Apparent diffusion coefficient map of a case of extramedullary plasmacytoma

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ABSTRACT

Plasmacytomas are rare tumors, which arise from the monoclonal proliferation of malignant plasma cells. They may affect either the bony skeleton or rarely the soft tissues, the latter being referred to as extramedullary or extraosseous. We report a case of an extramedullary plasmacytoma that presented as a soft tissue mass involving the muscles of the left leg, in a patient who was previously treated for multiple myeloma. We describe the MR Imaging characteristics of the tumor and highlight the usefulness of diffusion-weighted imaging with apparent diffusion coefficient mapping.

CASE REPORT

CASE REPORT

This case is of a 77 year old male patient who was diagnosed with IgG (lambda) multiple myeloma at age 72. Following the initial diagnosis, he was treated with bortezomib and dexamethasone. After completion of treatment, he was in remission for 9 months when he developed anterior chest masses that were biopsied and found to be plasmacytomas. These masses rapidly resolved following chemotherapy with bortezomib and lenalidomide. After another period of remission lasting 2 years, he developed a posterior left thigh mass, for which he was started on carfilzonib and dexamethasone. The posterior thigh mass slowly decreased in size, but the lower leg subsequently became swollen.

An MRI of the left lower extremity was performed 22 days after presentation of the leg swelling to evaluate it. In light of the patient's comorbidities and declining renal function, gadolinium was not administered. The MRI demonstrated two large infiltrative intramuscular solid masses involving the anterolateral and posterior compartments. The masses were homogeneously T1 isointense and T2 hyperintense relative to skeletal muscle (Fig. 1 & Fig. 2). The anterior compartment mass was centered in the extensor hallucis longus and extensor digitorum longus muscles, and extended laterally to involve the peroneus brevis and longus. In addition, it extended posteromedially through the interosseous membrane and involved the popliteus muscle. At this level, it abutted the posterior tibial artery and tibial nerve in the deep posterior compartment. The mass partially encased the fibula and abutted the lateral tibia, which revealed adjacent cortical scalloping. The fibular nerve distal to the fibular tunnel was completely encased by the mass. The mass measured 5.3 x 6.4 x 9.4 cm in the anteroposterior (AP), transverse and craniocaudal (CC) dimensions.

The second mass was centered in the lateral head of the gastrocnemius with similar signal characteristics. It approximated but did not encase the adjacent tibial neurovascular bundle (Fig. 3). It measured 7.9 x 5.6 x 9.4 cm (AP x transverse x CC). The masses both showed high signal intensity on diffusion weighted imaging (DWI) and corresponding low values on apparent diffusion coefficient (ADC) maps, consistent with restricted diffusion (Fig 6 a-c). The ADC values of the tumor ranged from 0.88 to 1.1 as compared to ADC values of approximately 1.7 for regional normal musculature. These imaging findings suggested a differential diagnosis, including extramedullary plasmacytoma,

sarcoma, lymphoma, sarcoidosis, abscess, and muscle infarction.

An ultrasound-guided core needle biopsy of the left leg posterior compartment mass showed diffuse infiltration of the skeletal muscle and fibro-connective tissue by sheets of monotonous neoplastic plasma cells (Fig 10). By immunohistochemistry, the neoplastic plasma cells were positive for CD138, with lambda light chain restriction. There was focal weak expression of CD3, which has been reported in plasma cell neoplasms [1].

The patient was treated with palliative radiation therapy to the left leg and Lenalidomide, to which he responded well with decreasing lower extremity swelling. In this case, clinical examination allowed sufficient assessment of therapeutic response. Hence, post-treatment MRI/ADC maps were not available.

DISCUSSION

Etiology & Demographics:

Plasmacytoma is one form of plasma cell neoplasia that results from dysregulated proliferation of plasma cells, others including multiple myeloma and monoclonal gammopathy of undetermined significance (MGUS). Plasmacytomas may arise from osseous structures (medullary) or soft tissues (extramedullary). Extramedullary plasmacytomas account for only 4% of all plasma cell tumors and the most common site of their occurrence (over 80%) is the upper aerodigestive tract. It usually occurs between the fourth and seventh decades of life, and is more common in men [2]. Cases of solitary extramedullary plasmacytomas affecting the skeletal muscles are rare, as these tumors are usually seen late in the course of relapsed myeloma patients. Imaging descriptions of extramedullary myeloma are restricted to case reports and small case series [3,4].

