Alveolar Rhabdomyosarcoma Causing Acute Compartment Syndrome of the Forearm: A Case Report and Review of the Literature

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Introduction

Acute compartment syndrome of the forearm has multiple etiologies affecting patients of all ages, but is most commonly caused by fractures, penetrating trauma, drug abuse, crush injuries and intravenous infiltration. Less common etiologies include tourniquet use, hemophilia, snakebites, phlebitis, burns, and postsurgical issues [1–8]. Tumors, while well-documented as an etiology of abdominal compartment syndrome [9], are an extremely rare cause of compartment syndrome in the upper or lower extremities. Herein, we describe the case of an acute forearm compartment syndrome caused by alveolar rhabdomyosarcoma. This is followed by a review of the surgical literature on tumors as an etiology of forearm compartment syndrome and subsequent wound management.

Case Presentation

An 18 year old male with a 1 year history of a left upper extremity alveolar rhabdomyosarcoma was referred to our tertiary care center for chemotherapeutic treatment of his tumor. Outside magnetic resonance imaging revealed a 6.7×6.9×5.9 cm enhancing mass encasing the median nerve. The

orthopaedic surgery service was consulted the morning after admission by the medical oncology service to evaluate for left forearm compartment syndrome.

The patient reported increasing pain and ulnar neuropathy with paresthesias in the left small finger beginning 4 days prior to his presentation to the hospital. The patient's symptoms were not improved by elevation, and there had been no provoking incident or trauma to his forearm. The patient did not notify the admitting oncology service of the change in his symptoms until the morning following his admission because of an acute increase in paresthesias in his ulnar nerve distribution overnight. The Orthopaedic service was consulted immediately after the patient's admission of his change in symptoms. On physical examination significant swelling and taught compartments were appreciated in the proximal left volar forearm. Sensation was intact to light touch in the medial and radial nerve distribution, despite the tumors proximity to median nerve. However, decreased sensation was noted in the ulnar distribution with two point discrimination >15 mm throughout the ring and small fingers. Full thumb and finger flexion was intact, however pain was elicited with active and passive extension of all fingers.

Venous thrombosis had been excluded prior to our consultation by venous ultrasound, and bedside intracompartmental pressure measurements obtained distal to the tumor mass were 58 mm Hg and 68 mmHg in the dorsal and volar compartments, respectively. The patient's blood pressure at the time of compartment pressure measurements was 120/68, demonstrating a delta p of 10 in the dorsal compartment, and 0 in the volar compartment.

The orthopaedic oncology department was emergently consulted regarding the risks of contamination of the remaining volar compartment and potentially the dorsal compartment versus the known complications of an untreated compartment syndrome. Based on the clinical findings, compartmental pressures and with input from our orthopaedic oncology

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Fig. 1 **a** and **b** Axial and Coronal T2 weighted MRI of the patient's forearm demonstrating a 6.7×6.9×5.9 cm enhancing mass

colleagues the patient was taken to the operating room for emergent forearm fasciotomies.

The dorsal compartment was addressed first to minimize contamination from the volar tumor mass. A longitudinal dorsal incision was made to release all dorsal fascial compartments. The muscle was noted to extrude through the fascia once released, consistent with significant compartment pressure elevation, but no evidence of a hematoma was present. The incision was then thoroughly irrigated and closed with nylon suture.

Fig. 2 Alveolar Rhabdomyosarcoma, magnification 20. Dyshesive and infiltrative small round blue tumor cells divided by collagenous fibrovascular septa; mitotic figures are easily identified

Fig. 3 Alveolar Rhabdomyosarcoma, magnification 20. The tumor cell nuclei are highlighted by an immunohistochemical stain that targets myogenin

The volar compartment was then visualized, and demonstrated tight swollen compartments around the muscles. Both superficial and deep volar compartments, as well as the carpal tunnel and distal forearm fascia were released. The volar incision could not be closed primarily. Temporary dermatotraction with vessel loupes was incorporated. A wound vacuum-assisted closure was avoided because of concern of increased vascularity in the area of the tumor.

