

Calcifying Aponeurotic Fibroma in the left pectoral region in an adult: A Case report.

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Abstract:

Calcified aponeurotic fibroma is a rare benign fibroblastic tumor. The lesion has property of local invasion and high recurrence. Because of this property, tumor should be accurately diagnosed pre-operatively. We report a case of 29 years old male, with a palpable painful mass on the anterior chest wall near left pectoral region, describes radiographic and MR findings. No skin defect was detected but patient feels discomfort. An excision procedure was done. The excised tissue was sent for biopsy and calcified aponeurotic fibroma was diagnosed.

Key Words: Calcifying aponeurotic fibroma, Fibroma, Fibroblastic, Soft tissue tumor

Introduction:

Calcifying aponeurotic fibroma was first described by KEASBEY in 1953 and referred as juvenile aponeurotic fibroma.¹ It is a rare, benign locally aggressive fibroblastic soft tissue tumor.¹⁻³ It commonly, occurs in first and second decade of life, with higher prevalence in males over females. Usually, all incidences are closely associated with aponeurosis, tendons or fascia but often invade adjacent bone.^{3,4} The tumor has predilection for local recurrence after surgical excision.^{2,3} We report a very rare case of aponeurotic fibroma in a 29 years old male with 6 months clinical history that presented with a relatively densely calcified mass affecting his anterior chest wall near left pectoral region.

Case Report:

A 29-years old healthy male presented with history of 6 months history of painful, slow growing mass in anterior chest wall near left pectoral region associated with left upper limb tingling. No history of trauma or previous any significant medical history. On physical examination, a firm, non-tender, palpable mass of 6 X 4.5 cm mass was noted at the left pectoral region.(Fig 1)



Fig 1: Clinical Picture of Left pectoral mass lesion.(arrow)

Imaging findings:

On ultrasonography a well defined 53 x 40 x 46 mm (ML x AP x CC) round to oval heterogeneously hypoechoic lesion noted in left pectoral region under the pectoralis muscle. Multiple echogenic foci of calcification noted within the lesion with internal vascularity. (Fig 2)

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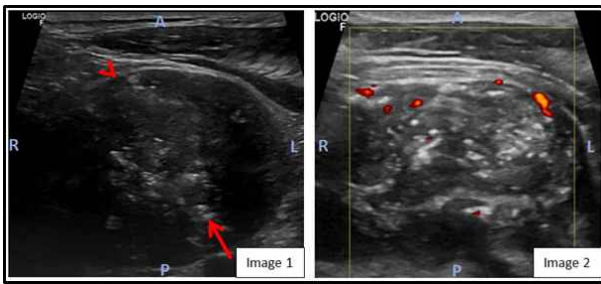


Fig 2: Image 1=USG image showing well defined,round to oval, heterogeneously hypoechoic lesion (arrowhead) with multiple echogenic calcific foci within (arrow).Image 2=Pulsed wave doppler image showing internal vascularity within the lesion.

On CT scan a 58 x 41 x 49 mm (ML x AP x CC) well defined isodense lesion with calcification noted.No evidence of vascular involvement was noted.(Fig 3)

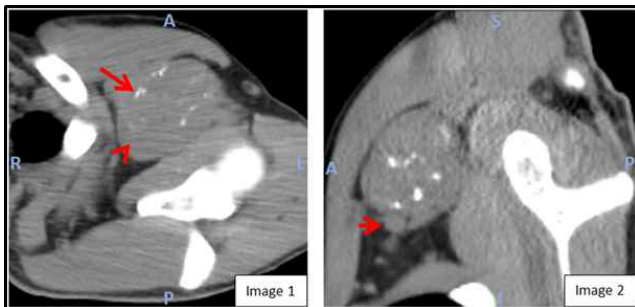


Fig 3: Image 1=Axial Plain CT scan image showing well defined,round to oval, isodense lesion (arrowhead) with multiple echogenic calcific foci within (arrow). Image 2=Sagittal Plain CT scan image showing axillary vessels at the inferior margin of the lesion.(arrow)

