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Alveolar Rhabdomyosarcoma

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Abstract

Alveolar Rhabdomyosarcoma is a rare soft tissue tumor seen in children and adolescents. In this case report, we discuss how the common emergency department chief complaint of abdominal pain lead to the uncommon diagnosis of a rare tumor. This Emergency Department (ED) workup and appropriate imaging allowed for identification and necessary transfer to a specialty center, which lead to definitive diagnosis and prompt treatment of this rare condition.

Introduction

We present a patient with a new diagnosis of alveolar rhabdomyosarcoma. Although this is an extremely rare disorder, we report how careful physical examination and appropriate workup with imaging, facilitated timely diagnosis and treatment of this patient. Prompt treatment with chemotherapy was started and is still ongoing.

Case Description

A 13-year-old male with no past medical history presented to the emergency department with his parents for evaluation of three days of abdominal pain. The pain initially began around his umbilicus and then migrated to the right upper quadrant with radiation into the upper back. The pain had suddenly worsened that day after playing basketball. He endorsed associated nausea and decreased oral intake, but denied any other symptoms. He reported last bowel movement was yesterday night. He denied any recent falls or trauma. There was no history of abdominal surgeries.

Upon arrival to the emergency department (ED), the patient's vitals were blood pressure 135/72 mmHg, pulse of 62 beats per minute, temperature was 98.6 ° F, respiratory rate 14 breaths per minute, and oxygen saturation of 98% on room air. Physical exam demonstrated an uncomfortable appearing male with right upper quadrant and right lower quadrant tenderness with involuntary guarding. The patient had no rebound tenderness. The patient had positive McBurney's point tenderness, negative Murphy's exam. There was no costovertebral angle tenderness to palpation. He had a normal genitourinary examination. The rest of the physical exam was unremarkable.

Laboratory values of complete blood count, basic metabolic profile and liver profile demonstrated a white blood cell count of 16.1 while all other results were within normal range. A complete abdominal ultrasound was obtained which showed no abnormalities however the appendix was not visualized. Given the patient's leukocytosis and continued involuntary guarding, a CT scan of the abdomen and pelvis with contrast was obtained, and demonstrated a 5.8 x 3.2 x 5.0 cm mass in the right upper abdomen.

The patient was transferred to a nearby children's hospital, and was evaluated by pediatric surgery and hematology/oncology. He had a biopsy of the mass performed that confirmed an alveolar rhabdomyosarcoma. He was started on high intensity chemotherapy in an attempt to shrink the tumor prior to operating, and discharged after a one-week hospitalization. Chemotherapy was continued in the outpatient setting. Three months after discharge, the tumor was surgically resected and was found to be necrotic secondary to the intensive chemotherapy that was started on initial admission. Currently, the patient is continuing treatment with chemotherapy.

DISCUSSION

- Alveolar rhabdomyosarcomas occur in skeletal muscle and are derived from mesoderm.
- These tumors most commonly present as a painless growing mass and are usually detected secondary to symptoms of compression at the primary site.
- The most common locations for this tumor are the limbs, spinal cord, and pelvic region, followed by the urogenital area. Other, not so frequent locations include the skin and abdomen (such as in our case) (4).
- Rhabdomyosarcomas make up less than 3% of all childhood cancers. Alveolar rhabdomyosarcomas account for 20-30% of all rhabdomyosarcomas, therefore alveolar rhabdomyosarcoma represents less than 1% of all childhood malignancies. They have an annual incident rate of less than 1 per million children and adolescence (1).
- Diagnosis of Alveolar Rhabdomyosarcoma is made by histopathological studies conducted on a biopsy specimen.
- Diagnostic features of the tumor cells on histology include relatively small cells with scant cytoplasm. They have round regular nuclei with a monotonous chromatin pattern. The cells form aggregates interrupted by fibrovascular septae, and within these aggregates, areas of discohesion often form, resulting in spaces that resemble alveoli of the lung (1).
- The standard treatment is a combination of radiation, intensive chemotherapy and surgery. The 4-year survival rates for children with localized and metastatic alveolar rhabdomyosarcoma are 65% and 15% respectively (1).
- This case demonstrates the need for the community ED physician to diligently work up patient's with red flag exam findings.