Clinical & Imaging findings:

The clinical presentation of solitary extramedullary plasmacytoma of skeletal muscles depends on the location and size of the tumor, as well as compression and/or involvement of surrounding structures. These soft tissue masses usually appear hypo- to isointense on T1 weighted imaging and iso- to hyperintense on T2 weighted imaging in comparison to muscle, with heterogeneous enhancement [3,5]. Large tumors may have areas of necrosis and are often associated with infiltration, destruction or encasement of adjacent structures. CT may outperform MRI in detecting subtle bone erosions [5].

DWI is an MRI technique based on the random Brownian motion of water molecules in the region of interest. The ADC value is a measure of the magnitude of diffusion, and can be used to assess the cellularity within a region. The ADC value is generally low in highly cellular tissues as diffusion is restricted by the presence of cell membranes. Conversely, it is high in regions with less cellularity, wherein diffusion is not as restricted. Decreased ADC values are also noted with other processes that increase the viscosity of a region such as cellular damage with fragmentation, as seen in areas of infarction, or the presence of mucoid proteins, cellular debris and bacteria as within abscesses. The DWI sequences can be easily incorporated into the routine MRI protocol with little extra scanning time. Further, in cases where the use of gadolinium-based contrast material is contraindicated (as in this patient), DWI can be useful in differentiating solid from cystic lesions. This technique can provide useful information for characterizing musculoskeletal soft tissue lesions and evaluating response to treatment [6], with longitudinal increases in tumor ADC values thought to reflect histologic necrosis and positive treatment response [7].

Treatment & Prognosis:

Solitary extramedullary plasmacytomas may be treated by surgery alone, however this is often not feasible due to the presence of vital structures adjacent to the tumor. Combining surgery with radiotherapy seems to be the best approach for effective treatment [2]. Alkylating chemotherapy may be used when extramedullary plasmacytoma develops in the presence of a systemic. Chemotherapy or radiation therapy can be used palliatively. Autologous stem cell transplant may be performed in appropriate candidates [8]. However, even after successful therapy, patients need to be under life-long observation for relapses, which may appear even several years later [2]. The median survival time for a patient with pre-existing multiple myeloma who is newly diagnosed with extramedullary plasmacytoma is 24 months, whereas, if extramedullary plasmacytoma is present at the time of diagnosis of multiple myeloma, the median survival time is 12 months [9]. Older patients with extramedullary plasmacytoma have a poorer prognosis, with the 5-year survival decreasing from 78.9% for patients younger than 60 years to 70.5% for those over the age of 60 [10]. Extramedullary plasmacytoma relapse in soft tissues, as our patient had, is unfortunately associated with a dismal prognosis with overall survival being 5 months [11].

Differential Diagnoses:

- Plasmacytoma
- Sarcoma
- Lymphoma
- Muscle infarction
- Sarcoidosis
- Abscess

The imaging findings can be similar among these entities, which exhibit high T2 signal intensity with decreased ADC values compared to normal musculature. Higher grade sarcomas often exhibit internal signal heterogeneity due to varying degrees of spontaneous necrosis, as well as peritumoral soft tissue edema [12]. Recently sarcomas have been shown to have a higher range of ADC values, whereas intramuscular lymphoma shows more homogeneously low ADC values [13], similar to our case (Fig 6). Muscle infarction generally presents with edematous swelling of muscle, with geographic hypoenhancement on post-contrast imaging, unlike the solid enhancement exhibited here. Nodular sarcoidal myopathy exhibits low signal intensity centrally on T2-weighted imaging with contrast enhancement of peripheral area, described as a 'dark star appearance' [14]. Clinical history and course might aid in distinguishing

sarcoidosis and abscess from neoplasm, but ultimately biopsy is necessary for definitive diagnosis.

TEACHING POINT

Extramedullary plasmacytoma, although rare, must be considered in the differential diagnosis when evaluating a soft tissue mass in patients with multiple myeloma. Including Diffusion-Weighted Imaging sequences in the MRI protocol for soft tissue tumors can facilitate identification of highly cellular tumors, manifested as low signal on ADC maps, and aid in selecting tissue targets for biopsy.

