

CASE REPORT

EUS Diagnosis of a Primary Pancreatic Metastasis of Alveolar Rhabdomyosarcoma

Ryan C Vanwoerkom¹, Joel Bentz², Lei Chen¹, Douglas G Adler¹

¹Division of Gastroenterology, Department of Internal Medicine, and ²Department of Pathology; University of Utah School of Medicine, Huntsman Cancer Center. Salt Lake City, UT, USA

ABSTRACT

Context Alveolar rhabdomyosarcoma are rare malignancies. These lesions typically metastasize to the lungs, liver, and bone marrow. Pancreatic metastases from alveolar rhabdomyosarcoma are exceptionally uncommon. **Case report** An 18-year-old man with a history of right orbital alveolar rhabdomyosarcoma, which had been treated with neoadjuvant therapy, surgery and adjuvant chemotherapy developed an episode of pancreatitis. CT at that time demonstrated acute pancreatitis with no mass lesions. Two months later, the patient developed abdominal pain and an MRI documented a 6.4 cm mass in the pancreatic body and tail. EUS guided FNA confirmed the diagnosis of alveolar rhabdomyosarcoma metastatic to the pancreas. **Conclusion** To our knowledge this is the first reported case of EUS guided FNA diagnosis of alveolar rhabdomyosarcoma metastatic to the pancreas. This is also the only case report of a primary pancreatic metastasis of this type of tumor in a male patient (which occurs less commonly than in females).

INTRODUCTION

Alveolar rhabdomyosarcomas are rare tumors. We present a case of an 18-year-old man who developed a primary pancreatic metastasis of alveolar rhabdomyosarcoma two years after initial diagnosis and treatment. The lesion was diagnosed via EUS guided FNA.

CASE REPORT

An 18-year-old man presented with a one-month history of double vision and swelling within the right orbit. Past medical history is significant for childhood asthma. A head CT revealed a soft tissue mass centered within the right maxillary sinus invading the right orbit and adjacent nasal cavity, maxillary alveolar process, buccal space and cheek. A biopsy was performed and revealed diffuse reactivity for myogenin, desmin, and CD56 and no reactivity for cytokeratin, CD45, CD99, or S-100. Rare cells were reactive for neurospecific enolase, chromogranin, and synaptophysin which is consistent with alveolar rhabdomyosarcoma as were the hematoxylin and eosin stains demonstrated in Figures 1 and 2.

Received August 1st, 2009 - Accepted September 16th, 2009

Key words Neoplasm Metastasis; Pancreas; Rhabdomyosarcoma

Correspondence Douglas G Adler

Therapeutic Endoscopy, Gastroenterology and Hepatology, Huntsman Cancer Center, University of Utah, Salt Lake City, UT, USA

Phone: +1-801.581.5036; Fax: +1-801.581.8007

E-mail: douglas.adler@hsc.utah.edu

Document URL <http://www.joplink.net/prev/200911/13.html>

The patient received ifosfamide, mesna, vincristine, and etoposide (replaced with adriamycin later in the therapy) with concurrent radiation therapy. Following neoadjuvant therapy, the tumor was resected. Soft tissue densities within the retromaxillary fat pad and retromolar fat pad were concerning for persistent tumor and these were resected again. The patient underwent a total of 12 chemotherapy cycles.

Nine months later, the patient began developing midepigastic, unremitting pain, which was initially attributed to an episode of binge drinking on the patient's birthday. He presented to an urgent care facility where his lipase was 3,500 (reference range: 0-

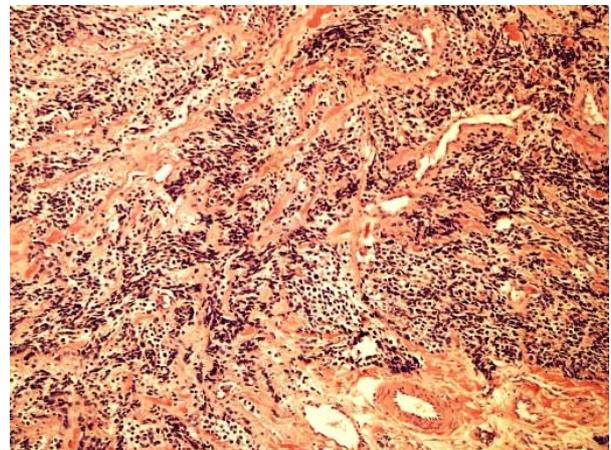


Figure 1. Low-power view of alveolar rhabdomyosarcoma with alternating cellular and myxoid areas, characteristic of this tumor. This specimen was obtained from the patient's right maxillary sinus at the time of initial diagnosis. (H&E stain, 20x)