The patient successfully underwent delayed primary closure 2 months after the initial surgery following 1 cycle of Vincristine and Irinotecan chemotherapy. Following this final surgery the patient worked with our pediatric occupational therapy department. Follow-up physical examination 15 months following the patient's initial fasciotomy revealed painless full active flexion and extension of his fingers, with minimal subjective sensory changes only in the left small finger. However, the patient did continue to have weakness in his adductor pollicis musculature consistent with a positive Froment's sign, and two point discrimination in the ulnar nerve distribution between 10 and 15 mm in his affected hand, compared with 5 mm in the contralateral extremity. The patient's median and radial nerve function have remained intact.

Discussion

While rhabdomyosarcoma has been described as the most common tissue sarcoma in the pediatric population [10], to our knowledge it has not been previously cited as an etiology of compartment syndrome. Few case reports have described tumors causing compartment syndrome in the extremities, with the majority being sarcomas of different histopathology [11–13] and extramedullary manifestations of lymphoma infiltrates [14, 15]. One case of compartment syndrome has also

been presented as a late manifestation of metastatic melanoma [16]. In each of these cases, it is likely that the critical pressure for perfusion of blood was exceeded by the mass effect of the tumor within an enclosed fascial sheath.

Rhabdomyosarcoma may be further classified into botryoid, spindle cell, embryonal, alveolar and undifferentiated types. Each subtype has a predilection for a particular age group; for example, the alveolar subtype is more common in adolescents, whereas the embryonal type occurs more frequently in children less than 8 years old [17]. Perhaps more importantly, the classification has an important correlate with prognosis. According to the International Classification of Rhabdomyosarcoma (ICR) alveolar rhabdomyosarcomas, which comprise 31 % of all rhabdomyosarcomas, have a more aggressive clinical course and worse prognosis and are usually high stage at initial presentation [17, 18]. These tumors typically arise in the deep soft tissues of the extremities; and as they expand entrap normal muscle fibers.

Microscopically, the appearance of the tumor raises a differential diagnosis for small round blue cell tumors, including Ewing sarcoma and lymphoma [17] (Fig. 1). Immunohistochemical stains that target proteins of the myogenic lineage such as myogenin and desmin are useful adjuncts to the histopathologic features [19] (Figs. 2 and 3). Cytogenetic analysis may also be performed to differentiate alveolar from other subtypes, given the presence of a common translocation in the majority of these tumors of t(2;13) [18].

Given the vascularity of this patient's tumor, we initially considered a chronically expanding hematoma within the tumor itself, or an acutely expanding hematoma secondary to some minor trauma as the etiology of the compartment syndrome. Large hematomas have been shown within the lesions of angiogenic tumors such as angiosarcoma [20], synovial sarcoma [21–23], epitheloid sarcoma [24, 25], extraskeletal Ewing's sarcoma [26], leiomyosarcoma [27], liposarcoma [28], or malignant fibrous histiocytoma [29]. However, the timing of the acute increase in pain and the features of the magnetic resonance imaging did not suggest a hematoma as the etiology. Additionally, no hematoma was found intraoperatively.

It is noteworthy to discuss our choice to proceed with first a dorsal then volar compartment release. In compartment syndromes involving both compartments of the forearm, releasing the volar compartment first may additionally decompresses the dorsal compartment, negating the need for a separate dorsal incision [30]. In this case, however, given the tumor location on the volar aspect of the forearm, and the risk of wound contamination with tumor cells [31–33], we did not want to risk seeding the dorsal compartment.