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FIGURES

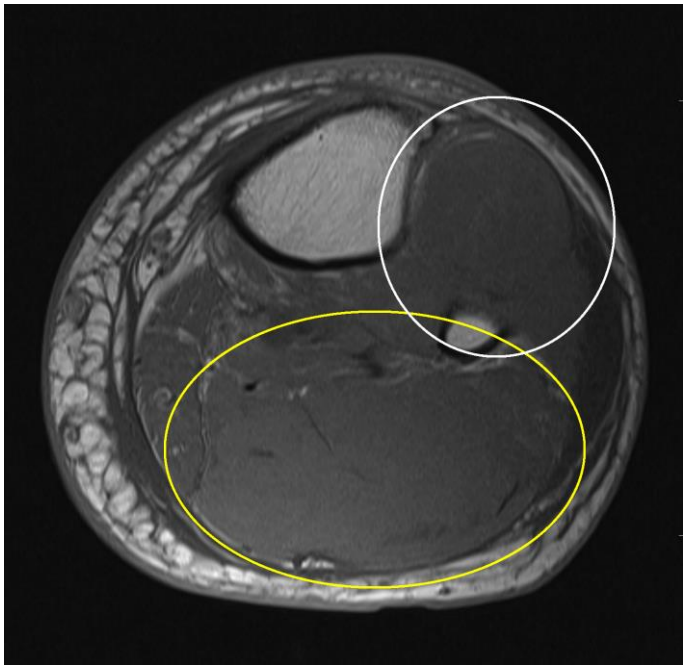


Figure 1: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: Two large infiltrative intramuscular solid masses involving the anterolateral (white circle) and posterior (yellow circle) compartments. The masses appear iso-intense when compared to normal musculature.
Technique: T1-weighted axial image (1.5 Tesla, TR/TE = 679msec/9msec, slice thickness=7mm)

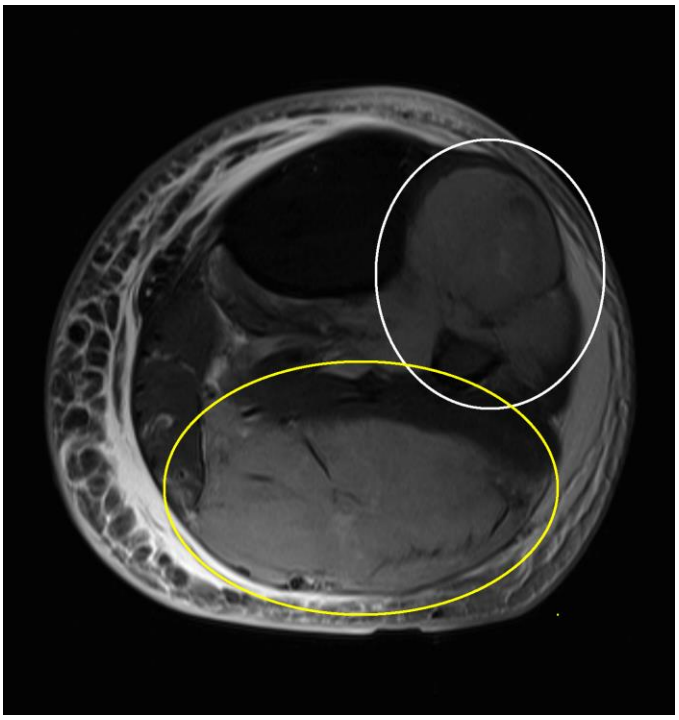


Figure 2: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: The masses appear hyperintense when compared to normal musculature.
The anterolateral mass (white circle) is eroding the outer cortex of the fibula. However, there is no evidence of intramedullary fibular or tibial involvement.
Technique: T2-weighted axial image with fat saturation (1.5 Tesla, TR/TE = 6400msec/87msec, slice thickness = 7mm).