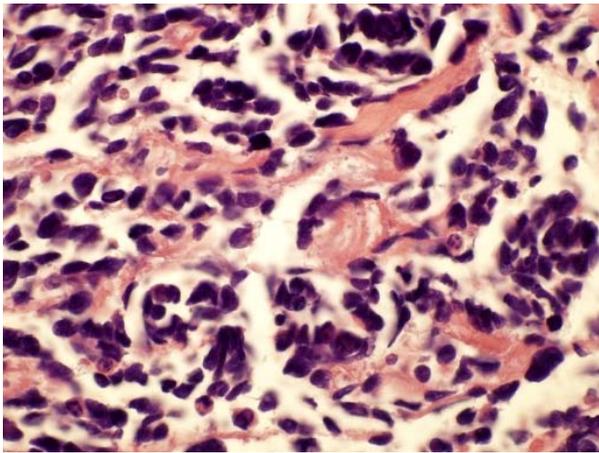


Figure 2. High power view of alveolar rhabdomyosarcoma composed predominately of primitive ovoid cells and devoid of rhabdomyoblastic differentiation, also obtained from the patient's right maxillary sinus at the time of initial diagnosis. (H&E stain, 60x)

160 U/mL). A CT demonstrated findings consistent with acute pancreatitis. He denied any nausea, vomiting, diarrhea, weight loss, jaundice or decreased appetite. Two months later, by this time nearly two years after initial presentation, routine imaging by PET/CT had findings consistent with a new lesion in his pancreas and para-aortic region a SUV of 11.4. Abdominal MRI confirmed a pancreatic mass 3.6x6.4

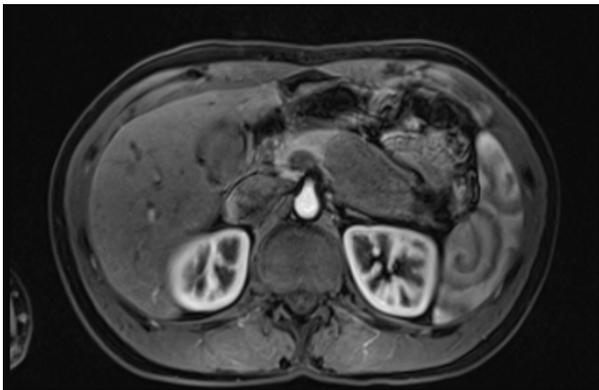


Figure 3. Abdominal MRI image of 3.6x6.4 cm mass in the body/tail of the pancreas.



Figure 4. EUS (7.5MHz) image of large hypoechoic pancreatic mass lesion.

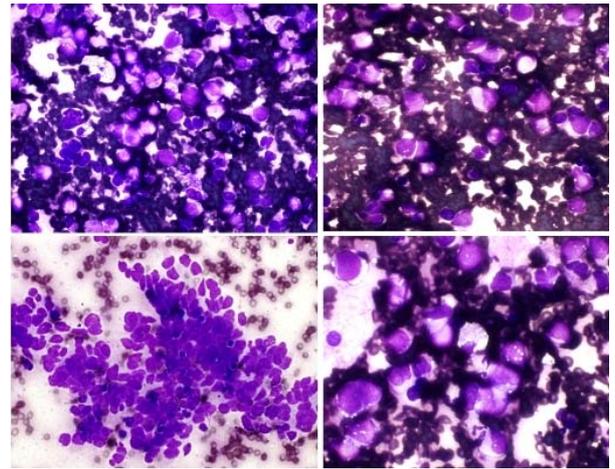


Figure 5. EUS-guided fine needle aspiration of metastatic alveolar rhabdomyosarcoma. Aspirate smears are highly cellular with neoplastic cells lying in sheets and individually. Necrotic debris is present in the background. The neoplastic cells have fragile cytoplasm, and naked nuclei are common. The majority of cells resemble small lymphocytes with compact, hyperchromatic nuclei and scanty cytoplasm. (Romanowsky stain, 60x)

cm and an enlarged left para-aortic lymph node (Figure 3).

The patient was referred for EUS evaluation. Linear EUS was performed at 7.5 and 10 MHz with Doppler. A few small peripancreatic nodes were observed. All were less than 1 cm with mixed echogenicity but round and well-demarcated. The body and tail of the pancreas were essentially replaced by a large, well defined, well demarcated, hypoechoic, solid mass lesion. The mass abutted the splenic vasculature without evidence of invasion. The remainder of the pancreas had a normal appearance (Figure 4). EUS guided FNA was performed using a 22 gauge needle via a transgastric

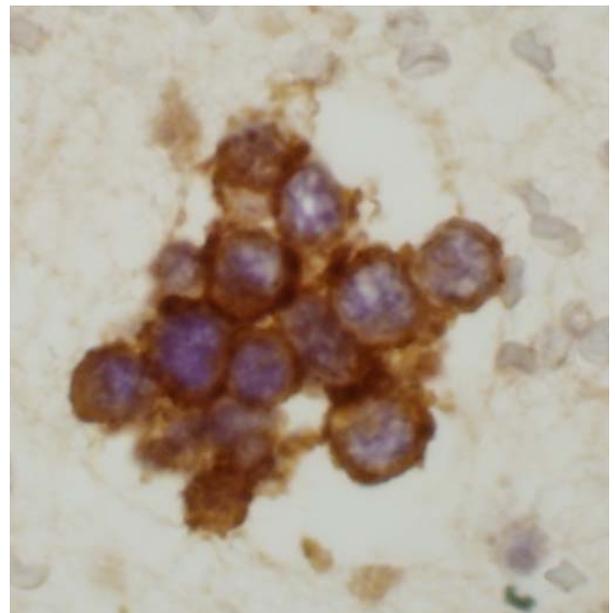


Figure 6. EUS-guided fine needle aspiration of metastatic alveolar rhabdomyosarcoma. The neoplastic cells demonstrated positive staining with antibodies directed against CD56. This antigen is present in many cases of alveolar rhabdomyosarcoma, including the original excisional biopsy specimen removed 2 years previously.

