Nuclear Medicine /

Molecular Imaging:

Systemic Mastocytosis: A Rare Cause of Single Vertebral Body Uptake on Bone Scan

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ABSTRACT

Systemic Mastocytosis is a rare condition characterized by the abnormal proliferation of Mast Cells. Presentation as a solitary vertebral body lesion is extremely uncommon and may be confused with more ominous conditions such as metastasis. Familiarity with the condition can heighten clinical suspicion, direct tissue diagnosis, guide management and indicate appropriate follow up. We present a case of a 64-year-old woman undergoing staging for recently diagnosed breast cancer who was found to have Systemic Mastocytosis of a single vertebral body.

CASE REPORT

CASE REPORT

A 64 year old woman was referred to our radiology department for evaluation of a palpable right breast mass. Diagnostic mammogram showed a spiculated mass in the right upper outer quadrant (Figure 1a and 1b). Targeted ultrasound demonstrated an irregularly-shaped hypoechoic mass at the 10 o'clock position, highly suspicious for malignancy (Figure 2). Core needle biopsy revealed infiltrating, moderately differentiated ductal carcinoma. Sentinel lymph node biopsy was positive for adenocarcinoma. On metastatic workup, a Tc99m MDP bone scan demonstrated a unifocal, horizontally oriented focus of increased radiotracer uptake at the T10 vertebral body (Figure 3). Correlation with available radiographs demonstrated no osseous abnormality at that site (Figure 4). An FDG PET/CT demonstrated heterogeneous osteosclerosis without significant FDG avidity (Figures 5-7). There was no evidence of visceral involvement. The presence of a single non-FDG avid metastatic focus remained of primary concern. Because such a finding would significantly alter treatment options and prognosis (upstaging the patient to

referred for CT guided biopsy of the T10 vertebral body (Figure 8). Pathology showed extensive proliferation of spindle mast cells confirmed by immunophenotype (positive for CD117, CD25, and mast cell tryptase), satisfying the WHO diagnostic criteria for Systemic Mastocytosis (Figures 9-11). She did report a recent fall that aggravated her chronic low back pain although she did not seek treatment. She reported no other recent illnesses or hospitalization. The patient's past medical history included hyperlipidemia, but she denied any history of hypersensitivity, urticaria or allergies. Physical examination revealed an afebrile woman with blood pressure of 145/78, heart rate of 76 beats per minute without palpable organomegaly or other abnormality. Lab evaluation with Complete Blood Count (CBC), Basic Metabolic Panel (BMP), Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP) were unremarkable. DEXA scan demonstrated osteopenia. Treatment was initiated for T2N1M0 breast cancer. In the absence of systemic symptoms, no treatment of her Systemic Mastocytosis was given.

stage IV), tissue sampling was deemed necessary. She was

DISCUSSION

Etiology and Demographics

Systemic Mastocytosis (SM) is a rare condition with an incidence of 5-10/100,000 per year [1]. Seven World Health Organization variants have been described [2]. These range from a benign Mastocytic Tumor to myeloproliferative leukemia [2]. Seventy five percent of cases are diagnosed in children as Urticaria Pigmentosa (UP) with a peak incidence of 2 years [3]. The most commonly encountered form in adults is the Indolent Type (Indolent System Mastocytosis) with a median age of 49 years at diagnosis [4]. There is no gender predilection [4]. Pathogenesis appears to result from abnormal mast cell proliferation due to a random mutation in the kit gene [5].

Clinical and Imaging Findings

Diagnosis of Systemic Mastocytosis depends on fulfillment of one major and one minor or 3 minor WHO criteria [1]. The major criterion is the presence of multifocal dense mast cell infiltrates in one or more extra-cutaneous organs, most commonly the bone marrow. Minor criteria include the following: i) abnormal morphology of extracutaneous mast cells (spindle-shaped cells); ii) increased serum tryptase level (>20 ng/ml); iii) abnormal expression of CD2 and/or CD25 on bone marrow mast cells; iv) detection of a kit mutation at codon 816 in an extra-cutaneous organ [6]. Our patient was classified as Isolated Bone Marrow Mastocytosis (BMM) subtype in the absence of cutaneous symptoms. Interestingly, she denied symptoms of mast cell degranulation (bronchospasm, anaphylaxis, atopy) seen in 86% of patients with this variant [2]. Assessment for kit mutation was not undertaken as the tissue specimen was sufficient for the diagnosis (dense mast cell aggregates with abnormal CD25 expression).

