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Calcifying fibrous tumor involving the paraspinal region of a child: A rare case report

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ABSTRACT

Introduction: Calcifying fibrous tumor (CFT) is a rare, benign tumor characterized by hyalinized fibrous tissue with distinctive psammomatous and dystrophic calcifications. It is a soft-tissue tumor found in various organ systems. There are only a few reported cases of CFT involving the paraspinal region.

Case Report: We report a 28-month-old female with a paravertebral soft tissue mass. Ultrasound and magnetic resonance imaging (MRI) revealed a well-defined, lobulated lesion in the left paravertebral muscles extending approximately from T9 to L2. The patient had a needle core biopsy and a surgical resection of the tumor. The pathologic diagnosis reported was CFT of the longissimus thoracic muscle.

Conclusion: Postoperative recovery was uneventful and there was no recurrence after follow-up.

Keywords: Calcifying, Case study, Fibrous, Spine, Tumor

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INTRODUCTION

Calcifying fibrous tumors (CFT) are rare, benign mesenchymal tumors that originate within subcutaneous and deep soft tissues [1]. These tumors are histologically categorized by a non-encapsulated mass that contains a blend of bland spindle cells, dystrophic calcifications, dense hyalinized collagen, lymphoplasmacytic infiltrates, and psammoma bodies [2]. While the tumor was originally considered a tumor of soft tissue sites, it has been recorded in various sites around the body, including the spine, mandible, neck, heart, pleura, mediastinum, back, arm, thigh, and other oral, intrascrotal, and paratesticular locations [2].

Recently, CFTs are being documented in the muscles surrounding the spine as lumbar tumors [3]. Calcifying fibrous tumors in the lumbar region are rare in nature, therefore, it is extremely important that surgeons are aware of this pathologic entity, as CFT responds well to surgical treatment alone [3]. Herein, we describe a case of a CFT involving the left paraspinal muscle tissue.

CASE REPORT

A 28-month-old female presented to the orthopedic clinic for evaluation of a mass found on the left side of her

spine. The operating surgeon obtained informed consent from the patient's family prior to her participation in this case report. The mass had been growing progressively but did not seem to cause pain. She had no history of trauma or other pertinent symptoms or medical conditions. The physical exam revealed a 7×32 cm mass on the left side of her spine that was firm and did not move when the patient extended or bent her back. Her neurological status was within normal limits as she could move each limb, react to touch in all distributions, and did not have any functional issues with her arms, legs, bladder, or bowel. The patient received an ultrasound (US) and magnetic resonance imaging (MRI) before referral to the orthopedic clinic.

Ultrasound imaging of the lumbar region revealed a complex soft tissue mass lesion infiltrating the left paravertebral muscle, longissimus thoracic muscle. The tumor measured approximately 5.2×2.8×2 cm with areas of calcification and increased peripheral vascularity. The overlying skin and surrounding subcutaneous fat appeared unremarkable. Multiplanar and multi-sequence non-enhanced and enhanced MRI of the entire spine was conducted. A solid mass infiltrating the left paravertebral muscles that extended from T9 to L2 with two linear enhancements extending toward the left neural foramina at T11-T12 was revealed (Figure 1). No extensions expanded into the neural foramina, spinal canal, thorax, or abdomen. The vertebrae and intervertebral discs had normal morphology with no segmentation abnormality, aggressive marrow lesion, disc herniation, or spinal canal stenosis. Based on these findings, a diagnosis of paravertebral soft tissue mass was made, such as solitary myofibroma, nodular fasciitis, other fibromatosis, fibrosarcoma or other sarcomas, and surgical consultation was recommended.

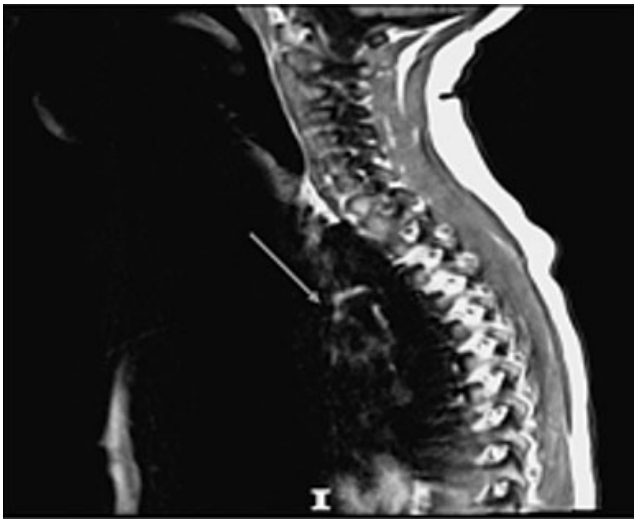

Figure 1: Multiplanar and multi-sequence non-enhanced and enhanced MRI of the entire spine revealed a well-defined, lobulated lesion in the left paravertebral muscles extending approximately from T9 to L2. It is isointense to the muscle on T1-and T2-weighted sequences, with some areas of subtle T1 and T2 hyperintensity.