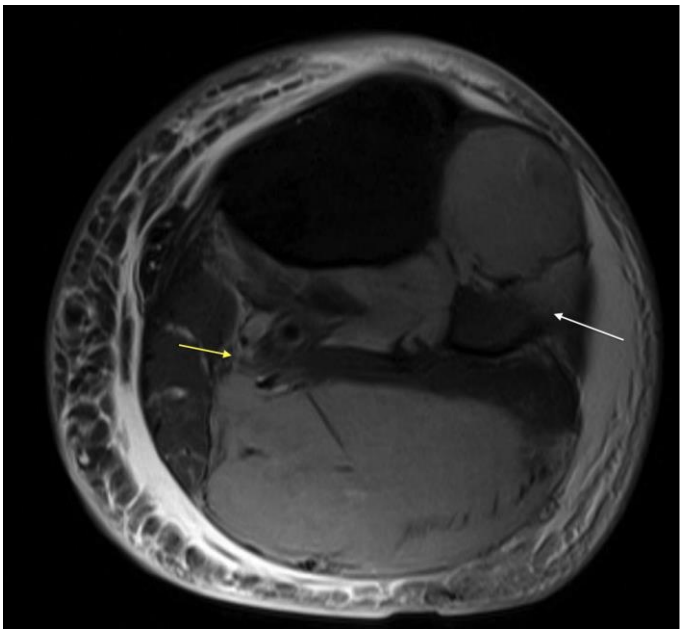


Figure 3 (left): A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: The posterior compartment tumor is abutting, but not encasing the tibial neurovascular bundle (yellow arrow). The anterior compartment tumor is encasing the fibular nerve distal to the fibular tunnel (white arrow; The nerve itself is not well seen due to encasement).
Technique: T2-weighted axial image with fat saturation (1.5 Tesla, TR/TE = 6400msec/87msec, slice thickness = 7mm).

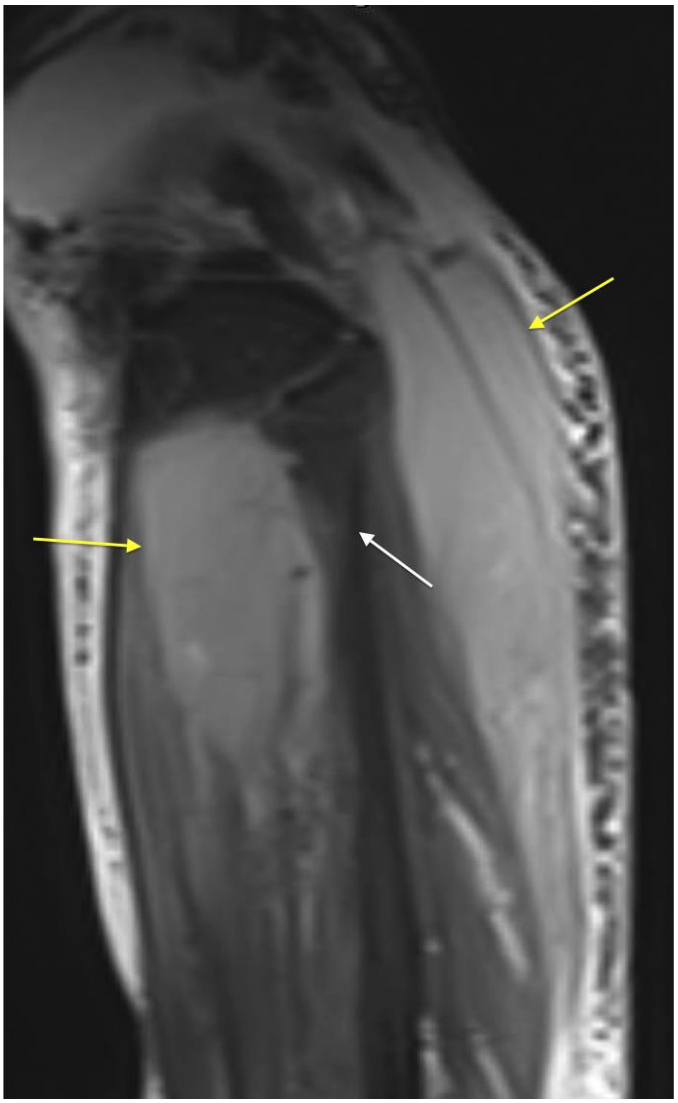


Figure 4: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: Left leg masses (yellow arrows) both posteriorly and anteriorly. The fibula has been marked with a white arrow for orientation.
Technique: Sagittal image from STIR sequence (1.5 Tesla, TR 6800, TE 29, TI 120 ms, 4 mm slice thickness)