Symptomatology is widely variable and dependent on the WHO type. The most commonly involved organ is the skin with pruritus, erythema or orticaroid lesions erupting with mechanical stress (Darier's sign) [6]. Physical disfigurement may lead to development of neurosychiatric symptoms such as depression and even suicidal ideation [7]. Skeletal manifestations are present in up to 70% of SM cases as osteopenia, bone pain and insufficiency fractures [9]. Bone marrow infiltration, if diffuse, may lead to pancytopenia [6]. Visceral involvement most commonly affects the lymph nodes, liver and spleen and presents with organomegaly [2]. Frank organ failure is possible, typically associated with more aggressive forms of the condition [1]. Peptic ulcer disease, abdominal pain and diarrhea have also been described as Mastocytosis Enterocolitis [10]. As the result of mast cell degranulation, anaphylaxis, hypotension and syncope can occur especially in the setting of physical stress [6]. Laboratory tests including an elevated serum tryptase (>20ng/ml⁻¹) can be helpful as one of the minor criteria [11]. Blood counts are important in monitoring for progression to bone marrow suppression or leukemia [7]. Peripheral eosinophilia is seen in up to 20% of cases [7]. Other lab values, including a normal ESR and CRP, may be useful in ruling out other conditions such as osteomyelitis. Because of the wide variability and non-specificity of symptoms, the

diagnosis may be elusive on clinical grounds alone as symptoms can be seen in a variety of more common conditions [12].

Radiographic findings have been reported in up to 70% of patients with SM [13]. The most commonly encountered pattern is that of diffuse osteoporosis and osteopenia, believed to result from heparin production by mast cells [14]. Osseous demineralization can be monitored with DEXA scans [2]. Ill defined, discrete, sclerotic lesions of varying shapes asymmetrically distributed throughout the axial and appendicular skeleton characterize another presentation, and can be mistaken for metastatic disease [15]. Frenzel reported a case of a SM presenting with the "Ivory Verytebra Sign" i.e densely sclerotic vertebral body which is more commonly seen with metastatic disease or osteomyelitis. [16]. Although frequently involved, skeletal lesions are not part of the diagnostic criteria [6].

CT findings of skeletal involvement resemble those of radiographs [13]. Although no specific CT signs exist, it offers superior resolution and can aid in differentiating SM from other disorders. Cross sectional imaging has the added benefit of imaging visceral manifestations of SM such as hepatosplenomegaly, and retroperitoneal and mesenteric adenopathy [17]. Despite her recent trauma, our patient had intact vertebral bodies without a linear lucency or cortical disruption to suggest a fracture. The absence of vertebral body trabeculations and sparing of the posterior elements made the diagnosis of Paget's disease less likely. The intact endplates ruled out a Schmorl's node. Preservation of the soft tissue planes immediately surrounding the vertebral body argued against osteomyelitis.

Findings of SM on MRI are nonspecific but may visualize bone marrow infiltration by mast cells. A mosaic pattern of low to intermediate signal on T1 and intermediate to hyperintense signal on T2 weighted images has been described [18,30]. Since there is no correlation between bone marrow signal changes and mast cell percentage in bone marrow biopsy, prospective suspicion of SM on MRI would be difficult [18]. Contrast enhancement is not typically seen with SM and, when present, should argue for an alternative diagnosis such as metastasis or Sarcoidosis [19]. An MRI was not felt to be indicated in our patient as definitive tissue sampling would ultimately be necessary given the clinical and prognostic implications associated with the presence of a distant metastatic focus (Stage IIB versus Stage IV).

Scintigraphy with Tc99 methylene disphosphonate (MDP) is more sensitive to osseous involvement than radiographs with up to 74% of SM patients demonstrating abnormal uptake [20]. The activity pattern varies and can include diffuse osseous uptake producing a "superscan" [21]. Unifocal uptake is much less common. In a study of 75 patients with SM, only 2 patients had uptake at a solitary site [21]. Focal Tc99m MDP uptake of a single vertebral body as in our patient (the so called "Black Ivory Sign") has yet to be described for SM [22]. The most commonly encountered pathology is focal metastasis followed by acute compression fracture, Paget's disease and Lymphoma [22]. Scintigraphy can be used to

assess disease progression in SM, even before the development of systemic symptoms or bone marrow findings [21]. Serial bone scans offer an efficient and effective alternative to cross sectional monitoring and have prognostic value, as patients with more extensive osseous involvement carry a worse prognosis [10].

