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Cystic Angiomatosis: A Radiological Dilemma

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Abstract

Cystic angiomatosis is a rare benign disorder with multifocal hemangiomatous or lymphangiomatous lesions of the axial or appendicular skeleton, spleen, liver, and lungs. Patients usually are asymptomatic and are incidentally detected on radiographs. The spectrum clinical presentation may include splenomegaly, hepatomegaly, cystic hygromas or effusions, and pathological fracture depending on the affected area, they may also have neurological symptoms with vertebral localization and skull lesions.

This case report illustrates the prompt and accurate diagnosis of cystic angiomatosis using radiological investigations, prior to definitive biopsy.

Keywords: Cystic angiomatosis, cystic angiomatosis radiology, cervical spine compression, osteolytic bone lesion, osteolytic lesion radiology, MRI spine

Introduction

Cystic Angiomatosis (CA) also known as Generalized lymphatic abnormality (GLA), is a benign vascular disorder which is characterized by cystic skeletal lesions often seen in conjunction with extra skeletal cysts. The most commonly involved bones are femur followed by pelvis, humerus, skull and vertebrae. Small bones like carpals, metacarpals, phalanges and tarsus bones are rarely involved. Spleen frequently shows visceral lymphangiomatosis in CA [1]. Lungs, pleura and liver are seldom involved, but cases showing the same have been reported in literature [2,3]. Cervical spine lesions in CA pose a huge threat to spinal cord compression. Pulmonary complications pivotally determine the prognosis of the patient.

The first features of CA are manifested in childhood and early adulthood but a few cases have also shown a

second frequency peak after the fifth decade of life. Males have a predilection of varying amounts towards CA, however this finding may be inconsistent.

The first line of investigation done is an X-ray. The findings of the same are quite ambiguous and confusing, thus leading to a plethora of differential diagnoses. Gorham Stout Disease (GSD) is one such entity that shows a close resemblance with CA. That being said, GSD is more severe than CA and holds a poor prognosis [4]. The radiological features of CA are also mistaken as bony metastasis, due to their overlapping features like osteolytic lesions, osteoblastic lesions, periosteal reaction, etc. Other differentials are Langerhan cell histiocytosis, multiple myeloma, TB osteomyelitis and multiple enchondromas as seen in Mafucci syndrome [5,6].

The most consistent findings of the bony lesion on X-Ray are intramedullary or subarticular multifocal osteolytic cysts with a well-demarcated sclerotic rim with little or no periosteal reaction. The cortex of the bone is thus usually spared [1]. The radiological features show variation with the progression in the phases of the disease, which are - a phase of osteolysis, followed by a phase of stabilization, fibrous scarring, and osteoformation [7].

MRI findings are almost diagnostic of the condition due to similar findings in most cases. T2 weighted scans very frequently show a hyperintense signal due to liquid contents of the cyst. The findings on T1 weighted sequences vary with the contents of the cyst (serum/blood). Enhancement is seen in the lesion or its rim after administering contrast [1,8].

FDG PET scan can be employed for ruling out the differentials of bony metastasis hardly any cases have been reported showing uptake.

The case described below is that of a 15-year-old male with cervical myelopathy due to cord compression in whom multiple bony cysts were identified and provisionally diagnosed as cystic angiomatosis on radiology which was then confirmed on histopathology after neurosurgical interventions.

Case Report

This case is of a 15-year-old male who was referred to a tertiary care hospital, presenting with the chief complaint of weakness in all four limbs since one year. It began in the bilateral upper limbs which then gradually progressed to the lower limbs. The onset was insidious and did not have any specific precipitating or relieving factors.

On neurological examination, the patient had decreased power (2/5) distally in bilateral upper limbs and power of (3/5) proximally in both upper limbs. The power in the lower limbs was 2/5 throughout except in the foot it was 4/5. The tone was normal in the upper limbs but was increased in the lower limbs. All four limbs showed decreased muscle bulk. The deep tendon reflexes (DTR) were brisk in the lower limb with bilaterally well-sustained plantar clonus. There were no other significant positive findings on neurological examination.