On long term follow up the patient demonstrated minimal complications, despite a change in the patient's symptoms for 4 days prior to presentation. Additionally, there were no intraoperative findings of prolonged compartment syndrome, such

as non viable tissue. In this case, the patient's initial change in symptoms may have begun as the mass effect neared equal to the perfusion pressures. However, the patient had significantly increasing dense ulnar neuropathy once admitted, and therefore likely did not develop full compartment syndrome until later in his course. This limited the time period wherein his perfusion pressures were exceeded by the mass effect of the tumor.

Compartment syndromes due to trauma or fluid infiltration from hyperosmolar IV solution are typically taken back to the operating room within a few days when the swelling has decreased to attempt delayed primary closure. However, in this case the tumor mass prevented closure until medical treatment was initiated. Additionally, a skin flap was not immediately considered given the imminent initiation of chemotherapy. One must also consider the timing of the skin flap with respect to chemotherapy and radiotherapy to avoid tissue complications. In the end, a skin flap was not required given a significant decrease in tumor mass after one cycle of chemotherapy.

In summary, although rarely encountered, alveolar rhabdomyosarcoma should be considered as a potential inciting factor of acute compartment syndrome. The immediate management of the compartment syndrome caused by a tumor involves releasing the involved compartments, however further skin management is dictated by the management of the tumor itself.

Informed Consent The patient was informed that data concerning the case would be submitted for publication, and has agreed to this.

References

1. Kalyani BS, Fisher BE, Roberts CS, Giannoudis PV (2011) Compartment syndrome of the forearm: a systematic review. *J Hand Surg [Am]* 36(3):535–543
2. Prasarn ML, Ouellette EA (2011) Acute compartment syndrome of the upper extremity. *J Am Acad Orthop Surg* 19(1):49–58
3. Eaton RG, Green WT (1975) Volkmann's Ischemia. A volar compartment syndrome of the forearm. *Clin Orthop Relat Res* 113:58–64
4. Mubarak SJ, Owen CA, Hargans AR, Garetto LP, Akeson WH (1978) Acute compartment syndromes: diagnosis and treatment with the aid of the wick catheter. *J Bone Joint Surg* 60A:1091–1095
5. Gelberman RH, Garfin SR, Hergenroeder PT, Mubarak SJ, Menon J (1981) Compartment syndromes of the forearm: diagnosis and treatment. *Clin Orthop Relat Res* 161:252–261
6. Geary N (1984) Late surgical decompression for compartment syndrome of the forearm. *J Bone Joint Surg* 66B:745–748
7. Sneyd JR, Lau W, McLaren ID (1993) Forearm compartment syndrome following intravenous infusion with a manual "Bulb" pump. *Anesth Analg* 76:1160–1161
8. Seiler JG 3rd, Valadie AL 3rd, Drvaric DM, Frederick RW, Whitesides TE Jr (1996) Perioperative compartment syndrome. A report of four cases. *J Bone Joint Surg* 78A:600–602
9. Reis R, Labas P, Vician M, Ziak M (2003) The abdominal compartment syndrome. *Bratisl Lek Listy* 104(1):32–35