Pathological findings

The patient underwent a US-guided needle core biopsy to facilitate diagnosis, which revealed a calcifying fibrous tumor with infiltration. A core needle biopsy was performed. Microscopically, the lesion consisted of hyalinized fibrous tissue with distinctive psammomatous and dystrophic calcifications (Figure 2A). The spindle cells show bland nuclei, fine chromatin, and inconspicuous nucleoli between the thick hyalinized collagenous stroma of the lesion (Figure 2B and C), all consistent with a CFT. Immunohistochemistry revealed that the tumor cells were expressing factor XIIIa (Figure 2D) and CD34 and positive for Desmin, but negative for SMA, MSA, ALK-1, S100, CD45, Cyclin-D1, H-caldesmon, Myogenin, IgG4, CD31, CD117, Cd68, CD163, Beta-catenin, myogenin, ERG, CD34, CD31, AE1/AE3, and pan-TRK. Molecular analysis of RNA-derived next-generation sequencing (NGS) libraries using the Trusight Pan-Cancer 1385-gene panel showed no oncogenic single nucleotide variants (SNVs) or gene mutations/fusions were detected. The immunohistochemical findings were typical of a bland hypocellular infiltrative lesion within the skeletal muscles, which was indicative of a CFT.

Given the growth potential of the tumor and the concern that it extended into the neuroforamen and to the ribs, the decision was made to proceed with surgical excision. The tumor was carefully removed with no complication. The mass consisted of a firm reddish tissue that infiltrated into background tissues, entrapping striated muscle bundles. The entire specimen was 7×3.5×2.5 cm and weighed 30 grams.

Microscopically, the lesion profile remained as a CFT. The lesion consisted of a dense collagenous stroma with infiltrating and unencapsulated hypocellular proliferation of bland spindle shaped fibroblast cells, and randomly distributed dystrophic calcification (Figure 2C). The tumor showed immunophenotypic features on the resection identical to those noted on the biopsy. All results taken together allowed for a final diagnosis of CFT. The patient's postoperative course was uncomplicated with no tumor recurrence during the seven years follow-up period.

DISCUSSION

Calcifying fibrous tumor is a rare, benign soft-tissue tumor that was originally reported in 1988 by Rosenthal and Abdul Karim [4]. It is commonly characterized by hypocellular collagen that is densely hyalinized and contains psammomatous or dystrophic calcification [2]. Calcifying fibrous tumor was originally described as a soft-tissue childhood tumor and termed "calcifying fibrous pseudotumor" due to the fibroinflammatory and reactive processes thought the lesion [5]. The belief that the tumor was that of "childhood" was also dropped as more cases were identified in individuals of various ages [5]. It was

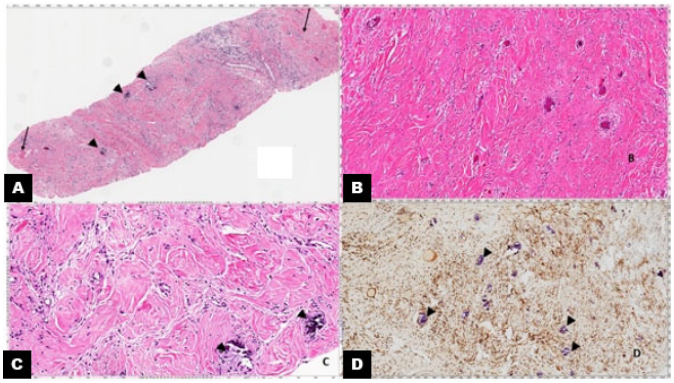

Figure 2: Dense fibrous tissue infiltrating skeletal muscle, with lymphoplasmacytic inflammatory infiltrates and dystrophic calcifications. (A) Core biopsy showing bland fibrous lesion infiltrating into the skeletal muscle fibers at the biopsy edges. Arrows highlighting skeletal muscle fibers, H&E ×40. (B) CFT resection showing hypocellular lesion with dense collagen fibers, H&E ×200. (C) CFT resection showing scattered bland fusiform fibroblast in a dense collagenous background with focal calcific deposits. Arrow heads highlight calcifications, H&E ×400. (D) Factor XIIIa showing positive staining in fibroblasts in CFT. Arrow heads highlight calcifications, factor XIIIa ×200.

only in 2002 that the World Health Organization (WHO) decided on a name for the lesion, which was "Calcifying Fibrous Tumors" [6].