The use of FDG-PET/CT in SM has been limited to a small number of cases [23]. Findings are similar to other myeloproliferative disorders, demonstrating no significant visceral or osseous FDG avidity [23, 24]. The presence of FDG avidity would argue for an alternative diagnosis such as metastasis. Although FDG-PET/CT has demonstrated excellent efficacy in detecting bony metastasis in breast cancer patients, its sensitivity is degraded with smaller primary breast lesions and sclerotic bony metastasis [31]. In our case, a non-FDG avid metastatic focus required exclusion and therefore tissue sampling was deemed necessary.

Treatment and Prognosis

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There are no curative therapies for SM. Symptomatic treatment with antihistamines, Disodium cromolyn, or leukotriene inhibitors provides good control in patients with mild symptoms [25]. Immunomodulators (prednisone, azathioprine, methotrexate, cyclosporine) could also be considered in patients, with formularies and combinations tailored to each patient [25]. Patients should be counseled to avoid triggers such as systemic stress. Epinephrine should be reserved for life threatening anaphylaxis [1]. Omalizumab is an effective antagonist to the IgE mediated symptoms reserved for patients who fail first line therapy [26]. Bisphosphonates have been proven safe in SM for treatment of patients with osteoporosis [27].

Cytoreductive agents (Interferon alpha (IFN), 2 Chlorodeoxyadenosin (2CdA), Fludarabine, Cytarabine, Mitoxantrone) are reserved for symptoms refractory to basic therapy with identifiable changes in hematologic profile [28]. Tyrosine Kinase Inhibitors (Imatinib, Dasatinib and Midostaurin) have also undergone evaluation although reports of their usefulness are limited [29]. Bone marrow transplant may offer a last resort [29].

Prognosis depends on the disease subtype. Children diagnosed with the condition typically see resolution by adolescence [2]. Aggressive forms have a 1% per year risk of leukemic transformation or development of concurrent lymphoma and carry a prognosis of less than 6 months [2]. Those with systemic involvement have a median survival of 5 years [3]. Close monitoring of disease progression using bone scan and serum tryptase levels can help identify systemic progression at an early stage and indicate cytoreductive or chemotherapeutic agents [29].

Differential Diagnosis

In the absence of typical cutaneous symptoms, the nonspecificity of clinical and imaging findings makes the differential diagnosis of SM quite broad. In the setting of isolated bone uptake on bone scan, differential considerations can be trimmed substantially, although tissue sampling is usually required for definitive diagnosis. The most important distinction is from sclerotic metastasis as the result of lung, breast or prostate cancer [30]. Trauma, infection and arthropathy are other common conditions that warrant consideration. A review of the differential diagnosis for a single Tc99m- MDP avid vertebral body is provided in table 1.

TEACHING POINT

Systemic Mastocytosis is a rare myeloproliferative disorder that can have visceral, cuteaneous and osseous manifestations. Imaging findings are non specific and laboratory/ pathologic evaluation of a tissue sample is required for diagnosis. Systemic Mastocytosis presenting as focal Tc99m-MDP radiotracer uptake of a solitary vertebral body is extremely uncommon and may be confused with more ominous conditions such as metastasis. The use of serial Tc99m-MDP bone scans is a practical means of monitoring disease burden and has prognostic indications.