An MRI carried out previously (a year prior to the present day) showed - osteolytic lesions in the C2 to D1 vertebrae involving both the anterior and posterior segments. The lesions were hypointense and hyperintense on T1 and T2 weighted scans respectively and showed mild enhancement after the administration of contrast. The pathology attributed wedging and moderate reduction in the vertebral height of the C6 and C7 vertebrae. Kyphotic deformity was noted which caused moderate spinal canal stenosis from C6 to D1 levels resulting in cord compression and edema. The right first rib and clavicle showed similar affection. No abnormal signal intensity was seen in the cord at any level. The

differential diagnoses were cystic angiomatosis and Langerhans cell histiocytosis. The rest of the spine were unremarkable. [Figure 1]

The patient was scheduled for Halo orthosis immobilization and observed for 4 months for improvement. The power in the upper limb improved over 4 months to 5/5 after Halo immobilization. No improvement was seen in the lower limb.

A second MRI scan was conducted 11 months after the first MRI scan to observe for progression. Varied-sized cystic lesions were noted in all cervical vertebrae and entire D1 vertebrae which were hyperintense on T2 weighted sagittal sequences, T2 weighted axial sequences and hypointense on T1 weighted sagittal sequence [Figures 2, 3 and 4]. The lesions showed patchy post-contrast enhancement. The C7 vertebra showed maximum collapse and vertebral body height reduction of 60-70%. All other cervical vertebrae show varying amounts of body height reduction. C5-C7 vertebrae were seen forming an angular gibbus resulting in cord compression and spinal canal narrowing [Figure 2]. T2 weighted hyperintense signal was noted within the spinal cord with thinning of the cord at the same level [Figure 2]. The findings were suggestive of cystic angiomatosis causing cervical myelopathy and myelomalacia.

An NCCT (Non-Contrast Computed Tomography) of the spine taken concurrently shows lower cervical kyphosis and spinal canal narrowing. Multiple lytic lesions were seen in C2-D1 vertebrae [Figures 5]. With decreased body height in C3-T1 vertebrae and partial collapse in C6, C7, and D1.

In view of worsening cord compression, the patient was posted for surgery. The patient was placed in supine position under general anesthesia (GA) with a traction of 6 kg weight with slight extension of the neck. The skin was painted and draped after which a skin incision was made. The planes between the trachea, esophagus medially, and carotid sheath laterally were identified and dissected meticulously with peanut to access the pre vertebral fascia. Beneath the Longus coli muscle, the vertebral bodies were exposed to the C4-D2 level. After dissecting the intervertebral spaces, a corpectomy was done in the C6 C7, and D1 vertebrae. A small dural defect at the C5 level was noted which was corrected by placing a collagen-based dural graft (Duragen). Then using an expandable cage, C5-D2 fusion was achieved by using spinal implants and screws. Antibiotic coverage was given and the wound was closed leaving two in-situ minivac drains. The patient was shifted to the critical care unit (CCU) on ventilatory support.

The post-operative history of the patient was unremarkable. The power in the lower limbs improved to 3/5 under critical care. The patient was discharged nine days post-surgery.

The post-operative histopathology revealed multiple bony fragments along with variable-size cystic spaces lined by a single layer of endothelial cells [Figures 6, 7]. A few spaces also showed red blood cells and blood vessel proliferation. Thus, the diagnosis of cystic angiomatosis was confirmed.

On follow-up, after 4 months now the patient has shown good improvement. The power in both upper and lower limbs is 5/5 and can do all daily activities efficiently.

Discussion

Cystic angiomatosis (CA) is primarily a skeletal system disorder but frequently affects the spleen, lungs, and pleura. The presentation can be variable depending on the site of the lesions. **Jacobs, Julian E.; Kimmelstiel, Paul** reported the first case of cystic angiomatosis in 1953. It involved the left elbow first and then the proximal part of the left femur [9].The most commonly involved bones are the femur followed by the pelvis, humerus, skull, and

vertebrae. But rare presentations involving smaller bones have also been reported. One such case was reported by **Papanagiotou P, Karantanas AH** which showed subchondral osteolytic cysts in the proximal and middle phalanges [5]. Bone pain, pathological fractures, etc. may be the common clinical features. It may also be an incidental finding in radiological investigations.