Calcifying fibrous tumor occurs in various age ranges and locations, a systematic review conducted by Chorti et al., in 2016, showed 157 well-documented cases of CFT from 1993 to 2015 into one report [2]. The mean age of patients reported with CFT as 33.58 years, with little difference between genders (1 men:1.27 female). The age ranges from 0 to 4, 25 to 29, and 30 to 34 years and range from 5 weeks to 84 years. None of the cases occurred in paraspinal location. Most common locations of CFT involved the stomach (18%), small intestine (8.7%), pleura (9.9%), mesentery (5%), and peritoneum (6.8%) [2]. More rarely, CFT tumor sites involved the lung, lymph node, breast, and spermatic cord [2]. The present case is extremely rare as it is one of the few reported cases of CFT in the paraspinal or lumbar region (making the case a rare pathologic diagnosis).

The majority of CFTs are asymptomatic, where the tumor is found during routine imaging or physicals, rather than through illness [2]. In the rare case of symptoms, clinical features of CFT are divided into two groups: acute and chronic [2]. Chronic features involve a large painless mass visible under the skin, rarely may be complicated by acute peritonitis or intussusception in the intestines. Other chronic symptoms can include foreign body sensation, weight loss, fever, fatigue, progressive weakness, lack of appetite, and in some cases, pain [1–3, 7]. Pain is a prevalent symptom of intra-abdominal CFT, characterized as a dull, progressive pain, and discomfort following the intake of food [1].

Calcifying fibrous tumors can be difficult to differentiate and diagnose, especially in the preoperative stage. Histologically, some differential diagnoses include solitary fibrous tumor (SFT), inflammatory myofibroblastic tumor (IMT), calcified granulomas, fibromatosis, fibrous hamartoma of infancy, Idiopathic intervertebral disc calcification in childhood, and calcifying aponeurotic fibroma [1, 2].

Solitary fibrous tumor, histologically, is a cellular tumor characterized by CD34 positive spindle cell proliferation and abundant stag horn proliferating vessels. Inflammatory myofibroblastic tumor, histologically, is a cellular tumor characterized by ALK-1 positive spindle cell proliferation inflammatory cellular infiltrate rich in lymphocytes and plasma cells. Both of these are not identified in the current case report. In addition, molecular testing for ALK mutation, that is associated with IMT, was negative. Calcified granulomas, histologically characterized by inflammatory cellular infiltrate and CD68 positive histiocytic granulomas. This is also not identified in the current case report.

In contrast to CFT, calcifying aponeurotic fibroma is most commonly on the palmar surfaces of the hands and fingers. Histologically the tumor is more cellular compared to CFT. It often extends into adjacent soft tissues, where it may be attached to tendons or entrap peripheral nerves and blood vessels and shows nodular calcified chondroid areas. By immunohistochemistry, the tumor cells react with SMA, MSA, CD99, and (in the chondroid areas) S100. On molecular testing, it is associated with detection of the *FN1-EGF* gene fusion.

In contrast to CFT, fibrous hamartoma of infancy shows an organoid, triphasic morphology with bundles of bland fibroblastic/myofibroblastic cells, nodules of primitive, rounded or stellate cells with myxoid stroma, and mature adipose tissue. Calcification is not a feature of fibrous hamartoma of infancy. By immunohistochemistry, fibrous hamartoma of infancy shows variable expression of SMA in the fibroblastic areas and occasionally in the primitive mesenchyme. CD34 is expressed within the primitive mesenchyme and in areas with giant cell fibroblastoma-like morphology.

Idiopathic intervertebral disc calcification in childhood is a calcification of the intervertebral disc and is strictly involving the cervical region. Histologically there is no associated spindle cells proliferation as in CFT.

Calcifying fibrous tumors can be differentiated via a histological and immunohistochemical evaluation. Microscopically, CFT is composed of hyalinized fibrous tissue with distinctive psammomatous and dystrophic calcifications. Among the thick hyalinized collagenous stroma of the lesion, there exists a proliferation of myofibroblastic spindle cells, mononuclear inflammatory infiltrate, and lymphoid aggregates [3]. Immunohistochemically, CFT is characterized by positive expression of factor XIIIa, CD34, and Desmin. Calcifying fibrous tumor is also associated with the negative expression of factor VIII, S100 protein, ALK-1, CD31, and

smooth muscle actin (SMA) [3]. Both the positive and negative expressions were identified in the paraspinal mass in the patient, indicating CFT.

