Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(2):114-117 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2016.v04i02.017

Unusual Pancreatic Mass in a Young Adult: Metastases from Alveolar Rhabdomyosarcoma

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Abstract: Obstructive jaundice in older children can be associated with multipl primary and secondary diseases. As a rare entity, pancreatic neoplasms may lead to biliary obstruction in pediatric population. Here, a case of a 15 year-old boy presented with jaundice was evaluated and discussed. In the presence of a jaundice in childhood, pancreatic mass should be included in the differential diagnosis particularly pancreatic metastases from rhabdomyosarcoma should be kept in mind. In diagnosis of rhabdomyosarcoma careful and active evaluation of pancreas should be performed.
Keywords: jaundice, pancreas, metastasis, rhabdomyosarcoma

INTRODUCTION:

Multipl causes of jaundice in older children can be divided into primary diseases of the hepatocytes and biliary tracts. Hepatitis and biliary atresia is the most common cause of non-obstructive jaundice whereas, obstructive jaundice is mainly related with neoplasms , benign strictures or stone diseases in childhood. Pancreatic masses may be a rare cause of obstructive jaundice in pediatric population however they commonly present as primary pancreatic tumours such as pancreatoblastoma. Pancreas is a rare site for metastases in pediatric population thus a few number of cases with pancreatic metastases from rhabdomyosarcoma was reported [1-5].

Rhabdomyosarcoma (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 [6]. RMS originates from striated muscle cells or their mesenchymal precursors and occurs in a large variety of locations, such as the head and neck, pelvis and extremities [7]. Histologically, RMS can be classified as either embryonal (60%), alveolar (20%), undifferentiated or miscellaneous subtypes [8]. Alveolar RMS is associated with an aggressive course and a poor outcome . Metastatic disease is thought to involve most commonly the lungs, bone and lymph nodes [9] and rhabdomyosarcomas have a propensity to involve unusual sites such as the breast, testes and subcutaneous tissue [10]. Additionally, isolated cases of patients with alveolar RMS and pancreatic metastases can be found in the radiology literature [2, 3, 7, 11] . Symptoms and presentation of patients depend on tumor location. A 15 year-old boy presented with jaundice was evaluated and discussed in this case report radiologically.

CASE REPORT

A 15-year-old boy presented with jaundice, abdominal pain and left inguinal swelling to Marmara University, Pediatric Onchology deparment. In physical examination, he had yellow sclera/skin; and marked left inguinal swelling. Laboratory test results showed elevated aspartate aminotransferase, alanine aminotransferase, serum gamma glutamyl transferase , and alkaline phosphatase levels. Total bilirubin was 6.9 mg/ dl and conjugated fraction was 4.9 mg/dl. Abdominal ultrasonography demonstrated dilated intra and extrahepatic bile ducts and an exactly 3 cm hypoechoic mass located in the pancreatic head. There was also an irreguler heterogenous hypoechoic solid mass in the left inguinal subcutaneous region consistent with malignancy. Contrast-enhanced whole body MRI using body coil (Verio 3.0 T field strength; Siemens, Germany) as a further radiologic evaluation was performed for characterization of pancreatic and inguinal mass. MRI demonstrated 32 x 37 mm encapsulated pancreatic mass hypointense on T1-weighted images, hyperintense on T2-weighted images and heterogenously enhancing on dynamic images (Figure 1A-B). Pancreatic mass was leading to biliary obstruction at the level of distal choleduct and pancreatic duct was dilated (choleduct: 11 mm, pancreatic duct:4 mm) (Figure1C). Paraaortic lymphadenopathies and enhancing bone masses indicative of metastasis also exist on MRI.

Additionally, MRI showed a heterogenously enhancing 92 x 38 x 70 mm solid mass in the left inguinal region extending into the pelvis and scrotum (Figure 2). Excisional biopsy from the inguinal mass is performed due to its availability and hystopathologic evaluation revealed alveolar RMS. In view of pathological and radiological findings, a diagnosis of metastases of alveolar RMS was considered for pancreatic mass. A chemotheraphy regimen with cyclophosphamide and vincristine was performed. During chemotherapy, mid-term evaluation (6th month) revealed that pancreatic mass disappeared; and left inguinal bulk significantly regressed (Figure 3).

