapy, after discussion with oncologist we decide to start SRS (stereotactic Radiosurgery) outside the country. After finishing the radiotherapy course, follow up PET scan showed no remaining disease. Till second year post surgery there was no recurrence indicating complete cure.

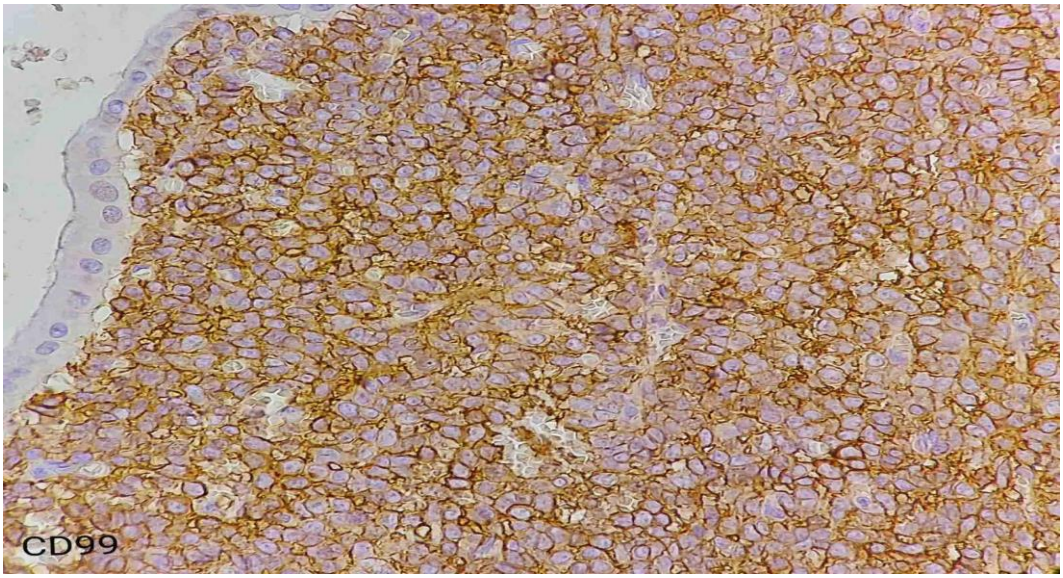

Figure (5): Slides of IHC staining CD99 showed diffuse membranous staining.

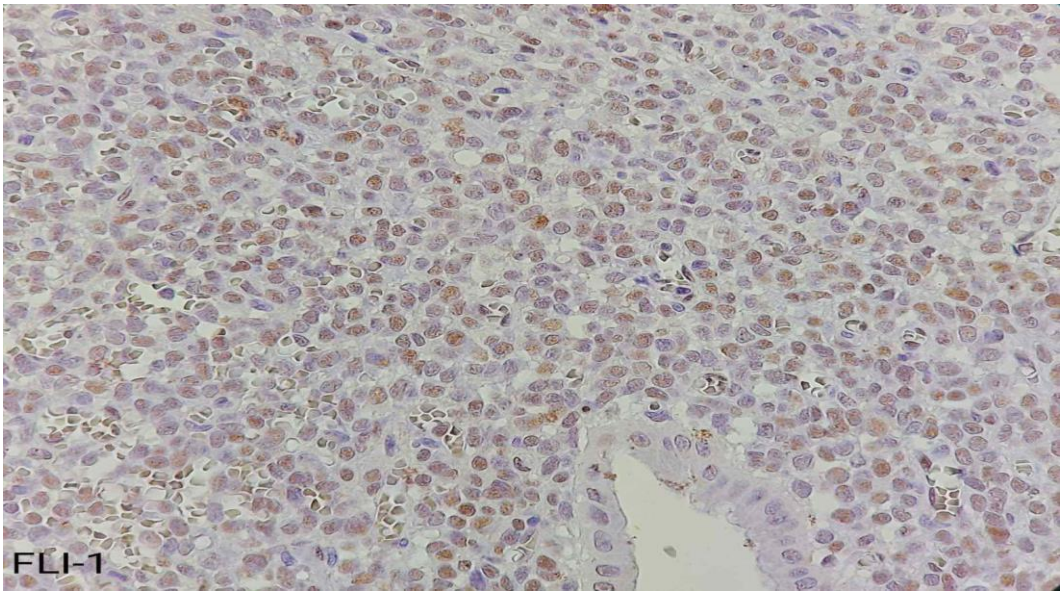

Figure (6): Slides of IHC FLI-1 staining showed nuclear staining.