Similarly, to the tumor removed in the case, the macroscopic appearance of CFTs is benign with a firm consistency. The tumors are regularly restricted but not encapsulated [2]. They occur in various organ systems; however, they are most recorded in the gastrointestinal tract, both in the stomach and the intestines [2]. The treatment choice for CFT is complete surgical resection, especially in soft tissues. This can be completed through open surgical excision or laparoscopic excision [2]. However, some cases have reported difficulty in surgically resecting the lesion due to extensions into surrounding tissues [2, 8]. A 2016 review of 157 CFT cases in the international literature found from those with available follow-up data, recurrence of the CFT occurred in 10 out of the 96 cases or roughly 10.4% of cases [2]. Out of these 10 cases, 6 recurrences occurred in pediatric patients [2]. In our case, total resection was achieved and there was no evidence of recurrence after follow-up. A second follow-up is still required to confirm no long-term recurrence and to examine for signs of scoliosis following the procedure.

Due to the rarity of CFT, there is little information surrounding the excision of these lesions and the rate of occurrence; however, in the case of recorded recurrence, the tumor usually remains nondestructive [3]. Our case demonstrates a rare finding of CFT in the paraspinal region. Despite the challenges in excision in such a location, our case did not show recurrence during the 7-year follow-up period. Since CFT was not defined by the World Health Organization until 2002, there is likely literature and historical data that is mislabeled [6]. Even today, CFT continues to be confused and mislabeled in literature, making research on the topic difficult to conduct [6]. The uniqueness of this case provides an added support within the CFT knowledge gap as there is little published data for this tumor type (CFT) in paraspinal location and arising in very young child as noted in our case.

To the best our knowledge, the largest study (not systematic review) included a total 22 cases, reported 13 males and 9 females, ranging in age from 7 to 72 years (mean, 48.5 years; median, 48 years), with only one case in retroperitoneal location and none in the thoracic or lumbar paravertebral region [7]. Another large study included 11 patients reported a male-to-female ratio was 5:6, with a mean age of 38 years and age range of 25–52 years [8]. We preset a rare and unusual case of calcifying fibrosis tumor occurring in a 28-month-old female patient involving the paravertebral thoracic region.

CONCLUSION

In conclusion, CFT is a rare, benign tumor characterized by dense hyalinized collagenous tissue and calcifications. We presented a complex soft tissue mass infiltrating the left paraspinal region, consistent with CFT. Although CFT in the paraspinal region is a rare diagnosis, it should be considered in the evaluation of tumors within this space. Surgical resection is the primary treatment for CFT due to its low percentage of recurrence. Awareness of the features and possible extensions of CFT in the lumbar region and its differential diagnoses will aid in diagnosis and preoperative surgical planning. Misdiagnosis and hence mistreatment are likely caused due to unfamiliarity to clinicians or junior pathologists.

REFERENCES

1. Turbiville D, Zhang X. Calcifying fibrous tumor of the gastrointestinal tract: A clinicopathologic review and update. World J Gastroenterol 2020;26(37):5597–605.
2. Chorti A, Papavramidis TS, Michalopoulos A. Calcifying fibrous tumor: Review of 157 patients reported in International literature. Medicine (Baltimore) 2016;95(20):e3690.
3. Nathoo N, Viloria A, Iwenofu OH, Mendel E. Calcifying fibrous tumor of the spine. World Neurosurg 2012;77(3–4):592.e1–4.
4. Rosenthal NS, Abdul-Karim FW. Childhood fibrous tumor with psammoma bodies. Clinicopathologic features in two cases. Arch Pathol Lab Med 1988;112(8):798–800.
5. Chen KT. Familial peritoneal multifocal calcifying fibrous tumor. Am J Clin Pathol 2003;119(6):811–5.
6. Montgomery E, Rubin S, Thomas G, et al. Calcifying fibrous tumor. World Health Organization Classification of Tumours, Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon, France: IARC Press; 2002. p. 77–8.
7. Emanuel P, Qin L, Harpaz N. Calcifying fibrous tumor of small intestine. Ann Diagn Pathol 2008;12(2):138–41.
8. Kimura M, Kato H, Sekino S, et al. Radical resection of a giant retroperitoneal calcifying fibrous tumor combined with right hepatectomy and reconstruction of the inferior vena cava and bilateral renal veins. Surg Case Rep 2018;4(1):7.

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Author Contributions

Youssef Nasr – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Abbigail King – Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Lauren Tristani – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Moaz Alowami – Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Kevin Smit – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Joseph de Nanassy – Conception of the work, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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The corresponding author is the guarantor of submission.

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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.