Lesions in the lungs, pleura, or spine worsen the prognosis and need to be followed in consecutive years. A total of twenty-one cases involving the vertebra have been reported, out of which only five cases involving the cervical spine have been reported in the literature [1].The spinal lesions show considerable loss of bony mass leading to deformities and cord compression [10]. Thus, they come with a significant risk of mortality. **N K Sferopoulos, D Anagnostopoulos, J K Webb** reported a case of a 2 ½-year-old male with CA involving the cervical spine who died of neurological alterations due to massive osteolysis at the age of 15 [11]. In the case illustrated by us, the patient was diagnosed promptly and treated without any present neurological deficits.

The main challenge is to distinguish it radiologically from various other differential diagnoses like GSD, metastasis, other primary bone tumors, Langerhans cell histiocytosis, multiple myeloma, TB (tuberculosis) osteomyelitis as it plays a pivotal role in the management of the patient. Out of all the radiological investigations, an MRI of the suspected lesion gives the most reliable and consistent findings. Some highlighting and frequently encountered features of CA on MRI also seen in our case are – hyperintense signal on T2W scan, enhancement on administration of contrast, and preserved bony cortex with little to no periosteal reaction. Osteolytic bone metastases are unlikely to show a preserved bony cortex.

Although X-ray is the first investigation in most cases, it gives little insight into the lesion but still holds importance due to its ability to localize the lesion.The cysts appear well circumscribed with a sclerotic rim preserving the bony cortex on X-Ray. At times, X-rays may also give a falsely normal appearance as in a case illustrated by **Aurélie Najm, Elise Soltner-Neel, Benoît Le Goff, Pascale Guillot, Yves Maugars, Jean-Marie Berthelot** [1]. Other modalities like CT, PET, and 99 mtc, hold little importance. The case of CA given by **Caroline Souza Dos Anjos, Rafaela Melo Campos Borges, Ananda Castro Chaves, William Hiromi Fuzita, Carlos Augusto Moreira Silva, Ubirata Gomes Loureiro de Oliveira** revealed glycolytic hypermetabolism on PET which falsely raises suspicion of a malignant etiology [2].

In our case, no significant interval changes were seen in the two MRI scans. The condition of the patient improved drastically after the neurosurgical interventions which relieved the cord compression. Thus, it proves that the symptoms of the patient were primarily due to cord compression.

Conclusion

Despite a wide range of imaging features, lesions of cystic angiomatosis have characteristic features on MRI scans.

This case illustrates the importance of the prompt and accurate diagnosis of cystic angiomatosis from a wide range of imaging features by using various radio diagnostic tools for optimum patient management.

References

1. Najm A, Soltner-Neel E, Le Goff B, Guillot P, Maugars Y, Berthelot J-M (2016) Cystic angiomatosis, a heterogeneous condition: Four new cases and a literature review. Medicine (Baltimore) 95:e5213