approach. Immunohistochemical staining revealed strong diffuse positive staining with antibodies against CD56 and focally positive staining with vimentin and myo-D1; negative staining included keratin, CD45, S-100, and desmin with an equivocal myogenin, all of which were consistent with metastatic rhabdomyosarcoma (Figures 5 and 6).

The patient has since undergone four rounds of neoadjuvant chemotherapy. He subsequently underwent surgical resection of the pancreatic lesion followed by four more cycles of his doxorubicin, ifosfamide and vincristine. A follow-up PET/CT demonstrated 75-80% reduction in tumor mass. He then underwent laparoscopic distal pancreatectomy and splenectomy with 1/8 peripancreatic lymph nodes containing metastasis. The patient subsequently began adjuvant chemotherapy consisting of vincristine, dactinomycin, and cyclophosphamide with concurrent radiotherapy in the first cycle, cyclophosphamide and topotecan for an additional cycle with radiotherapy for each cycle, and ending with a temozolomide/irinotecan cycle. The patient is currently doing well.

DISCUSSION

Alveolar rhabdomyosarcomas are rare tumors. They are most commonly encountered in adolescents and young adults. Alveolar rhabdomyosarcoma tends to involve the extremity and axial musculature, and can spread both locally and metastatically. Metastases typically spread to the lungs but also the liver, bone marrow, and bone. Of all the rhabdomyosarcomas, alveolar rhabdomyosarcoma (with exception of the undifferentiated type) portends the poorest prognosis [1]. Morphologically, rhabdomyosarcomas appear similar to other small round blue cell tumors such as Ewing sarcoma, lymphoma and desmoplastic small round blue-cell. Alveolar rhabdomyosarcoma comprises 31%, of all rhabdomyosarcomas [2]. Alveolar rhabdomyosarcomas histologically present as loose round to oval cells which appear within fibrous septa [3]. PAX-3 PAX-7 (so-called "master switches of myogenesis" and regulators of proto-oncogenes) and FOXO1a (FKHR) are the genes typically involved in

alveolar rhabdomyosarcomas; although less than 25% are without testable translocations [3, 4, 5].

To our knowledge this is the first reported case of EUS guided fine needle aspirated diagnosis of an alveolar rhabdomyosarcoma metastatic to the pancreas. Case series involving rhabdomyosarcoma metastasis to the pancreas have been reported although they were neither the primary metastasis nor the alveolar subtype [6, 7]. Additionally, this is the only case report of an alveolar rhabdomyosarcoma with a primary pancreatic metastasis in a male patient which occurs less commonly than in women [6, 8].

Conflict of interest The authors have no potential conflicts of interest

References

1. Tsokos M, Webber BL, Parham DM, Wesley RA, Miser A, Miser JS, et al. Rhabdomyosarcoma. A new classification scheme related to prognosis. *Arch Pathol Lab Med* 1992; 116:847-55. [PMID 1497467]
2. Qualman SJ, Coffin CM, Newton WA, Hojo H, Triche TJ, Parham DM, Crist WM. Intergroup Rhabdomyosarcoma Study: update for pathologists. *Pediatr Dev Pathol* 1998; 1:550-61. [PMID 9724344]
3. Breitfeld PP, Meyer WH. Rhabdomyosarcoma: new windows of opportunity. *Oncologist* 2005; 10:518-27. [PMID 16079319]
4. Parham DM, Ellison DA. Rhabdomyosarcomas in adults and children: an update. *Arch Pathol Lab Med* 2006; 130:1454-65. [PMID 17090187]
5. Clark MA, Fisher C, Judson I, Thomas JM. Soft tissue sarcomas in adults. *N Engl J Med* 2005; 353:701-11. [PMID 16107623]
6. Miller DV, Coffin CM, Zhou H. Rhabdomyosarcoma arising in the hand or foot: a clinicopathologic analysis. *Pediatr Dev Pathol* 2004; 7:361-9. [PMID 15383931]
7. Khalbuss WE, Gherson J, Zaman M. Pancreatic metastasis of cardiac rhabdomyosarcoma diagnosed by fine needle aspiration. A case report. *Acta Cytol* 1999; 43:447-51. [PMID 10349379]
8. Farah RA, Kamen BA. Parameningeal alveolar rhabdomyosarcoma with an isolated pancreatic metastasis. *Pediatr Hematol Oncol* 1999; 16:463-7. [PMID 10505324]