Pancreatic Metastasis of Alveolar Rhabdomyosarcoma Presenting as Acute Pancreatitis

پرستو ملایی توانا

SBMU

Case presentation

- **A 14-year-old girl, known case of alveolar rhabdomyosarcoma (RMS) of head and neck (zygomatic region accompanied by cervical lymphadenopathies)**
- **She was receiving chemotherapy for intermediate-risk RMS with standard VAC for the last eight months**
- **She was admitted with severe epigastric unrelenting pain.**

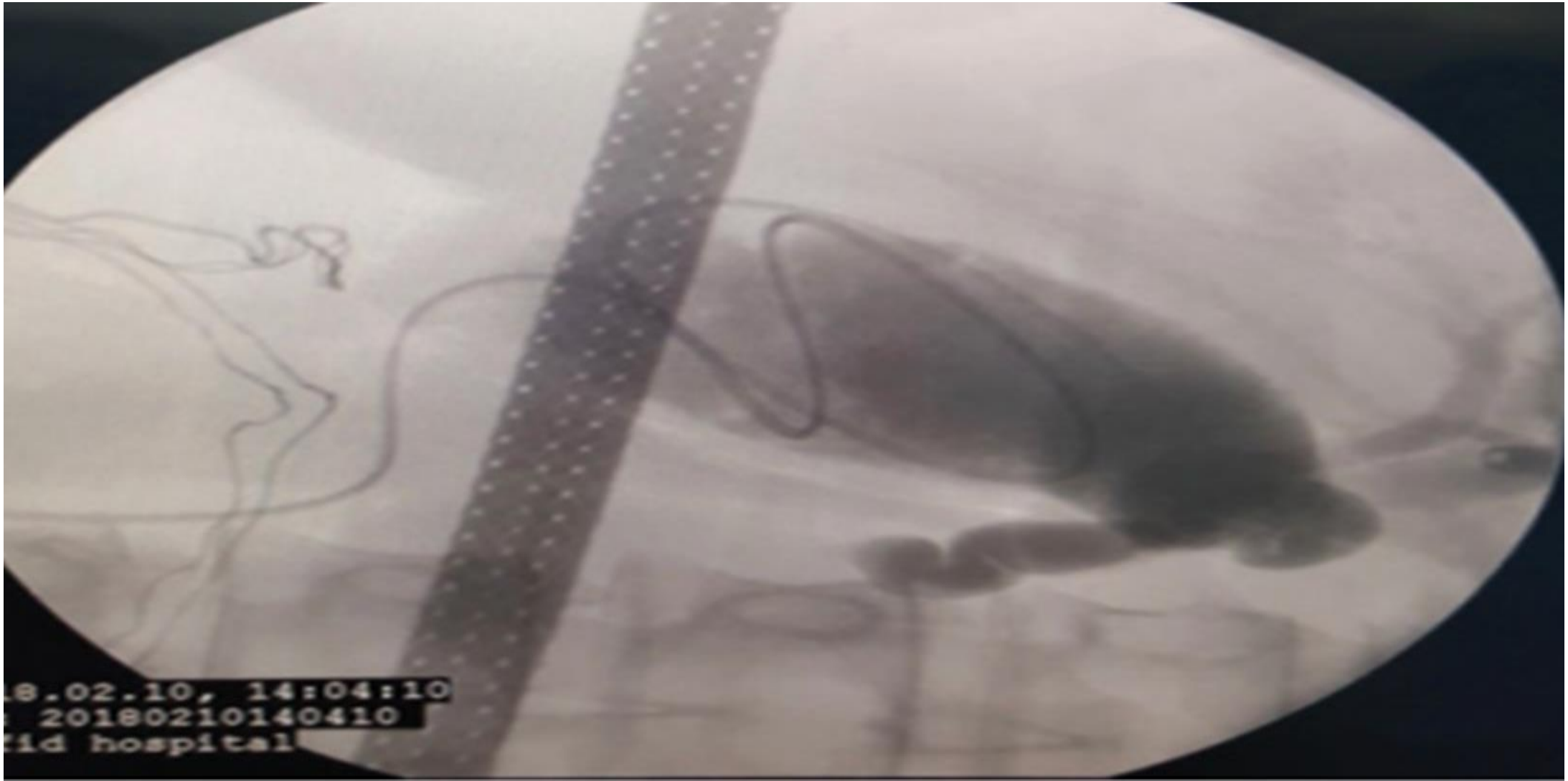
Case presentation

- **On admission, she was mildly icteric and afebrile.**

- **Ultrasound and CT scan: minimally dilated intra and extra hepatic and common bile ducts. Head and neck of the pancreas had increased size**