REFERENCES

- 1. Patnaik MM1, Rindos M, Kouides PA, Tefferi A. Systemic mastocytosis: a concise clinical and laboratory review. Arch Pathol Lab Med. 2007May;131(5):784-91. PMID: 17488167
- 2. Pardanani A. Systemic Mastocytosis: Disease overview, pathogenesis and treatment. Hematol Oncol Clin N Am 2012; 26(5):1117-1128. PMID:23009941
- 3. Vardiman J, Thiele J, Arber D, Brunning RD. The 2008 Revision of the WHO Classification of Myeloid Neoplasms and Acute Leukemia: Rationale and Important Changes. Blood 2009; 114(5):937-51. PMID:19357394
- Akin C, Metcalfe D. Systemic mastocytosis. Annu Rev Med 2004; 55:419-32. PMID:14746529
- 5. Garcia-Montero, A. Jara-Acevedo, M. Teodosio, C. (2006) KIT mutation in mast cells and other bone marrow hematopoietic cell lineages in systemic mast cell disorders: a prospective study of the Spanish Network on Mastocytosis (REMA) in a series of 113 patients, Blood,Vol.108, pp.2366-2372,
- Valent P, Horny H, Escribano L, Longley B. Diagnostic criteria and classification of mastocytosis: a consensus proposal. Leuk Res. 2001;25(7):603-25. PMID:11377686
- Amon U, Hartmann K, Horny HP, Nowak A. Mastocytosis an update. J Dtsch Dermatol Ges 2010; 8(9):695-711. PMID:20678151
- Fritz J, Fishman EK, Carrino JA, Horger MS. Advanced imaging of skeletal manifestations of systemic mastocytosis. Skeletal Radiol 2012;41(8):887-97. PMID:22366736

- Ramsay DB, Stephen S, Borum M, Voltaggio L, Doman DB. Mast cells in gastrointestinal disease. Gastroenterol Hepatol (N Y). 2010; 6(12):772-7. PMID 21301631
- Van der Goot W, de Monchy JG, Kluin-Nelemans HC, van Doormaal JJ. High prevalence of fractures and osteopersosis in patients with indolent systemic mastocytosis. Allergy. 2012; 67(3):431-8. PMID:22229787
- 11. Metcalfe DD. Cells and Mastocytosis. Blood. 2008; 112(4):946-56. PMID:18684881
- 12. Genovese A, Spadaro G, Triggiani M, Marone G. Clinical advances in mastocytosis. Int J Clin Lab Res. 1995;25(4):178-88. PMID:8788545
- 13. Deramond H, Remond A, Grumbach Y, Dieval M, Coevoet B, Duriez J, Katz M. Bone locations of systemic mastocytosis: radiological and scintigraphic study. J Radiol. 1980; 61(8-9):503-8. PMID:7463392
- Huang TY, Yam LT, Li CY. Radiological features of systemic mast-cell disease. Br J Radiol. 1987;60(716):765-70. PMID:3664176
- 15. Braga FS, de Oliviera GA, de Melo AM, de Assis Barbosa LG, Fraga JV. Indolent systemic mastocytosis: is there a reliable radiographic pattern? Pediatr Radiol. 2012;42(8):1017-20. PMID:22281758
- Frenzel L, Suarez F, Chandesris M. Ivory Vertebra and Systemic Mastocytosis. Joint Bone Spine. 2012;79(3):319-21. PMID: 22366148
- Avila NA, Ling A, Worobec AS, Mican JM, Metcalfe DD. Systemic Mastocytosis: CT and US Features of Abdominal Manifestations. Radiology. 1997;202(2):367-72. PMID:9015059
- Roca M, Mota J, Giraldo P, García Erce JA. Systemic Mastocytosis: MRI of bone marrow involvement. Eur Radiol 1999; 9:1094-1097. PMID:10415240
- 19. Haney K, Russell W, Raila FA. MRI characteristics of systemic Mastocytosis of the lumbosacral spine. Skeletal Radiology (1996) 25; 171-173. PMID: 8848749
- Arrington ER, Eisenberg B, Hartshorne MF, Vela S, Dorin RI. Nuclear Medicine Imaging of Systemic Mastocytosis. J Nucl Med. 1989;30(12):2046-8. PMID:2585108
- 21. Chen C, Andrich M, Mican J, Metcalfe D. A retrospective analysis of Bone scan abnormalities in Mastocystosis: Correlation with Disease Category and Prognosis. J Nucl Med 1994; 35(9):1471-5. PMID:8071694
- 22. Greene GS, Maurer AH. Solitary vertebral body uptake on Bone Scintigraphy. The black ivory" Vertebrae Sign. Semin Nucl Med. 1985;15(3):317-21. PMID:2930900