2. Dos Anjos CS, Borges RMC, Chaves AC, Fuzita WH, Silva CAM, de Oliveira UGL (2019) Cystic angiomatosis, pleural effusion and multiple bone lesions mimicking a metastatic malignant neoplasia: a case report. J Med Case Rep 13:265
3. Sawalgi V, Amin D, Arora AJ, Srivastava M, Uppin M (2022) A rare case of systemic cystic angiomatosis involving the bones, spleen, liver, and lungs. Cureus14:e30414
4. Albayrak B, Aydın Şimşek Ş, Cengiz T, Muslu O, Büyükceran İ, Coşkun HS, Dabak N (2023) Gorham-Stout disease: A case report and review of the literature. Cureus15:e38756
5. Papanagiotou P Cystic angiomatosis of the hand. In: Eurorad - Brought to you by the ESR. https://www.eurorad.org/case/2442.
6. Weerakkody, Yuranga & Gilcrease-Garcia, Brian 2018 'Generalised lymphatic anomaly' Radiopaedia.org
7. Reid AB, Reid IL, Johnson G, Hamonic M, Major P (1989) Familial diffuse cystic angiomatosis of bone. Clin OrthopRelat Res 238:211–218
8. Schlecht I, Mäurer J, Schulte-Overberg U, Kenzel P, Zystische Angiomatose FR Erscheinungsbildim Magnetresonanztomogramm
9. Jacobs JE, Kimmelstiel P: Cystic angiomatosis of the skeletal system. J Bone Joint Surg Br. 1953, 35:409-464.
10. Pavanello M, Piatelli G, Ravegnani M, Consales A, Rossi A, Nozza P, Milanaccio C, Carbone M, Cama A. Cystic angiomatosis of the craniocervical junction associated with Chiari I malformation: case report and review of the literature. Childs Nerv Syst. 2007 Jun; 23(6):697-700.
11. S feropoulos NK, Anagnostopoulos D, Webb JK. Cystic angiomatosis of bone with massive osteolysis of the cervical spine. Eur Spine J. 1998;7(3):257-9.

Legend Figures

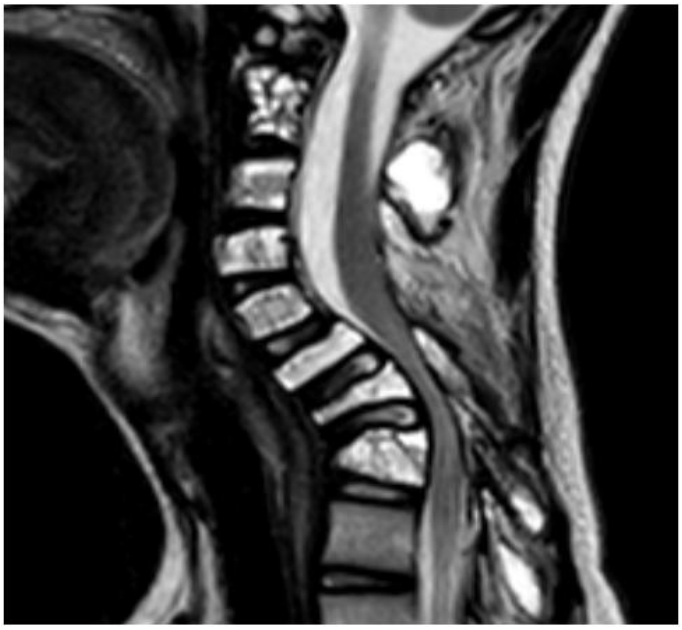

Figure 1: First MRI conducted of the patient showing T2W hyperintense cystic lesions from C2 to D1 with spinal canal stenosis from C6 to D1.

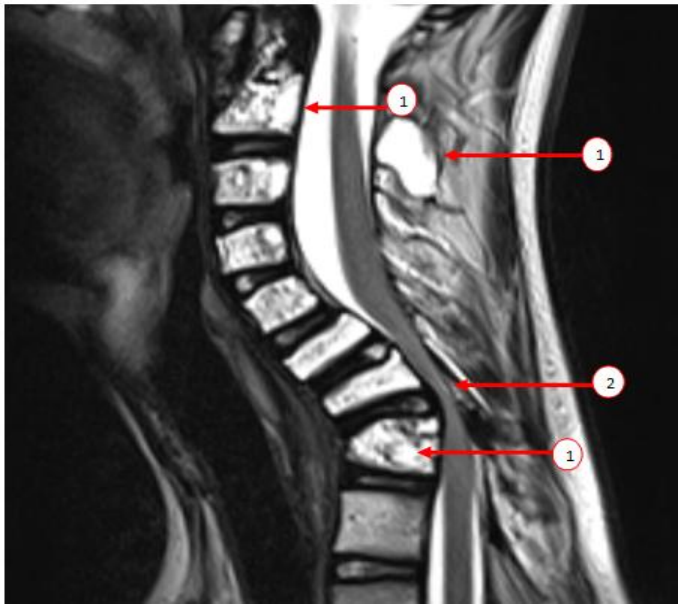

Figure 2: T2W image of the latest MRI scan available showing hyperintense cystic lesion (1) and angular gibbus and cord compression at C5-C7 levels (2).

