

Intramuscular Extramedullary Plasmacytoma in Gluteus Medius Muscle: A Case Report and Literature Review

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ABSTRACT

Plasmacytomas occur in bone marrow or in extramedullary sites, typically in the head and neck. Intramuscular plasmacytoma is rare and reported occurring in the orbits and upper limbs. We report a rare case of intramuscular extramedullary plasmacytoma (EMP) occurring in the gluteus medius muscle. In this case, an elderly lady with known multiple myeloma, presented with a lump in her right buttock. Subsequent imaging demonstrated a non-specific intramuscular mass in the right gluteus medius. Treatment is traditionally surgical resection plus/or adjuvant chemotherapy or autologous stem cell transplant. Plasmacytoma was considered among the differentials, despite the non-specific imaging features, and atypical location. It allowed for a rapid multi-disciplinary team discussion about the risks and benefits of a biopsy. Unfortunately, the patient died before treatment could be started. The purpose of this report is to remind clinicians to include plasmacytoma as a differential for soft tissue masses, even in atypical locations. We also discuss the relevant imaging features and management of EMPs.

Key words: Biopsy, diagnosis, differential, multiple myeloma, muscles, neoplasms, plasmacytoma, plasma cell

INTRODUCTION

A plasmacytoma is a descriptive term applied to tumors consisting primarily of plasma cells. Plasmacytomas are now incorporated in the World Health Organization (WHO) classification and are associated with the production of a monoclonal immunoglobulin or light chains in an extraskeletal area.^[1,2]


In the absence of systemic involvement, the WHO describes two types of plasmacytomas: solitary

plasmacytoma of the bone and extramedullary plasmacytoma (EMP). More than 80% of EMPs arise in the abundant lymphatic tissue of the upper respiratory tract. In 17-33% of plasmacytomas, the extramedullary disease will develop into multiple myelomas (MM). It is mostly seen in head and neck region, but is most frequently in nasopharynx and nasal cavity.

The mean age of growing EMP is 50-55 years, with supremacy of the female sex. Solitary EMPs are highly radiosensitive and many case reports have shown good disease control with external beam radiation therapy, with a 10-year overall survival rate of 70%.^[3]

Plasmacytomas are monoclonal plasma cell neoplasms that are one of the rare plasma cell neoplasms.^[1-5] When they occur in the absence of MM, they are termed solitary plasmacytoma (SP) but can be associated with MM either at the time of diagnosis or with disease

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progression. SP can either occur in bone marrow or soft tissues (extramedullary). EMP have a predilection for the head and neck, particularly the upper aerodigestive tract with few cases involving the rest of the body.^[6] We report an intramuscular EMP occurring in the gluteus medius muscle in a 76-year-old female. Intramuscular EMP is extremely rare, and we emphasise the need to consider plasmacytoma in the differential diagnosis of soft tissue masses even in atypical locations.

CASE REPORT

A 76-year-old Caucasian female with a 5 years history of immunoglobulin A lambda myeloma associated with significant Bence-Jones proteinuria was being managed actively with the following chemotherapy: Melphalan, cyclophosphamide, dexamethasone, and lenalidomide. She presented to the hematologists with right buttock pain. On clinical examination, a palpable lump was felt in the right gluteal region, and she was referred for imaging.

Ultrasound (US) revealed a 6 cm × 3 cm mixed echoic lesion within the right gluteus medius [Figure 1]. Contrast-enhanced multidetector row computed tomography (CT) demonstrated a solitary focal high attenuation intramuscular lesion [Figure 2]. After multi-disciplinary team (MDT) discussion, an ultrasound-guided biopsy was performed. This revealed plasmacytoid cells with eccentric nuclei and a moderate degree of pleomorphism. The cells were positive for a number of plasma cell markers on immunohistochemistry, consistent with a plasma cell neoplasm [Figure 3]. Palliative management was decided, and local radiotherapy was planned. Unfortunately, she died a few weeks later.

DISCUSSION

Plasmacytomas can occur in the absence of MM, or can be a complication of myeloma either at diagnosis or with disease progression.^[4] When they occur alone, they are termed SP. SP is a plasma cell tumor without the features of disseminated myeloma. When a SP occurs in the bone it is termed as a solitary bone plasmacytoma (SBP). When present outside the marrow, it is termed as an EMP. The spectrum of plasma cell disorders includes SP, MM and plasma cell leukemia.^[5,6] The nature and prognosis of the spectrum of plasma cell dyscrasias differ, prompting some to consider these as being separate entities.^[7] SPs are rare with SBPs affecting approximately

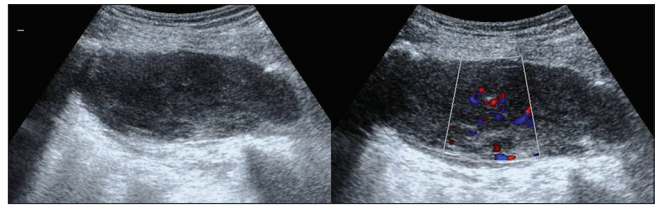


Figure 1: Ultrasonography demonstrating well-defined intramuscular lesion with increased vascularity on doppler