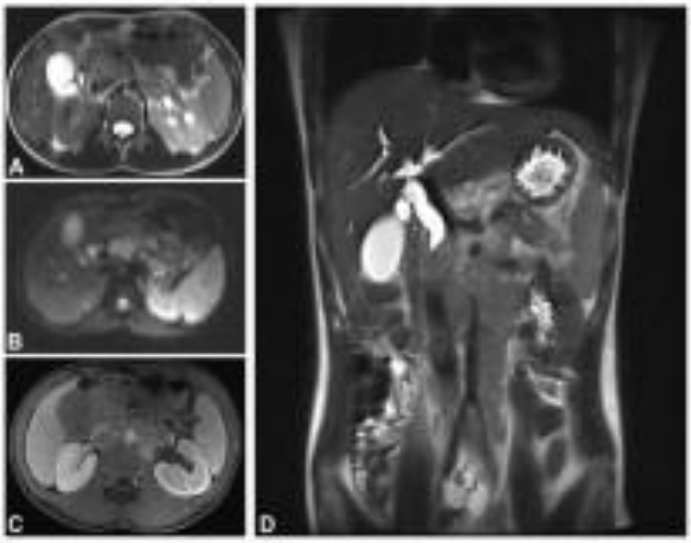

Fig-1: There is a mass in the pancreatic head, hyperintense on axial T2W MRI images (A), restricted in DWI images (B) heteregenously enhancing (C) and causing a biliary obstruction as shown in the coronal T2W images (D).

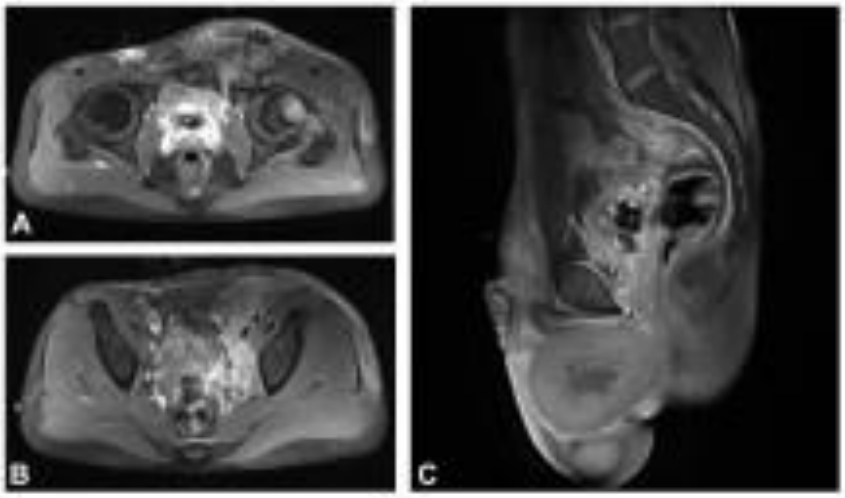

Fig-2: Contrast enhanced abdominal MRI shows heterogenously enhancing left inguinal mass (A) extending into pelvis (B) and scrotum (C).

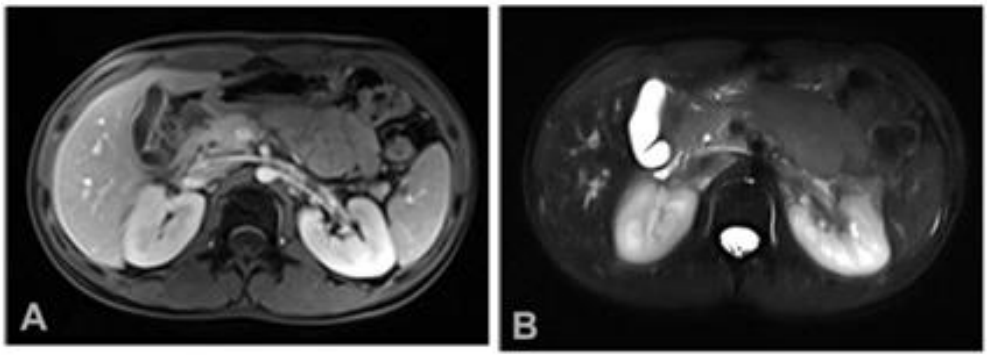

Fig-3: As shown on contrast enhanced axial T1-weighted (A) and axial T2-weighted images (B)pancreatic mass totally regressed after chemotheraphy regiman.

DISCUSSION

Jaundice is an uncommon symptom in childhood. It is usually associated with benign diseases such as hepatitits and biliary atresia which leads to non-obstructive jaundice. Stone disease is the most common etiology of obstructive jaundice in young adults. However, as a rare entity, pancreatic neoplasms may lead to biliary obstruction in pediatric population.

Multiple institutional reviews in the literature yielded a relatively small number of pancreatic neoplasms as a cause of obstructive jaundice. Only a small subset of which are malignant and which mostly present as primary pancreatic tumours. Pancreatic involvement with tumors arising from other organs generally occurs by direct extension of tumor from a contiguous organ, such as neuroblastoma invasion. However, metastases to pancreas from primary malignancies is rarely defined in the literature especially from leukemia and lymphoma [5]. Rhabdomyosarcoma present with pancreatic metastases leading to malign obstructive jaundice is defined in extremely rare case reports in the literature [1-3,11].