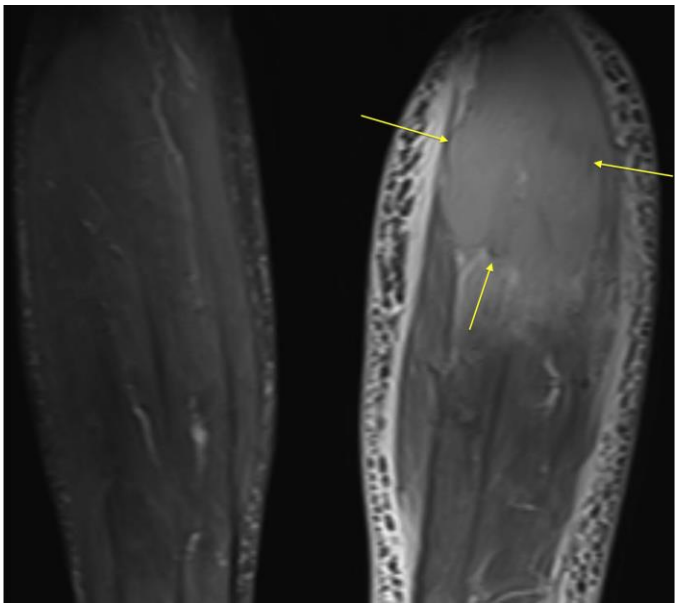
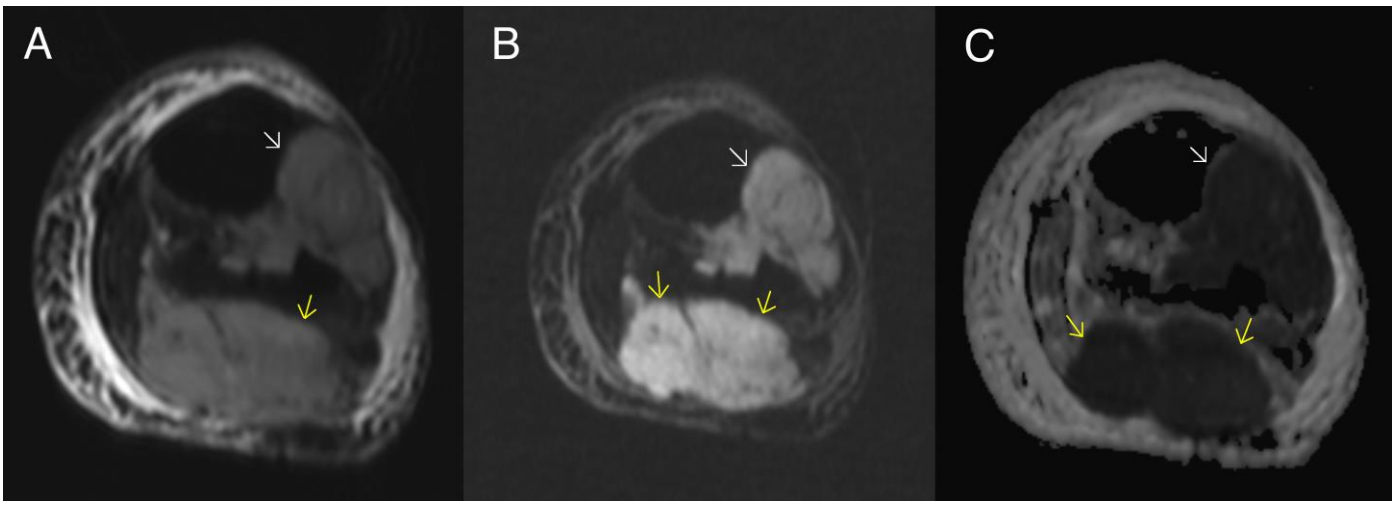


Figure 5: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: T2 hyperintense mass in the posterior calf (arrows), infiltrating adjacent gastrocnemius. There is extensive subcutaneous edema in the left lower leg compared to the right. The patient had a left iliac and femoral vein thrombosis due to extensive disease in the pelvis.
Technique: Coronal image from STIR sequence (1.5 Tesla, TR 6300, TE 29, TI 120 ms, 4 mm slice thickness)