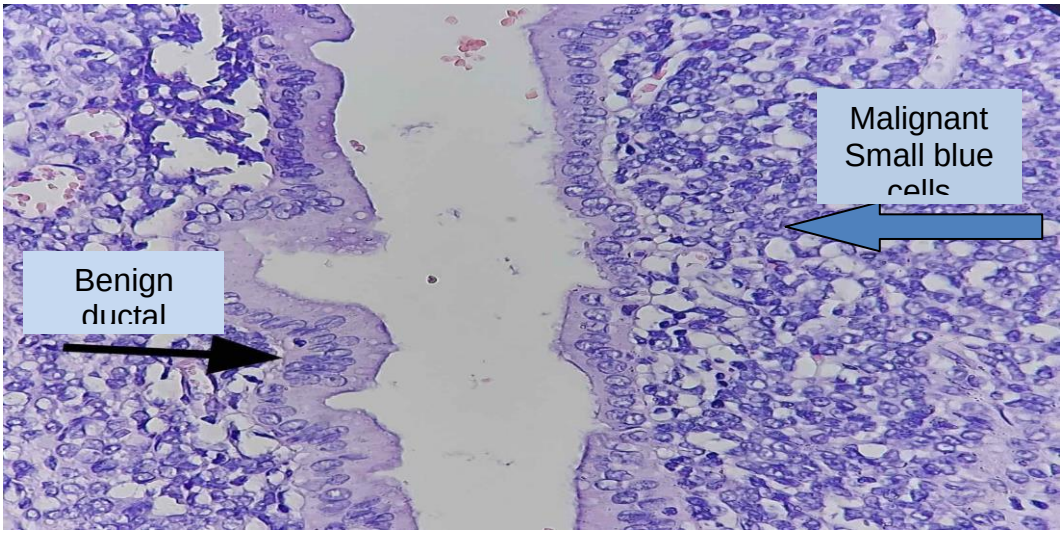

Figure 7: Slides of IHC showed blue cells surrounds pancreatic duct.

DISCUSSION

Since there is less data on the prevalence and clinical features of EES in children, a 13-year-old with gallstones, epigastric discomfort, and a recurring cystic lesion in the pancreatic tail was more likely to have a pancreatic pseudocyst. According to Kosmahl et al.[8], a significant proportion of pancreatic ductal adenocarcinomas and their variations exhibit cystic characteristics, and as such, have to be taken into account when making a differential diagnosis of pancreatic cystic neoplasms. Furthermore, not every one of the cystic formations they saw was malignant. They could also be non-cancerous alterations like inflammatory pseudocysts and retention cysts. According to other reports, the majority of cystic pancreatic ductal adenocarcinomas that have been documented in the literature are hypocystic tumors that are poorly differentiated.[9–11] However, the radiologist considered the multilobulated cyst in our case to be a pseudocyst because it lacked any solid components or alarming characteristics. We suspected recurrent pancreatic pseudocyst since the patient had a history of pancreatitis, which was treated by cystogastrostomy. However, since our patient is a youngster and the recurrence incidence of pancreatic pseudocyst is 10–14%, I was unsure if malignancy was present. Additionally, tumor markers were not increased. Because TAUS also showed a gallstone, I originally thought there could have been a pancreatic pseudocyst recurrence. Due to the family's refusal of EUS, I was unable to make a firm diagnosis before surgery. Since an intraoperative frozen section was not accessible because of the COVID19 pandemic, I thus scheduled a cholecystectomy and Roux en Y cystojejunostomy after obtaining fluid for amylase, CEA, cytology, and wall biopsy for histology. The cyst fluid's postoperative investigation revealed elevated levels of amylase, CEA, and malignant cyst wall. No distant metastases were detected by staging CT, and surgery was decisive. Since surgical resection has a significant impact on the prognosis of the disease, it is imperative to detect these instances as soon as possible.

The availability of EUS would have been very important in this case, it was not available in my city and the COVID19 pandemic impacted the management approach.

CONCLUSION

Ewing sarcoma of the pancreas is rare but should be in the differential diagnosis of pancreatic lesions, particularly in case of recurrent pancreatic pseudocyst. EUS is crucial and intraoperative frozen section for the cyst wall in recurrent pancreatic pseudocyst is mandatory to exclude carcinoma if the EUS biopsy inconclusive and there are no radiologic signs of malignancy.

Ethics statement

The Ethics Committee of the Rizgary Teaching Hospital in Erbil Province gave its approval to the human subjects' study. The research were carried out in compliance with institutional norms and local laws. According to national laws and institutional standards, neither the participant nor their legal guardians or next of kin were needed to provide written informed permission for participation. Any potentially identifying photographs or data featured in this article were published with the individual's written informed consent.

Author contributions

Dr. Mohammed Niyazi Gheni : Writing – original draft, Writing – review & editing.

Dr. Janan wadea hurmiz Zora and Dr. Kawa Mohammed Mawlood Bajalan: Conceptualization, Data curation, Supervision, Writing – review & editing. Dr. Mohammed Niyazi Gheni: Methodology, Visualization, Writing – review & editing.

Dr. Mohammed Niyazi Gheni: Data curation, Formal analysis, Writing – review & editing.

Dr. Mohammed Niyazi Gheni and Dr. Janan wadea hurmiz Zora: Conceptualization, Project administration, Writing – review & editing.

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Conflict of interest

The authors declared that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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