Radiologic features of pancreatic metastases were similar with the primary malignancy .Radiologic features of pancreatic metastases from rhabdomyosarcoma is defined as well-circumscribed, nodular, homogenously enhancing masses with hypointensity on T1-weighted and hyperintensity on T2-weighted images as in our case [2,3,11].

Alveolar RMS is a highly aggressive tumor and prognosis is poor. It is encountered commonly in adolescents and young adults [7]. It mostly arises in the neck, trunk, and extremities. Most common metastatic sites are lungs, bone [13] . Enzinger *et al.;* demonstrated that according to an autopsy report of 57 children who died from alveolar RMS, 67 % had pancreatic metastases, making the pancreas the third most common site for metastases [10]. In spite of this high number, the presence of pancreatic metastases in RMS has been rarely reported in the radiology literature [7,12,13,14]. This discrepancy between pathology and radiology observations may be due to different evaluated patient populations , limited radiologic evaluation of pancreas as an uncommon metastatic organ.In these cases, the usual symptoms were nonspecific; such as abdominal discomfort, weight loss and palpable abdominal mass . To the best of our knowledge, only limited number of patients were presented with jaundice during the first diagnosis [1-3,11]. They were commonly detected during the staging of the primary malignancy. In current case jaundice was the first presentation of the patient.

As a conclusion; in the presence of a jaundice in childhood, pancreatic mass should be included in the differential diagnosis particularly pancreatic metastases from RMS should be evaluated .Radiologists and clinicians should be alerted in the diagnosis of RMS for careful and active evaluation of pancreas

Declaration of Interest

The authors report no conflicts of interest.The authors alone are responsible for the content and writing of the paper.

REFERENCES

1. Miller DV, Coffin CM, Zhou H; Rhabdomyosarcoma arising in the hand or foot: a clinicopathologic analysis. Pediatr Dev Pathol. 2004;7:361-369.
2. Khalbuss WE1, Gherson J, Zaman M; Pancreatic metastasis of cardiac rhabdomyosarcoma diagnosed by fine needle aspiration. A case report. Acta Cytol. 1999;43:447-451.
3. Jha P1, Frölich AM, McCarville, Navarro OM, Babyn P, Goldsby R *et al.;* Unusual association of alveolar rhabdomyosarcoma with pancreatic metastasis: emerging role of PET-CT in tumor staging. Pediatr Radiol. 2010;40(8):1380-1386.
4. Yu DC, Kozakewich HP, Perez-Atayde AR, Shamberger RC, Weldon CB; Childhood pancreatic tumors: a single institution experience. J Pediatr Surg. 2009 ;44(12):2267-2272.
5. Chung EM, Travis MD, Conran RM; Pancreatic tumors in children: radiologic-pathologic correlation. RadioGraphics 2006; 26:1211–1238.

6. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A; Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: an analysis of 2, 600 patients. J Clin Oncol. 2009;27(20):3391-3397.
7. McCarville MB, Spunt SL, Pappo AS; Rhabdomyosarcoma in pediatric patients: the good, the bad, and the unusual. Am J Roentgenol. 2000;176:1563-1569.
8. Pappo AS, Shapiro DN, Crist WM; Rhabdomyosarcoma. Biology and treatment. Pediatr Clin North Am 44:953–972.
9. Dagher R, Helman L; Rhabdomyosarcoma: an overview. Oncologist. 1999;4:34-44.
10. Enzinger FM, Shiraki M; Alveolar rhabdomyosarcoma. An analysis of 110 cases. Cancer. 1969;24:18-31.
11. Farah RA, Kamen BA; Parameningeal alveolar rhabdomyosarcoma with an isolated pancreatic metastasis. Pediatr Hematol Oncol. 1999;16:463-467.
12. Volker T, Denecke T, Steffen I, Misch D, Schönberger S, Plotkin M *et al.;* Positron emission tomography for staging of pediatric sarcoma patients: results of a prospective multicenter trial. J Clin Oncol. 2007;25(34):5435-5441.
13. McCarville MB, Christie R, Daw N, Spunt SL, Kaste SC; PET/CT in the evaluation of childhood sarcomas. AJR Am J Roentgenol. 2005;184(4):1293-1304.
14. Tateishi U, Hosono A, Makimoto A, Nakamoto Y, Kaneta T, Fukuda H *et al.;* Comparative study of FDG PET/CT and conventional imaging in the staging of rhabdomyosarcoma.Ann Nucl Med. 2009;23(32):155-161.