Figure 6 (bottom): A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: The masses (arrows) show high signal intensity on DWI images, with progressively higher relative signal intensity on higher b-value images. ADC map reveals homogeneously low ADC values of the tumors, ranging from 0.88 to 1.1 ×?10-3 mm2/s, as compared to ADC values of approximately 1.7 for regional normal musculature, reflecting their high cellularity. Standard deviation of ROIs constructed within these masses were only 0.07 - 0.08 ×?10-3 mm2/s reflecting tumor homogeneity.
Technique: Fig 3A- Axial DWI with b value = 50 sec/mm2. Fig 3B- Axial DWI with b value = 600 sec/mm2. Fig 3C- ADC Mapping.

Diffusion Weighted Imaging Protocol for imaging of left leg:

Parameter for DWI	**Description**
Plane of acquisition	Axial
Magnetic field strength	1.5 T
Gradient strength	30 mT/m
Sequence	Single-shot multiecho echoplanar (EPI factor = 102)
Slice thickness/interslice gap	7/1.8 mm
TR/TE	8100/86 ms
Field of view	220 mm2
Scanning time (min:sec)	5:48
Number of signals acquired	6
Matrix size	128 x 102
b-values	50, 400, 600



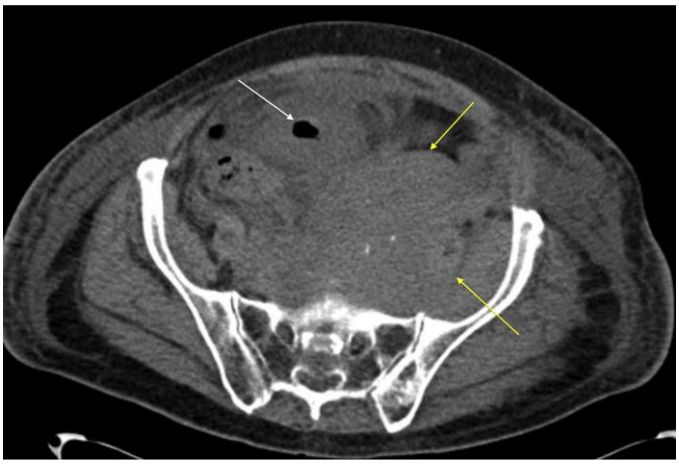


Figure 7: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: Massive extramedullary myeloma in the pelvis and retroperitoneum (yellow arrows). There is air in the displaced bladder (white arrow) due to the presence of Foley catheter. This site was included for radiation therapy, and after treatment the patient reported clinical improvement in left lower extremity pain and swelling.
Technique: Axial noncontrast CT (keV 120, effective mAs 159, 5 mm slice thickness)

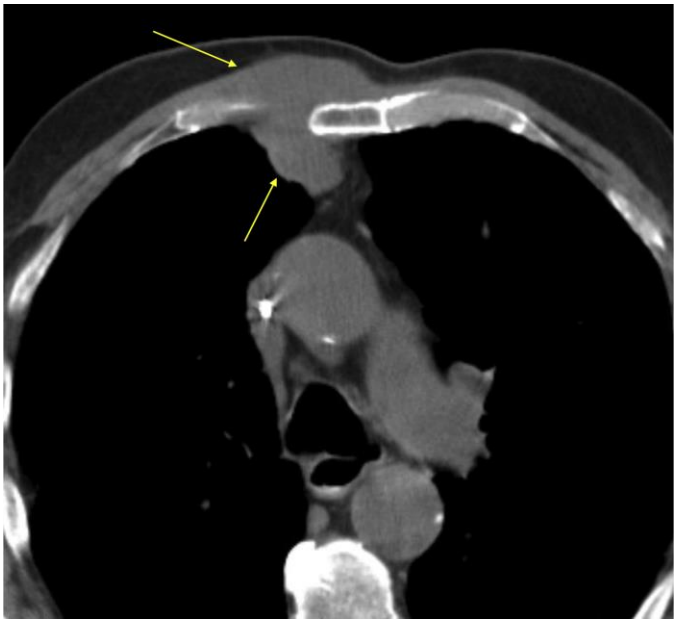


Figure 8: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: CT Chest showing an anterior mediastinal/chest wall mass centered at the right costosternal junction. A biopsy confirmed plasmacytoma.
Technique: Axial noncontrast CT (keV 120, effective mAs 87, 5 mm slice thickness)