Photos

Figure 1



Figure 2

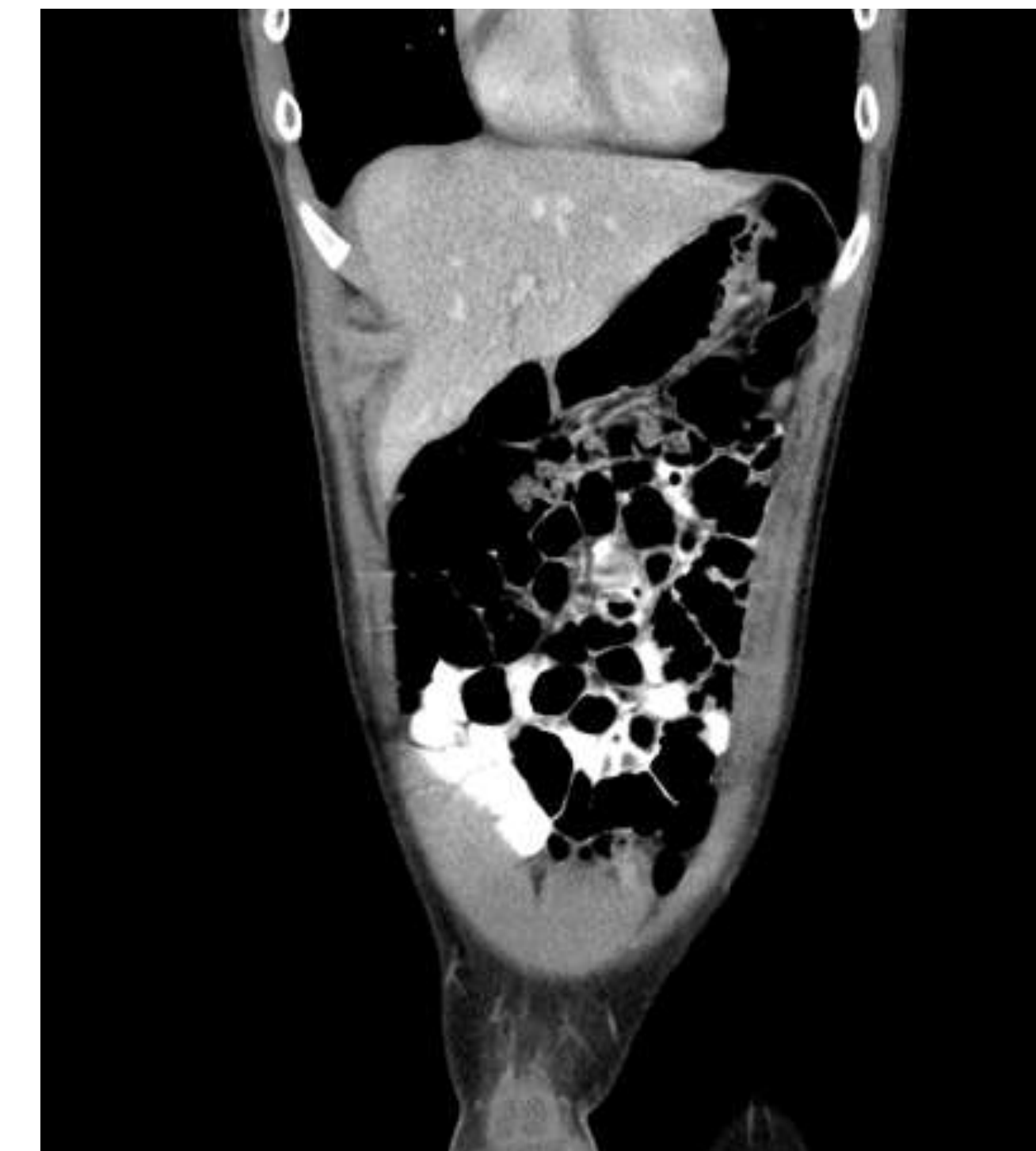


Figure 1 . Axial view of the patient's CT of the abdomen demonstrating tumor with mass effect on the liver.
Figure 2: Coronal view of the patient's CT of the abdomen demonstrating tumor with mass effect on the liver.