- 23. Zettinig G, Becherer A, Szabo M, Uffmann M, Dudczak R, Valent P, Kletter K. FDG Positron Emission Tomography in Patients with Systemic Mastocytosis. AJR Am J Roentgenol. 2002;179(5):1235-7. PMID:12388504
- 24. Tomihama RT, McEachen JC, Kuo PH. Imaging of Systemic Mastocytosis by FDG-PET/CT Demonstrates increased activity in cortical bone. Clin Nucl Med. 2008;33(3):220-3. PMID:18287854
- 25. Quintas-Cardama A, Aribi A, Cortes J. Novel Approaches in the Treatment of Systemic Mastocytosis. Cancer. 2006:107(7):1429-39. PMID:16948123
- 26. Molderings GJ, Raithel M, Kratz F, Azemar M, Haenisch B, Harzer S, Homann J. Omalizumab treatment of systemic mast cell activation disease: experiences from four cases. Intern Med. 2011;50(6):611-5. PMID:21422688
- 27. Valent P, Sperr WR, Akin C. How I treat patients with advanced systemic mastocytosis. Blood. 2010;116(26):5812-7. PMID:20855864
- 28. Droogendijk HJ, Kluin-Nelemans HJ, van Doormaal JJ, Oranje AP, van de Loosdrecht AA, van Daele PL. Imatinib mesylate in the treatment of systemic mastocytosis: A phase II trial. Cancer. 2006;107(2):345-51. PMID:16779792
- 29. Metzgeroth G, Dinter D, Erben P, La Rosée P, Hehlmann R, Hastka J. Systemic mastocytosis simulating osseous metastatic disease. Br J Haematol. 2007;136(1) PMID:17222193
- Deb, A, Tefferi, A. Systemic Mastocytosis. N England Journal of Medicine 2003; 349(7) e7. PMID: 12917319.
- Groheux D. Espie M, Giacchetti S, Hindie E. Performance of FDG PET/CT in the clinical management of Breast Cancer. Radiology; 266 (2) PMID: 23220901

FIGURES



Figure 1: 64 year old female with stage IIB breast cancer who has found to have systemic mastocytosis for a single vertebral body. FINDINGS: A) Medial- Lateral Oblique and B) Craniocaudal views of a diagnostic mammogram show a spiculated mass within the right upper outer quadrant (arrow). Microscopic evaluation of this mass demonstrated infiltrating ductal carcinoma. TECHNIQUE: images obtained on a General Electric Senographe 2000D® Digital mammography unit. Image A) 45 degree MLO angle, breast compressed to 69mm using 100N; rhodium filter: kV 30, mAs 306, rhodium anode: kV55 mAS 239. Image B) CC view compressed to 59mm by 80N. Rhodium filter: kV 29 mas 306, rhodium anode: kV 55, mAs 239.



Figure 2 (left): 64 year old female with stage IIB breast cancer who has found to have systemic mastocytosis for a single vertebral body. During cancer staging, she was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: Diagnostic right breast ultrasound demonstrates a hypoechoic, 2.2 x1.8 x 1.3cm irregular mass (outlined by calipers) with posterior shadowing. Microscopic evaluation of this mass demonstrated infiltrating ductal carcinoma. TECHNIQUE: 12.5 mHz linear array transducer, Philips iU22 xMatrix [®] unit.



Figure 3: 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: A) Anterior and B) posterior whole body and C) magnified anterior and D) magnified posterior Tc99m MDP bone scan demonstrates horizontally oriented, solitary focus of increased radiotracer uptake at the T10 vertebral body (black arrow) without any other osseous abnormalities. TECHNIQUE: Images obtained 3 hours after injection of 20.0 mCi Tc99m MDP on Low Energy, High Resolution Collimator by a Siemens E-CAMS ® dual head gamma camera utilizing a 20% window centered on 140 keV in a 256x256x16 matrix for 1,000,000 counts.



Figure 4: 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: A) Lateral chest radiograph and B) Lateral lumbosacral spine radiographs show degenerative end plate changes throughout the visualized spine without any cortical abnormality or height loss of the T10 vertebral body of interest (arrows).