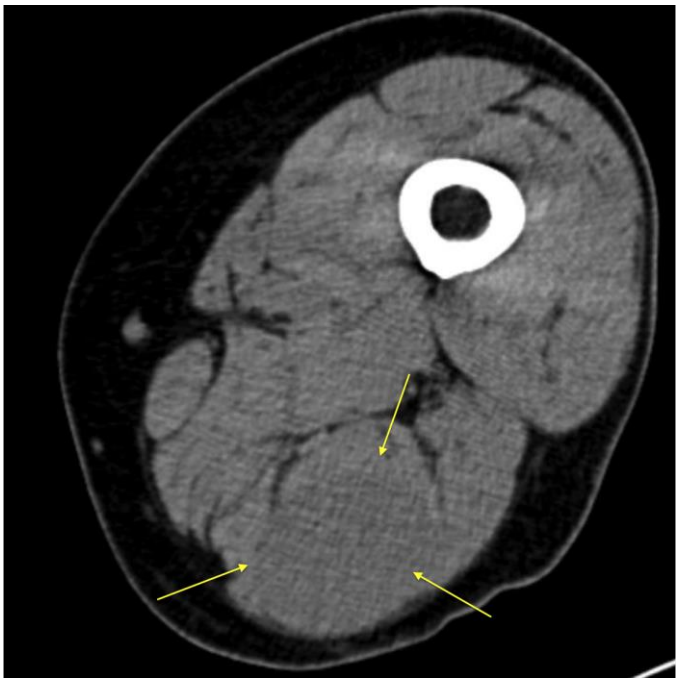


Figure 9: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: CT of the thigh shows a 4.8 x 4.1 cm soft tissue mass (arrows) in the posterior thigh, measuring 38 Hu in density. This was presumed to be extramedullary plasmacytoma, and clinically responded to therapy.
Technique: Axial noncontrast CT (keV 100, effective mAs 128, 5 mm slice thickness)

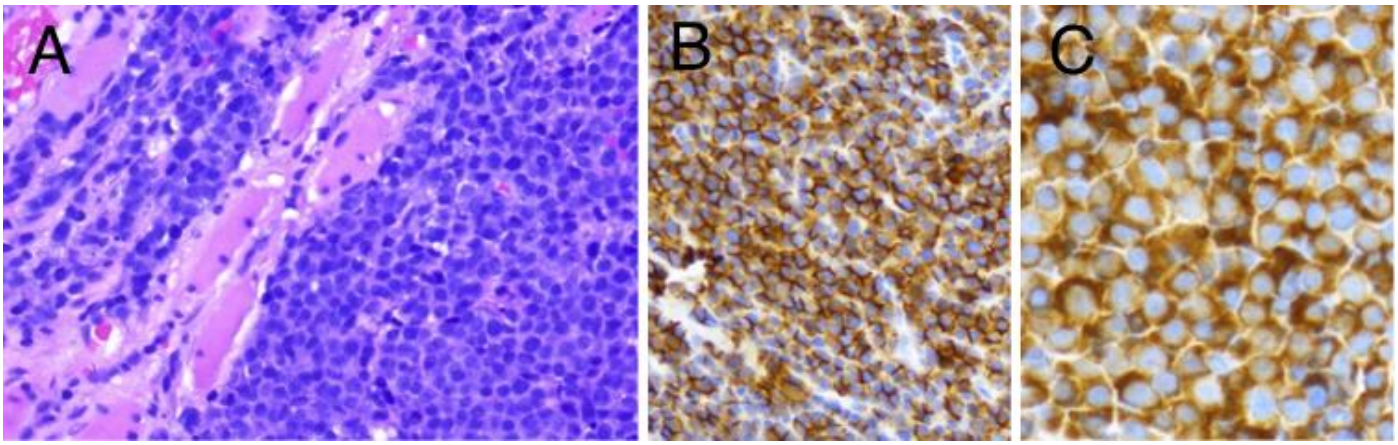


Figure 10: A 77 year old male with extramedullary plasmacytomas of the left leg.
Findings: Fig 4A- Monotonous population of neoplastic plasma cells with numerous prominent nucleoli and some mitoses are seen, splaying apart skeletal muscle bundles. Fig 4B- Neoplastic plasma cells are CD138 positive. Fig 4C- lambda light chain restriction.
Technique: Fig 4A- Hematoxylin and eosin staining of core needle biopsy specimen. Fig 4B and Fig 4C- Immunohistochemistry for CD138.