10. Huh WW, Skapek SX (2010) Childhood rhabdomyosarcoma: new insight on biology and treatment. *Curr Oncol Rep* 12(6):402–410
11. Shapiro SA, Brindley GW (1995) Synovial sarcoma presenting as an acute compartment syndrome. *J Bone Joint Surg Am* 77(8):1249–1250
12. Niimi R, Matsumine A, Kusuzaki K, Okamura A, Matsubara T, Uchida A, Fukutome K (2006) Soft-tissue sarcoma mimicking large haematoma: a report of two cases and review of the literature. *J Orthop Surg (Hong Kong)* 14(1):90–95
13. Scheipl S, Leithner A, Radl R, Beham-Schmid C, Ranner G, Linkesch W, Windhager R (2007) Myeloid sarcoma presenting in muscle-tissue of the lower limb: unusual origin of a compartment-syndrome. *Am J Clin Oncol* 30(6):658–659
14. Southworth SR, O'Malley NP, Ebraheim NA, Zeff L, Cummings V (1990) Compartment syndrome as a presentation of non-Hodgkin's lymphoma. *J Orthop Trauma* 4(4):470–473
15. Chim CS, Choy C, Liang R (1999) Primary anaplastic large cell lymphoma of skeletal muscle presenting with compartment syndrome. *Leuk Lymphoma* 33(5–6):601–605
16. Simmons DJ, Wharton SM, Waters R (2000) Compartment syndrome complicating metastatic malignant melanoma. *Br J Plast Surg* 53(3):255–257
17. Weiss SW, Goldblum JR (2008) Enzinger and Weiss's soft tissue tumors. Chapter 21. Rhabdomyosarcoma. Fifth edition. Mosby
18. Parham DM, Barr FG (2002) Alveolar rhabdomyosarcoma. *World Health Organization* p. 150–152. Lyon
19. Agarwala S (2006) Pediatric rhabdomyosarcoma and nonrhabdomyosarcoma soft tissue sarcoma. *J Indian Assoc Pediatr Surg* 11(1):15–23
20. Enzinger FM, Weiss S (1995) Soft tissue tumors, 3rd edn. Mosby, St Louis, pp 929–964
21. Naito N, Ozaki T, Kunisada T, Kawai A, Dan'ura T, Morimoto Y et al (2000) Synovial sarcoma with a large hematoma in the inguinal region. *Arch Orthop Trauma Surg* 120:533–534
22. Engel C, Kelm J, Olinger A (2001) Blunt trauma in soccer. The initial manifestation of synovial sarcoma [in German]. *Zentralbl Chir* 126: 68–71
23. Nakanishi H, Araki N, Sawai Y, Kudawara I, Mano M, Ishiguro S et al (2003) Cystic synovial sarcoma: imaging features with clinical and histopathologic correlation. *Skelet Radiol* 32:701–707
24. Romero JA, Kim EE, Moral IS (1994) MR characteristics of epithelioid sarcoma. *J Comput Assist Tomogr* 18:929–931
25. Hanna SL, Kaste S, Jenkins JJ, Hewan-Lowe K, Spence JV, Gupta M et al (2002) Epithelioid sarcoma: clinical, MR imaging and pathologic findings. *Skelet Radiol* 31:400–412
26. Ogose A, Hotta T, Yamamura S, Shioya Y, Yazawa T (1998) Extraskelletal Ewing's sarcoma mimicking traumatic hematoma. *Arch Orthop Trauma Surg* 118:172–173, Vol. 14 No. 1, April 2006
- Soft-tissue sarcoma mimicking large haematoma 95
27. Imaizumi S, Morita T, Ogose A, Hotta T, Kobayashi H, Ito T et al (2002) Soft tissue sarcoma mimicking chronic hematoma: value of magnetic resonance imaging in differential diagnosis. *J Orthop Sci* 7: 33–37
28. Gomez P, Morcuende J (2004) High-grade sarcomas mimicking traumatic intramuscular hematomas: a report of three cases. *Iowa Orthop J* 24:106–110
29. Weiss SW, Enzinger FM (1978) Malignant fibrous histiocytoma: an analysis of 200 cases. *Cancer* 41:2250–2266
30. Ronel DN, Mtui E, Nolan WB III (2004) Forearm compartment syndrome: anatomical analysis of surgical approaches to the deep space. *Plast Reconstr Surg* 114:697–705
31. Saphir O (1954) The transfer of tumour cells by the surgical knife. *Surg Gynecol Obstet* 63:775
32. Brandes WW, White WC, Sutton JB (1946) Accidental transplantation of cancer in the operating room. *Surg Gynecol Obstet* 82:212
33. Enneking WF, Maale GE (1988) The effects of inadvertent tumour contamination of wounds during surgical resection of musculoskeletal neoplasms. *Cancer* 62:1251–1256