- **She was diagnosed with “acute pancreatitis” due to associated signs and symptoms and increased levels of amylase and lipase to more than three times.**

Magnetic resonance cholangiopancreatography :diffuse enlargement of the pancreas with heterogeneous signal intensity suggestive of diffuse pancreatitis or infiltrative process along with obstruction at the distal end of the common bile duct



Case presentation

- **The patient underwent laparotomy which a tumoral mass in the head of the pancreas was detected.**

- **Histopathology : metastatic alveolar RMS.**

EUS Diagnosis of a Primary Pancreatic Metastasis of Alveolar Rhabdomyosarcoma

University of Utah School of Medicine, Huntsman Cancer Center. Salt Lake City, UT, USA

- **An 18-year-old boy with a history of right orbital alveolar RMS has also been reported who developed an episode of pancreatitis similar to our case.**
- **His CT-scan demonstrated acute pancreatitis with no mass lesion.**
- **Two months later, he developed abdominal pain and an MRI indicated a 6.4 cm mass in the body and tail of the pancreas. Endoscopic ultrasound guided FNA in that case confirmed the diagnosis of alveolar RMS metastatic to the pancreas.**

Discussion

- **RMS represents the most common soft tissue sarcoma in children, with an incidence of 4.3 new cases per million children and adolescents younger than 20 years.**

- **Alveolar RMS is associated with an aggressive course and a poor outcome. Metastatic disease is thought to involve most commonly the lungs, bone, bone marrow and lymph nodes.**
- **RMSs have a propensity to involve unusual sites such as the breast, testes and subcutaneous tissues.**

Discussion

- **Acute pancreatitis (AP) is a rare manifestation of pancreatic cancer (PC).**
- **The relationship between pancreatitis and PC remains is not well defined.**
- **Pancreatic metastases are uncommon and are only found in a minority (3-12%) of patients with widespread metastatic disease at autopsy.**

ALVEOLAR RHABDOMYOSARCOMA An Analysis of 110 Cases F. M. ENZINGER,MD, AND M. SHIRAKI,MD

From the Armed Forces Institute of Pathology and the Veterans Administration Special Reference Laboratory for Pathology at the AFIP, Washington, D. C.

Cancer
An International Interdisciplinary Journal of the American Cancer Society

- From 110 cases of ARMS Autopsy data were available in 57 patients
- Most common metastatic sites : lymph nodes (74%),the lungs and the pleurae(74%) ,the pancreas (67%),and the bones (51%).
- Less frequently metastatic sites : heart, the adrenals, the testes, the liver and kidneys, and the spleen
- The cause of the exceptionally high incidence of metastasis to the pancreas remains unexplained

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- **In spite of this high number, the presence of pancreatic metastases in RMS has been rarely reported in the radiology literature .**
- **This discrepancy between pathology and radiology observations may be due to different evaluated patient populations (selected patients with fatal outcome in the cited pathology report versus all patients with RMS evaluated on imaging studies), limited evaluations of the pancreas with standard imaging protocols and limited sensitivity of unenhanced CTs for the detection of pancreatic metastases among other parenchymal lesions.**

Unusual association of alveolar rhabdomyosarcoma with pancreatic metastasis: emerging role of PET-CT in tumor staging

Pediatr Radiol (2010)

- **The Committee on Human Research at three major pediatric oncology centers approved this retrospective evaluation. Pediatric radiologists at UCSF Children's Hospital, Hospital for Sick Children, which is affiliated with the University of Toronto, and St. Jude Children's Research Hospital reviewed their records of the last 11 years (1998–2009) and identified eight patients with biopsy proven alveolar RMS and pancreatic metastases out of a total of 71 cases.**

Unusual association of alveolar rhabdomyosarcoma with pancreatic metastasis: emerging role of PET-CT in tumor staging

Pediatr Radiol (2010)

- **Four of these presented at diagnosis and four with disease recurrence.**
- **In recurrent disease, the duration between the diagnosis of the primary tumor and pancreatic metastases varied from 8 months to 6 years (mean±SD: 2.38±2.49 years).**
- **In all patients who received PET scans, pancreatic metastases showed a marked FDGuptake, but had variable detectability with CT.**
- **Pancreatic metastases were not associated with certain primary tumor locations or presence of other metastases, mandating an evaluation of the pancreas in all cases of ARMS.**

Suggestion

- **Our case was the first case of a teenage girl with head and neck alveolar RMS with pancreatic metastasis who was presented as acute pancreatitis.**

- **We suggest that in patients with ARMS; in follow-up, pancreas and associated symptoms should be taken seriously in account.**

با تشکر از توجه شما