Metastasis of Ewing Sarcoma to the Pancreas: An Exceedingly Rare Case report and Literature Review

Rose Al Bacha[1], Karam Karam[1], Claude Ghorra[2], Bassam Abboud[2], and Joseph Bou Jaoude[2]

[1]University of Balamand
[2]Lebanese University

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Rose Al Bacha[1*], Karam Karam[2], Claude Ghorra[3], Bassam Abboud[4], Joseph Bou Jaoude[5]

[1] Clinical Educator, Department of Gastroenterology, University of Balamand, El Koura, Lebanon. Email: Rosealbacha@outlook.com *CORRESPONDING AUTHOR

[2] Department of Gastroenterology, University of Balamand, El Koura, Lebanon.

[3] Professor of anatomical pathology, Lebanese University, Lebanon.

[4] Professor, Department of General Surgery, Geitaoui Hospital, Lebanese University, Lebanon.

[5] Professor of Gastroenterology and Hepatology, Lebanese University, Lebanon.

Key Clinical Message: Physicians should have a high index of suspicion for a pancreatic metastasis when approaching a patient with Ewing sarcoma presenting with pancreatitis. Pancreatic metastasis from Ewing sarcoma is an exceedingly rare condition that physicians should be aware of to ensure prompt diagnosis and timely management.

Keywords : Ewing Sarcoma; Pancreatitis; Pancreatic Metastasis.

1-Introduction

The Ewing sarcoma family of tumours (ESFT) represents a group of classic Ewing's sarcoma of bone, extra skeletal Ewing's sarcoma (EES), Askin tumours of the chest wall and primitive neuroectodermal tumours of bone or soft tissues[1]. The translocation t(11; 22) (q24; q12), which produces the EWS/FLI1 fusion gene is the genetic bases of ESFT[2]. The tumor cells are an undifferentiated small round blue cell with a low mitotic index[3].

Ewing sarcoma (ES) of the bone is the second most frequent malignant bone cancer in adolescents and young adults after osteosarcoma[4]. EES is an uncommon condition that can appear anywhere in the soft tissues and can alter the cortex of nearby bone. The trunk, extremities, and retroperitoneum are the areas where it occurs most frequently[5].

However, the existence of metastatic disease is the most crucial prognostic marker, and even patients who are diagnosed with locally limited cancer are at significant risk of developing metastatic disease while receiving local therapy[6].

While Ewing sarcoma is primarily a bone cancer, it rarely metastasizes to the pancreas[7]. In this case report, we present a 38-year-old man with cytogenetically confirmed metastatic ES to the pancreas.

2-Case presentation

A 38-year-old male patient presenting for severe epigastric pain radiating to the back of 3-days duration. He described his pain as pressure-like, intermittent with an intensity of 10 out of 10. Of note, the patient was initially diagnosed with EES of the soft tissue on the plantar surface of the left foot (8*3,6cm) with immunochemistry that showed expression of CD99. He underwent surgical resection below knee amputation with biopsy revealing small round blue cell destructing the bone and infiltrating the soft tissue.

3-Methods:

Computed tomography (CT) scan of the abdomen revealing edematous pancreatitis of the tail of the pancreas, corroborating a diagnosis of mild acute pancreatitis. Magnetic resonance imaging (MRI) showed a well circumscribed mass measuring 14 mm present at the junction of the body and tail (Figure 1). It is of high signal intensity on T2 low T1 with minimal internal septal enhancement. It is communicating with the pancreatic duct which distally in the tail is at the upper limits of normal measuring 3 mm.

Endoscopic ultrasound (EUS) showed a 13 mm cystic lesion obstructing the Wirsung duct (figure 2). FNA and core biopsy were positive for a small blue cell tumour and positive for CD99. These findings were consistent with extension of Ewing sarcoma (figure 3).

4-Conclusion and Outcome:

He was subsequently treated with Docetaxel-based chemotherapy. Unfortunately, patient had disease progression, developed lung metastasis with recurrent pleural effusions and died of pulmonary failure thereafter.