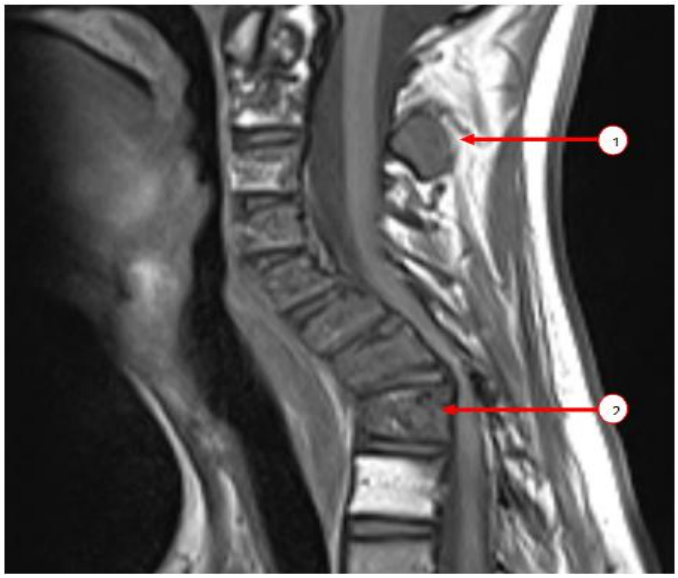

Figure 3: Latest T1W hypointense lesions in Cervical spine (1) and D1 vertebra (2)

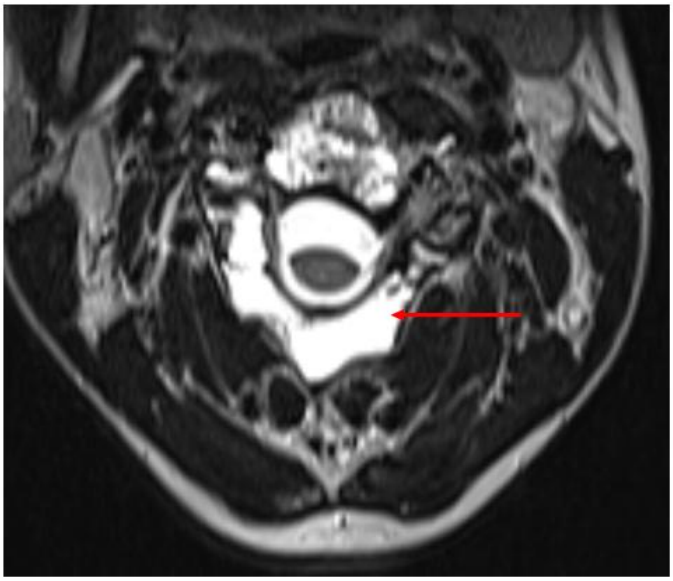

Figure 4: Latest T2W axial scan showing a hyperintense cystic lesion in the posterior segment of the vertebra