On MRI, 56 x 37 x 51 mm (MLxAPxCC), large lobulated lesion which is iso to hypointense on T1W images and isointense on T2W images and displacing adjacent nerves and vessels. Moderate heterogeneous enhancement is noted on contrast study. Adjacent fat planes are Normal. (Fig 4)

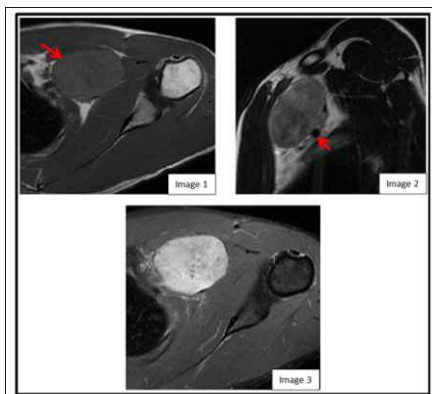


Fig 4: Image 1=Axial T1(TE:18.58 ms,TR:558 ms) scan image showing well defined,round to oval, iso to hypointense lesion (arrow). Image 2=Sagittal T2 (TE:122.628 ms TR:6355 ms) scan image shows isointense lesion displacing the vessels inferiorly.(arrow). Image 3=Post Gadolinium axial T1 FS image (TE:18.58 ms,TR:971 ms) showing enhancement of the lesion.

Patient underwent excision and specimen was sent for Histopathological correlation. (Fig 5) Microscopy confirmed the diagnosis of Calcifying aponeurotic fibroma.(Fig 6)



Fig 5: Image 1: Gross excised specimen Image 2: Cut section of specimen showing multiple pink to red congested areas.(arrow)

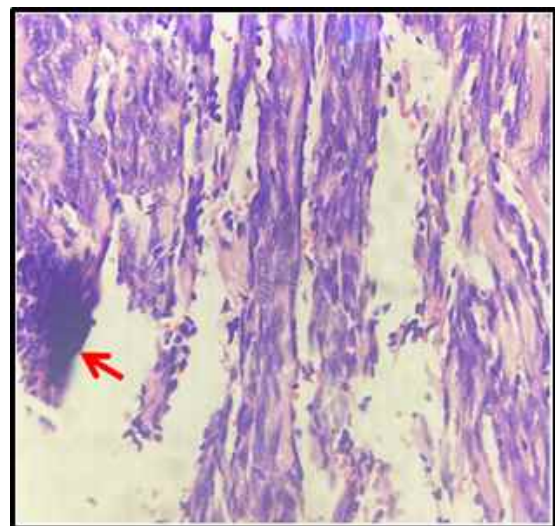


Fig 6: Microscopic image (40 x)=multiple spindle to oval cells arranged in short & long fascicles. Stroma shows thick rosy collagen at places and foci of patchy irregular calcification.(arrow)

Management

Treatment is surgical, with complete excision, in case of diffuse type, the tumor may not fully excised. 50% of tumors has reported to show local recurrence.

Recurrence rates are higher in younger patients aged less than 5 years. Radical excision to be avoided, regarding the benign characteristics of CAF, considering that tumor aggression decreases with time and growth.

Discussion:

CAF is a rare invasive fibroblastic tumor that was first reported as juvenile aponeurotic fibroma. Usually, tumor is painless rarely causes pain and decrease in range of motion. The most common site sites are palm, soles, rarely in chest region, back area, thigh and knee in areas closely related to aponeurosis, tendons or fascia.⁴⁻⁶

Generally, CAF does not metastasize. Rare cases had reported malignant transformation.⁷⁻⁹ Lafferty et al.⁷ reported the metastasis of a calcifying aponeurotic fibroma of the palm in a 3-year-old girl as a metastatic fibrosarcoma to the lungs and bones 5 years after a second local excision. Dr Sharon W. Weiss⁸ has also reviewed a single case of malignant transformation of a calcifying aponeurotic fibroma during consultation. Finally, Benichou et al.⁹ reported the case of a 7 year old boy with local recurrence for seven years leading to hand amputation and fibroblastic process extending to lung and pleura which caused the child's death.