5-Discussion

Pancreatic metastasis is rare, accounting for only 2% of all pancreatic cancer[8]. The primary tumors that metastasize frequently to the pancreas are lung cancer, renal cell carcinoma, breast cancer, melanoma and colon cancer[9].

Ewing sarcoma is a rare bone tumor and depending on whether metastases were present at diagnosis, patients with ESFT have considerably different five-year overall survival rates (OS) ranging from 70% if localized to 9–41% for metastatic disease[10].

Hyma et.al reviewed a 39 cases of ESFT involving the pancreas, with only four of them being metastatic lesions[11]. The 4 cases were younger than 30 years of age, with two of them testing positive for CD 99 and one testing positive for PAS in terms of pathologic features. There is also a male predominance with only one female case.

A review done by Saif et.al showed no gender predilection in Ewing sarcoma /primitive neuroectodermal tumour (ES/PNET) with the majority diagnosed at their teenage years with disseminated disease at the diagnoses[12].

The diagnosis of pancreatic metastases can be made safely and effectively with endoscopic ultrasonography (EUS) and confirming the diagnosis using immunohistochemistry[9]. Out of the 39 cases described in the literature, CD99 is the most frequently reported marker linked to ES. Other related markers that are less precise are synaptophysin, vimentin, and neuron-specific antigen[11].

Radiation, chemotherapy, and surgical resection were used to treat these individuals; however, the prognosis was poor with two resulting deaths from the disease. Similarly, our case was a young male patient with positive CD 99 in terms of pathologic features.

The prognosis for ESFTs has significantly improved after multiagent chemotherapy was added to surgery and/or radiation treatment. Children with localized illness who get combined-modality treatment have a 65% to 70% progression-free survival rate (PFS)[13]. The EURO-EWING 99 trial demonstrated that patients

who received local treatment for both the primary and metastatic disease had a three-year event-free survival rate that was significantly higher compared to patients who only received local treatment for the primary or metastatic disease[14].

Another differential diagnosis in young patients is small cell neuroendocrine carcinoma due to the similarity in morphological characteristics, imaging and immunohistochemistry which can delay the proper therapy. Hence, to confirm the diagnosis, molecular study should be employed.

In conclusion, we present a rare case of ES metastasis to the pancreas in a 38-year-old male with prior diagnosis and treatment of ES of the left foot unresponsive to chemotherapy. Our case emphasizes the rarity of pancreatic ES metastases, since only four previous cases have been documented in the literature. Pancreatic metastasis can be diagnosed with immunohistochemistry stain, with CD99 being the most frequently found marker linked to ES.

Figure 1 legend: Well circumscribed mass measuring 14 mm at the junction of the body and tail of the pancreas (white arrow).

Figure 2 legend: Figure 2a :13 mm cyctic lesion obstructing the Wirsung duct, which is dilated upstream.

Figure 2b: FNA needle 22G inside the lesion.

Figure 3 legend: *Figure 3a: Small round blue cell in H&E stain.*

Figure 3b: Cells showing diffuse membranous expression of CD99 on immunohistochemistry.

Authorship List:

1. Dr. Rose Al Bacha: *Corresponding author* Investigation, Methodology, Writing original draft and supervision.
2. Dr. Karam Karam: Investigation, Methodology, Writing original draft
3. Dr. Claude Ghorra: Investigation, Methodology, Writing original draft
4. Dr. Bassam Abboud: Data curation, investigation
5. Dr. Joseph Bou Jaoude*:* Supervision, writing review & editing

Declarations:

Ethics approval and consent to participate:

Not applicable.

Consent for publication:

Written informed consent was obtained from the patient for publication of

this case report and any accompanying images. A copy of the written consent

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Authors' contributions:

All authors have all contributed as authors to this manuscript in terms of planning, conception, writing and editing various drafts of the manuscript and read and approved the final manuscript.

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