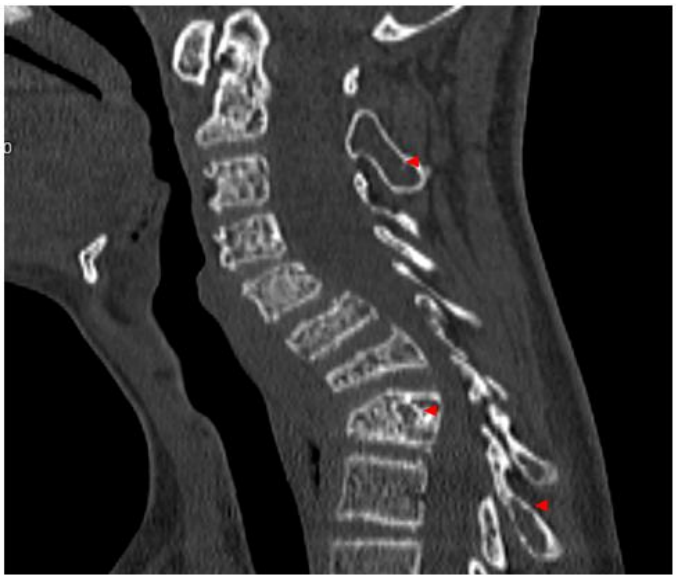

Figure 5: Sagittal CT of the cervical spine showing multiple osteolytic lesions (Arrow heads)

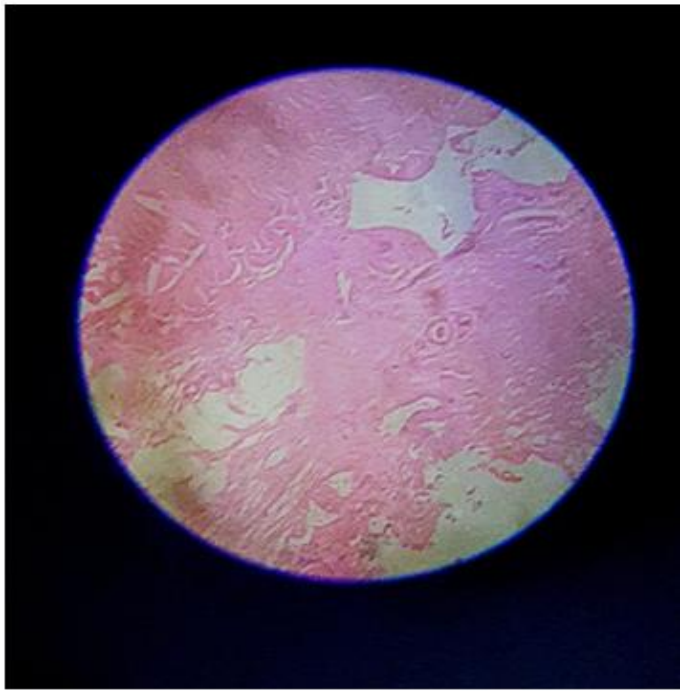

Figure 6: Histopathological slide seen at 10X power showing cancellous bone with cystic spaces

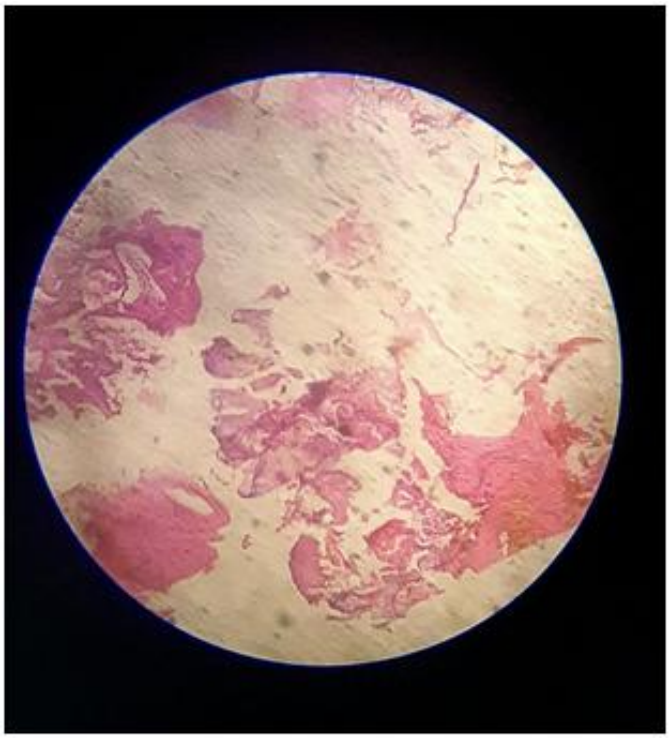

Figure 7: Histopathological slide seen at 10X magnification showing bony cysts filled with blood