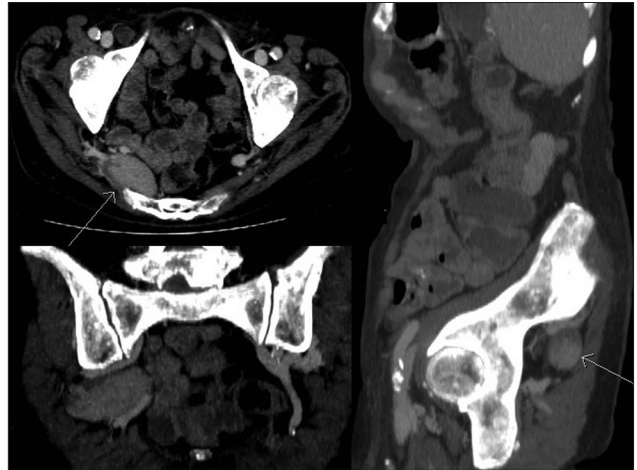


Figure 2: Multidetector row computed tomography axial, coronal, and sagittal reforms showing enhancing lesion within the right gluteus medius (arrow)

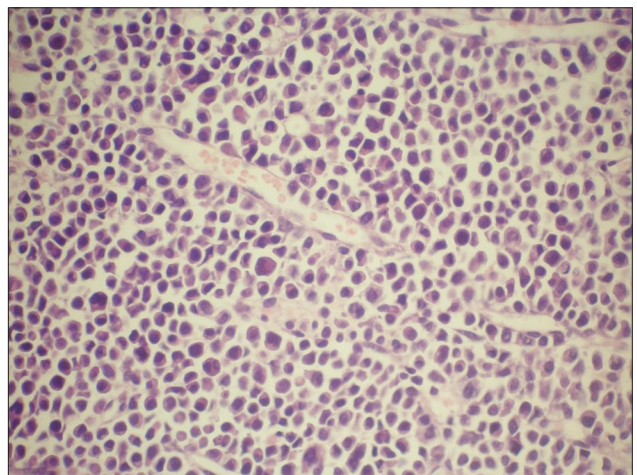


Figure 3: Histology showing plasmacytoid cells with eccentric nuclei with atypia

5% of patients with plasma cell disorders and EMPs representing approximately 3%.^[8,9] SBPs are seen most commonly in the axial skeleton with 90% of EMPs observed predominantly in the head and neck, commonly in the nasal cavity, sinuses, and nasopharynx.^[10] Other documented sites include the liver, vagina, pancreas, retroperitoneum, skin, breast, and gastrointestinal tract. The median age of the patients with either SBP or EMP is 55 years, in contrast to MM, which is seen in patients approximately 10-15 years older.^[9] It has been reported

that the incidence rate of SP in the Afro-Caribbean population is around 30% higher than in Caucasians.^[11]

Muscular involvement from adjacent bone lesions is common in MM. However, isolated de novo intramuscular plasmacytoma is rare, reported as 0.6%^[12] rising to 2.2% when associated with MM.^[13,14] Lesions may present as solitary masses or diffuse skeletal involvement. Documented sites include orbital muscles^[10] and the upper limb^[15-17] with nine different sites observed by Surov *et al.*^[18] with only two cases involving the gluteus muscle.

Differential diagnosis for a solitary intramuscular mass includes granuloma, sarcoma, neurofibroma, lymphoma, and metastases.^[19] In the appropriate clinical setting, abscess should also be considered.

US features of EMPs are variable with some demonstrating increased signal on color Doppler.^[5,7,20,21] Posterior acoustic enhancement, necrosis and calcification may also be seen.^[5,7,21,22] They are locally invasive and can demonstrate local infiltration into adjacent fat, bone, and even encase the vessels.^[3] The features on CT and magnetic resonance imaging are non-specific with variable enhancement and cannot be differentiated on imaging from carcinoma, sarcoma, and neurofibroma. As the appropriate clinical history was provided to radiology, despite the non-specific imaging features, a more relevant differential diagnosis was considered. This allowed an appropriate MDT for a timely diagnosis. Unfortunately, our patient succumbed to her disease and died.

A biopsy of the lesion, demonstrating plasma cell infiltration, is essential for confirming the diagnosis.^[1-6] However, careful planning is required; Kansar *et al.* described seeding of plasma cells in the tract following a percutaneous bone marrow biopsy.^[22,23] The primary role of imaging is, therefore, to identify other lesions, local infiltration, and lymphadenopathy.^[1,20] As this case was discussed in the MDT setting, appropriate expertise was sought to allow a biopsy to be undertaken safely.

Plasmacytomas are radiosensitive and the management is aimed to attain local control with the use of local radiotherapy. Surgery and a combination of radiation and surgery are the other options, which are primarily dependent upon the site of SP.^[3,20] Chemotherapy and marrow transplantation are reserved for recurrence, refractory, and multiple plasmacytomas.^[1-3] SBP

respond much more favorably to treatment in comparison to multiple plasmacytomas or to those that have adjacent nodal involvement and have a 5-year survival of 50%.^[1,2] A third of patients with SP develop features of MM subsequently.^[1] If a SP is associated with MM, prognosis is poor with limited treatment options, which often fail, as seen in our case.

CONCLUSION

We have presented a rare case of intramuscular EMP occurring in an atypical site. Our case highlights the non-specific imaging features of plasmacytomas. Although plasmacytomas commonly occur in the head and neck region, they should be included in the differential diagnosis of all solitary soft tissue masses, even in uncommon locations. The use of a MDT allows experts from various specialties to discuss the relevant investigations, treatments, and potential complications. Surgeons must also be alert to clinical signs of progression to MM, such as anemia, bone pain, and renal insufficiency, particularly in cases of relapse.

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