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Case Report

Metastasis of Ewing Sarcoma to the Pancreas: Case Report and Literature Review

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Abstract

Introduction: The Ewing sarcoma family of tumors (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. However,the existence of metastatic disease is the most crucial prognostic marker. While Ewing sarcoma is primarily a bone cancer, it rarely metastasize to the pancreas with 4 reported cases in the literature.

Case presentation: We represent a case of a 38 year old male patient diagnosed with Ewing sarcoma of the bone that developed acute pancreatitis after 6 months. MRI showed a well circumscribed mass measuring 14 mm is present at the junction of the body and tail . FNA and core biopsy was positive for a small blue cell tumor and positive for CD99. These findings were consistent of extension of Ewing sarcoma. Patient was unresponsive to chemotherapy , disease progression and died. . Our case emphasizes the rarity of pancreatic ES metastases, since only four previous cases have been documented in the literature. The necessity of advances in therapy is crucial.

Keywords: Ewing; Sarcoma; Pancreas; Metastasis

Introduction

The Ewing sarcoma family of tumors (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]. Whereas EES is an uncommon condition that can appear anywhere in the soft tissues but 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 metastasize to the pancreas [7]. In this case report, we present a 38-year-old man with cytogenetically confirmed metastatic ES to the pancreas.

Case Presentation

A 38-year-old male, was initially diagnosed with EES of the soft tissue on the plantar surface of the left foot (8*3,6cm) with immunochemistry showed expression of CD99. He underwent surgical resection below knee amputation with biopsy showed small round blue cell destructing the bone and infiltrating the soft tissue.

After 6 months he developed mild acute pancreatitis with CT scanner of the abdomen showed edematous pancreatitis of the tail of the pancreas. MRI (Figure 1) showed a well circumscribed mass measuring 14 mm is present at the junction of the body and tail. 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.

Echo endoscopic ultrasound showed a 13 mm cystic lesion obstructing the wirsung (Figure 2). FNA and core biopsy was positive for a small blue cell tumor and positive for CD99. These findings were consistent of extension of Ewing sarcoma (Figure 3).

He was then treated with chemotherapy Docetaxel basedbut unfortunately patient had disease progression, developed lung metastasis with recurrent pleural effusion and died for pulmonary failure.

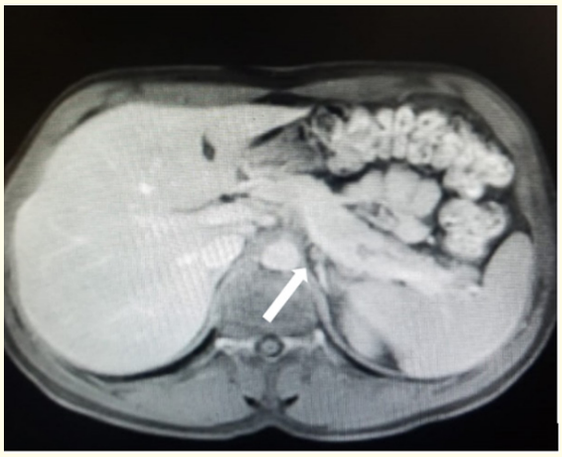

Figure 1: Well circumscribed mass measuring 14 mm at the junction of the body and tail of the pancreas (white arrow).

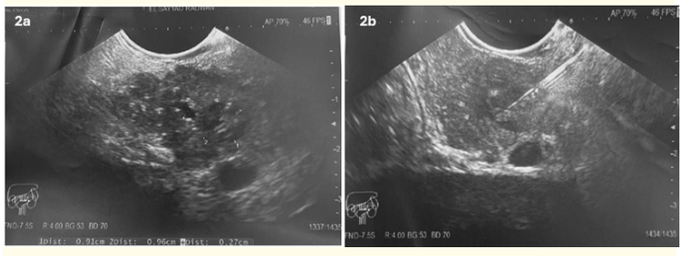

Figure 2: a: 13 mm cyctic lesion obstructing the wirsung which is dilated upstream, b: FNA needle 22G inside the lesion.

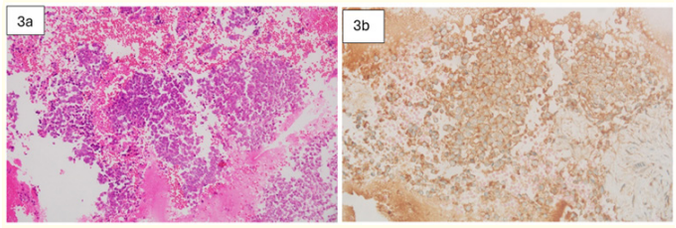

Figure 3: a: Small round blue cell in H and E stain, b: Cells showing diffuse membranous.

Discussion

Pancreatic metastasis are 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) with 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 in death from the disease. Similarly our case was young male patient with positive CD 99 in terms of pathologic features.

The prognosis for ESFTs has significantly improved after multi-agent chemotherapy was added to surgery and/or radiation treatment. Children with localized illness who get combined-modality treatment had a 65% to 70% progression-free survival rate (PFS) [13]. While 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 patient 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 obtained [15].

Conclusion

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.

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