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Figure 5: 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: A) Axial FDG B) Attenuation correction axial CT and C) Fusion images demonstrate heterogeneous sclerosis of the T10 vertebral body (Image B) without significant FDG avidity. No other FDG avid foci suspicious for metastasis were identified. TECHNIQUE: Images were obtained on General Electric Discover LS ® unit at 90 minutes after injection of 17.0 mCi of F-18-fluorodeoxyglucose with serum glucose 99mg/dL using 6 bed positions for 3 minutes per position to cover the base of the brain to the upper thighs in a 256 x 256 matrix. Iterative reconstruction was performed on the 3D data set into a 128 x 128 matrix by ordered subset maximization (OSEM) algorithm for 30 subsets with 2 iterations using a 7.0mm post reconstruction filter. Concominent noncontrast CT was obtained for attenuation correction using 140 kVp, 120 mAs, 750msec with a 1.75:1 pitch at 3mm slice thickness. Image B and C window level: W 2000 L400



Figure 6: 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. Findings: A) Sagittal FDG B) Attenuation correction sagittal CT reformats and C) Fusion images demonstrate heterogeneous sclerosis of the T10 vertebral body (Image B) without significant FDG avidity. No other FDG avid foci suspicious for metastasis were identified. TECHNIQUE: Images were obtained on General Electric Discover LS ® unit at 90 minutes after injection of 17.0 mCi of F-18-fluorodeoxyglucose with serum glucose 99mg/dL using 6 bed positions for 3 minutes per position to cover the base of the brain to the thighs in a 256 x 256 matrix. Iterative reconstruction was performed on the 3D data set into a 128 x 128 matrix by ordered subset maximization (OSEM) algorithm for 30 subsets with 2 iterations using a 7.0mm post reconstruction filter. Concominent noncontrast CT was obtained for attenuation correction using 140 kVp, 120 mAs, 750msec with a 1.75:1 pitch and reconstructed into 3 mm slices. Image B and C window level: W2000, L400

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Figure 7: 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: A) Coronal FDG B) Attenuation correction coronal CT reformats and C) Fusion images demonstrate heterogeneous sclerosis of the T10 vertebral body (Image B) without significant FDG avidity. No other FDG avid foci suspicious for metastasis were identified. TECHNIQUE: Images were obtained on General Electric Discover LS ® unit at 90 minutes after injection of 17.0 mCi of F-18-fluorodeoxyglucose with serum glucose 99mg/dL using 6 bed positions for 3 minutes per position to cover the base of the brain to the thighs in a 256 x 256 matrix. Iterative reconstruction was performed on the 3D data set into a 128 x 128 matrix by ordered subset maximization (OSEM) algorithm for 30 subsets with 2 iterations using a 7.0mm post reconstruction filter. Concominent noncontrast CT was obtained for attenuation correction using 140 kVp, 120 mAs, 750msec with a 1.75:1 pitch and reconstructed into 3 mm slices. Image B and C window level: W2000 L400



Figure 8 (left): 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. FINDINGS: CT fluoroscopy image during CT guided transpedicular biopsy of the T10 vertebral body of interest. Two 13 gauge core needles were used. Samples obtained with patient in the left lateral decubitus position. TECHNIQUE: Non contrast 1mm axial CT flouroscopy slices obtained at 100mA, 120 kVp, 500 msec, window level: W1500 L 450 on a Siemens SOMATOM® 16 slice scanner.

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Figure 9 (left): 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. High magnification micrograph of tissue sample obtained from CT guided biopsy of the T10 vertebral body (Hematoxylin and Eosin stain, original magnification 200x) shows extensive paratrabecular aggregates of spindle shaped mast cells in the bone marrow.



Figure 10 (left): 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis of a single vertebral body. High magnification micrograph of tissue sample obtained from CT guided biopsy of the T10 vertebral body (Hematoxylin and Eosin stain, original magnification 400x) demonstrates extensive paratrabecular aggregates of spindle shaped mast cells in the bone marrow.





Figure 11 (left): 64 year old female undergoing breast cancer staging who was found to have Systemic Mastocytosis involving a single vertebral body. High magnification micrograph of tissue sample obtained from CT guided T10 vertebral body biopsy. (Mast Cell Tryptase Immunohistochemical stain, original magnification 400 x) was positive for mast cells. These cells were also positive for CD117 and CD25. The morphology and immunohistochemical stains were consistent with Systemic Mastocytosis.