Photos

Figure 3

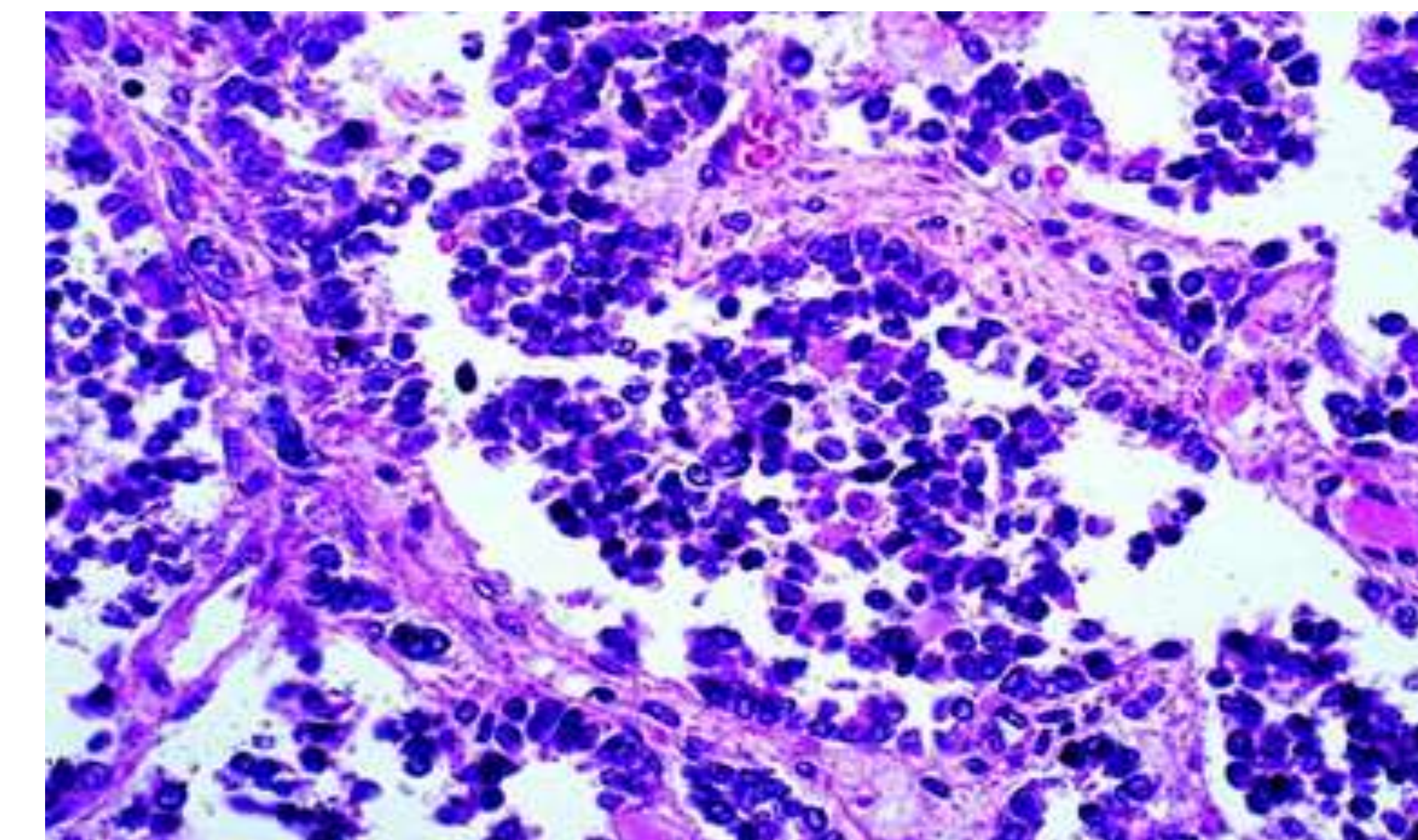


Figure 3 . Histopathology of Alveolar Rhabdomyosarcoma:
<http://atlasgeneticsoncology.org/Tumors/AlvRhabdomyosarcID5194.html>

Conclusion

In the emergency department, abdominal pain is a common chief complaint in the pediatric population with a wide differential diagnosis. A good history taking and physical exam guide imaging studies and work-up. The case of an alveolar rhabdomyosarcoma as described above, though extremely rare, demonstrates one of the numerous possibilities that should be considered when evaluating a child or adolescent with persistent, severe abdominal pain.

References

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