Imaging modality plays a limited role in accurate diagnosis of CAF. In some case on X-ray CAF shows soft tissue protrusion with calcific stippling¹ although it is not always related with pathological calcifications. CT shows non specific soft tissue mass with stippling of calcification. MRI provides information regarding preoperative planning and lesion extension.

The definitive diagnosis is based on histopathological biopsy. Tumor is although benign, it tends to invade surrounding fat, and engulfs nerves, vessels and muscle fibers without destroying them. CAF has shown the property of local recurrence even after excision.

References:

1. Keasbey L.E. Juvenile aponeurotic fibroma (calcifying fibroma); a distinctive tumor arising in the palms and soles of young children. *Cancer*. 1953 Mar;6(2):338-46. doi: 10.1002/1097-0142(195303)6:2<338::aid-cncr2820060218>3.0.co;2-m. PMID: 13032926.
2. Goldman RL. The cartilage analogue of fibromatosis (aponeurotic fibroma). Further observations based on 7 new cases. *Cancer*. 1970 Dec;26(6):1325-31. doi: 10.1002/1097-0142(197012)26:6<1325::aid-cncr2820260620>3.0.co;2-m. PMID: 5483662.
3. Murphey MD, Ruble CM, Tyszko SM, Zbojniewicz AM, Potter BK, Miettinen M. From the archives of the AFIP: musculoskeletal fibromatoses: radiologic-pathologic correlation. *Radiographics*. 2009 Nov;29(7):2143-73. doi: 10.1148/rg.297095138. PMID: 19926768.
4. Corominas L, Sanpera I Jr, Sanpera-Iglesias J, Ramos-Ansesio RF. Calcifying aponeurotic fibroma in children: our experience and a literature review. *J Pediatr Orthop B*. 2017 Nov;26(6):560-564. doi: 10.1097/BPB.0000000000000335. PMID: 27182752.
5. *J Pediatr Orthop B*, 26 (2017), p. 560. KA Lafferty, EL Nelson, RJ Demuth, et al. Juvenile aponeurotic fibroma with disseminated fibrosarcoma. *J Hand Surg Am*, 11 (737) (1986).
6. Kwak HS, Lee SY, Kim JR, Lee KB. MR imaging of calcifying aponeurotic fibroma of the thigh. *Pediatr Radiol*. 2004 May;34(5):438-40. doi: 10.1007/s00247-003-1110-7. Epub 2003 Dec 20. PMID: 14689252.
7. Lafferty KA, Nelson EL, Demuth RJ, Miller SH, Harrison MW. Juvenile aponeurotic fibroma with disseminated fibrosarcoma. *J Hand Surg Am*. 1986 Sep;11(5):737-40. doi: 10.1016/s0363-5023(86)80024-7. PMID: 3760506.
8. Enzinger FM, Weiss SW. Fibrous tumors of infancy and childhood. In: Enzinger FM, Weiss SW, editors. *Soft tissue tumors*, 7th ed. Elsevier; 2019. pp. 295–299. ISBN: 9780323610964.
9. M Benichou, P Balmes, CH Marty-Double, Y. Allieu Fibrome aponévrotique juvénile (tumeur de Keasbey) à évolution métastatique. A propos d'un cas [Juvenile aponeurotic fibroma (Keasbey's tumor) with metastatic progression. A propos of a case] *Ann Chir Main*, 8 (2) (1989), pp. 150-153, 10.1016/s0753-9053(89)80008-0 French. PMID: 2802841.