Entity	Non imaging findings	Radiography	СТ	MRI	Bone Scan	FDG- PET
Enostosis (Bone Island)	• Incidentally noted	MetaphysealFeatureless	• Similar to radiographs	 Low signal on all sequences No enhancement 	 Normal or very faint increased uptake 	• No FDG avidity
Bone or Marrow Metastasis	• Known history of cancer (typically breast or prostate)	• Sclerotic or lytic lesion without identifiable margins	 Similar to radiographs with higher sensitivity Bone expansion 	 Low T1 signal Post contrast enhancement 	 Increased uptake 	• Intense FDG avidity
Fracture	• History of osteopenia/ osteoporosis, trauma	 Linear lucency/ sclerosis Periosteal new bone Height loss 	 Similar to Radiographs 	High STIR signalNo enhancement	 Increased uptake 	• Mild FDG avidity
Osteomyelitis	 Fever Elevated ESR, CRP History of systemic infection 	 Thickened sclerotic bone Periosteal bone formation ± Sequestrum 	 Infiltration of surrounding soft tissue Nonuniform, enhancement Sequestrum (+/-) 	 Low T1 High STIR Diffuse enhancement involving multiple levels with disk involvement 	• Increased Uptake	• Moderate FDG avidity
Schmorl's node	• Chronic back pain	 Endplate discontinuity Other Degenerative changes: endplate sclerosis, marginal Osteophytosis 	 Smoothly marinated endplate disruption 	 Low T1 nodule protruding through endplate 	 Mildly increased Uptake 	 No FDG activity
Paget's Disease	 Deep constant pain worse at night Hearing loss 	 Course trabeculae Affects posterior elements 	• Enlarged thickened "picture frame" vertebrae	 Speckled T1 and T2 signal Inhomogeneous enhancement 	 Increased uptake; usually entire vertebra 	No FDG uptake
Sarcoidosis	 Arthropathy Pulmonary symptoms More common in African American females 	 Hilar Adenopathy Lacy sclerotic, lytic or mixed lesions in acral distribution without periosteal reaction 	 Ill-defined osseous "cloud like" sclerosis 	 Homogeneous Low T1 High STIR Enhance with contrast 	 Normal to increased uptake 	• Increased FDG avidity
Systemic Mastocytosis	 Cutaneous lesion Atopy Most common in children 	 Demineralization +/- Fracture Hetergeneous sclerosis 	Similar to radiographs	 Mosaic Low- intermediate T1 Intermediate to high T2 Non enhancing 	Typically photopenic	• No FDG uptake

Table 1: Differential diagnosis table of focal vertebral body uptake on Tc99 MDP bone scan.

Etiology	Mast Cell proliferation due to kit gene mutation		
Incidence	In adults, 5-10/100,000 per year		
Gender Ratio	No gender predilection		
Age Predilection	Peak incidence of cutaneous form: 2 years. Peak incidence of systemic form: 49 years		
Risk Factors	No known risk factors		
Treatment	Depend on symptom severity; range from Antihistamines to immunomodulators or monoclonal antibodies for Atopy. Cytoreductive agents and tyrosine kinase inhibitors may be considered for refractory cases. Progressive disease may indicate chemotherapeutic agents or bone marrow transplant.		
Prognosis	Ranges from excellent in isolated, asymptomatic forms to grave in rapidly progressing patients with leukemic transformation or lymphoid concurrence.		
Imaging Findings	Nonspecific; <u>Radiographs:</u> diffuse osteopenia. Mixed osteosclerotic or osteolytic lesions in a random distribution <u>CT:</u> Osteopenia <u>MRI:</u> Mosaic, low to intermediate marrow signal intensity on T1 and intermediate to high marrow signal intensity on T2 without post contrast enhancement <u>Bone Scan:</u> Solitary uptake in a single focus to diffuse uptake i.e. "Superscan" <u>FDG-PET:</u> typically no significant radiotracer uptake of involved segments		

 Table 2: Summary table for Systemic Mastocytosis.

ABBREVIATIONS

2 CdA = 2-Chlorodeoxyadenosin BMP = Basic Metabolic Panel CBC = Complete Blood Count CRP = C Reactive Protein CT = Computed Tomography DEXA = Dual Energy Xray Absorptiometry ESR = Erythrocyte Sedimentation Rate FDG = Fludeoxyglucose IFN = Interferon MDP = methylene disphosphonte MRI = Magnetic Resonance Imaging PET = Positron Emission Tomography SM = Systemic Mastocytosis STIR = Short Tau Inversion Recovery WBC = White Blood Cell Count WHO = World Health Organization

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KEYWORDS Systemic Mastocytosis; vertebral body; Solitary; Mast Cell; www.RadiologyCases.com