Etiology	Unknown
Incidence	4% of all plasma cell tumors
Gender ratio	Male preponderance
Age predilection	Mainly occurs between the fourth and seventh decades of life.
Risk factors	Age between 30 to 70 years, male gender, African-American race, family history and exposure to ionizing radiation.
Treatment	Alkylating chemotherapy when seen in context of systemic disease, i.e. multiple myeloma; surgery with radiotherapy for localized disease with adjuvant chemotherapy; chemotherapy or radiation therapy can be used palliatively.
Prognosis	The median survival time for a patient with multiple myeloma newly diagnosed with extramedullary plasmacytoma is 24 months. If extramedullary plasmacytoma is present at the time of diagnosis of multiple myeloma, the median survival time is 12 months. Extramedullary plasmacytoma relapse in the soft tissues is associated with a dismal prognosis with overall survival of 5 months.
Imaging findings	Appears hypo- to isointense on T1 weighted imaging and iso- to hyperintense on T2 weighted imaging in comparison to muscle. ADC values less than that of normal musculature

Table 1: Summary table for extramedullary plasmacytoma.

Diagnosis	MRI	CT	Ultrasound	PET
Plasmacytoma	• T1: Hypo- to isointense • T2: Hyperintense • DWI: Homogeneously decreased ADC values	• Well-defined soft-tissue mass with heterogeneous enhancement	• Solid well-demarcated mass with homogeneous echo texture	• Intensely increased Fluoro-deoxyglucose (FDG) uptake
Sarcoma	• T1: Hypo- to isointense (hyperintense if hemorrhage) • T2: Hyperintense • DWI: Variably decreased ADC values	• Soft tissue density. • Some enhancement with contrast	• Heterogeneous well-defined irregular mass of low to medium echogenicity	• Increased FDG uptake, depending on grade.
Lymphoma	• T1: Hypo- to isointense • T2: Hyperintense • DWI: Homogeneously decreased ADC values	• Iso – or hypoattenuating with variable postcontrast enhancement	• Heterogeneous, hypoechoic solid mass with irregular or poorly defined margins	• FDG avid
Sarcoidosis	• T1: Nodules that are iso- or hyperintense relative to muscle • T2: Low signal intensity centrally with contrast enhancement of peripheral area • DWI: ADC values are not as low as in tumors	• Iso – or hypoattenuating with slight postcontrast enhancement in the peripheral area	• Well-defined nodules with central hyperechoic area and peripheral hypoechoic area	• Increased FDG uptake
Abscess	• T1: Hypointense; may have hypointense capsule or internal gas • T2: Hyperintense; may have hypointense capsule or internal gas • DWI: Variable, can have decreased ADC values	• Iso – or hypoattenuating	• Variable. Colour Doppler demonstrates absence of central perfusion	• FDG uptake elevated.
Muscle infarction	• T1: Hypo- to isointense; geographic hypoenhancement post-contrast • T2: Hyperintense with marked surrounding soft tissue edema and swelling • DWI: Decreased ADC values but limited data	• Diffuse muscle enlargement with decreased attenuation	• Abnormal tissue architecture	• Reduced tracer uptake depending on severity

Table 2: Differential diagnosis for extramedullary plasmacytoma.

ABBREVIATIONS

99m Tc: Metastable nuclear isomer of technetium-99
AFIP: Armed Forces Institute of Pathology
C+: Contrast
CT: Computer Tomography
Gd: Gadolinium
IV: Intravenous
MRI: Magnetic resonance Imaging

KEYWORDS

Extramedullary plasmacytoma; DWI; ADC; Multiple myeloma; MRI

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