

Case study

SOFT TISSUE PLASMACYTOMA : SURGERY, RADIOTHERAPY OR IMMUNOCHEMOTHERAPY ?

Abstract

Soft tissue plasmacytoma is a rare tumor. It belongs to the extramedullary plasmacytomas. Diagnosis is based on biopsy, and treatment is mainly based on radiotherapy. Immunochemotherapy followed by autotransplantation, surgery are a possible weapon in its management.

We report the case of a 39-year-old man who had a thigh plasmacytoma on a femur fracture 10 years ago, diagnosed on biopsy, and treated with radiotherapy alone.

Key words: plasmacytoma; soft tissue; radiotherapy; immunochemotherapy; surgery

Introduction

Soft-tissue plasmacytoma is a rare monoclonal proliferation of plasma cells in soft tissues or organs[1]. The relationship between multiple myeloma, solitary bone plasmacytoma and soft-tissue plasmacytoma is not well understood. For some authors, these 3 entities represent different aspects of the same disease[2]. The incidence of soft-tissue plasmacytoma at diagnosis varies from 1.7% to 4.5% [3]; with the skin being the most frequent site, followed by the liver, pleura and CNS[4]. We report an observation of soft-tissue plasmacytoma and attempt to identify the main features of this entity through a review of the literature.

Case presentation :

This was a 39-year-old patient with a history of a left femur fracture operated on in 2013, treated with a screw-plate and orthopaedic cement, who presented with a symptomatology of progressive onset evolving for 6 months, made up of a painful swelling of the left thigh, more marked at the fracture

site. Clinical examination revealed a patient with a PS (statural performance) of 1, with a hard, firm, painful swelling 16cm long on the left thigh.

MRI of the left thigh revealed a tumoral mass in the middle third of the left thigh measuring 11*8cm by 18cm (Figure 1).

Soft tissue biopsy was consistent with a plasmacytoma with Kappa monotypy; CD138+, MUM1 +, CD45-, ERG-, CD20-, CD3-, CD99-(Figure 2).

The PET scan showed a hypermetabolic tissue process in the lower half of the left thigh with an SUV max of 9.05, and no suspicious hypermetabolism over the rest of the volume explored (Figure 3).

Bone radiology was normal, with normocytic normochromic anaemia at 8g/dl, normal renal function tests and serum calcium levels. Serum protein electrophoresis showed a narrow monoclonal band migrating into the gamma zone at 30g/l, with Kappa IgG immunoglobulin. Light chain assay showed a Kappa chain at 117mg/l, and a Lambda chain at 34.7mg/l; with a K/L ratio of 3.38. Myelogram, osteomedullary biopsy and 24-hour proteinuria were normal. Multidisciplinary management of our patient involved traumatologists, hematologists, radiologists, nuclear medicine physicians and radiation therapists. The decision of the RCP was to receive radiotherapy sessions only on the tumor mass.

Discussion

Multiple myeloma is the most common type of malignant plasma cell proliferation. Myeloma cells can form tumors, called plasmacytomas. Plasmacytomas can sometimes arise after surgery, from laparotomy scars or catheter insertions [5], as in our case.

It is possible that the inflammatory process of a wound may facilitate the migration of myeloma cells into the skin, creating a reservoir for proliferation [6].

The appearance of a soft-tissue plasmacytoma along a surgical scar 10 years prior to diagnosis was reported in a patient with multiple myeloma [7].

Positive diagnosis is essentially based on immunohistochemical studies.

MRI is useful in assessing the nature of the plasmacytoma (bony or extraosseous) and its extent.

Positron emission tomography is the most useful whole-body technique when soft tissue involvement is suspected [8], with a sensitivity of 96% and specificity of 87%. Surgical resection of plasmacytoma is generally not necessary, as these malignant tumors are radiosensitive. For the most part, surgical excision is reserved for cases where there is a loss of anatomical structural integrity or spinal cord compression [9]. If surgery is performed, it is usually carried out prior to radiotherapy and as a complement to definitive radiotherapy.

Treatment of plasmacytoma is therefore essentially based on radiotherapy. A response rate of 80-90% can be achieved with radiotherapy alone [10].

Immunochemotherapy has also been proposed to improve disease control and prevent progression to multiple myeloma following radiotherapy, although data are largely insufficient [11].

However, VMP or VRD regimens combined with Daratumumab appear to be the treatment of choice for patients who are not eligible for autotransplantation [12].

Intensive treatment with VTD or VRD/cisplatin, doxorubicin, cyclophosphamide and etoposide (PACE) is offered to patients eligible for autotransplantation.

The International Myeloma Group (IMWG) uniform response criteria require disappearance of soft-tissue masses for complete remission (CR) and $\geq 50\%$ reduction for partial response (PR) [13].

The presence of soft-tissue plasmacytoma is associated with significantly shorter progression-free survival and overall survival, although the incidence is only 6% and 10% respectively [14].

Conclusion :

Soft-tissue plasmacytoma is a rare tumor; its management is multidisciplinary. Radiotherapy remains the mainstay of treatment. The risk of local and distant relapse, and of transformation into multiple myeloma, warrants regular monitoring.

References

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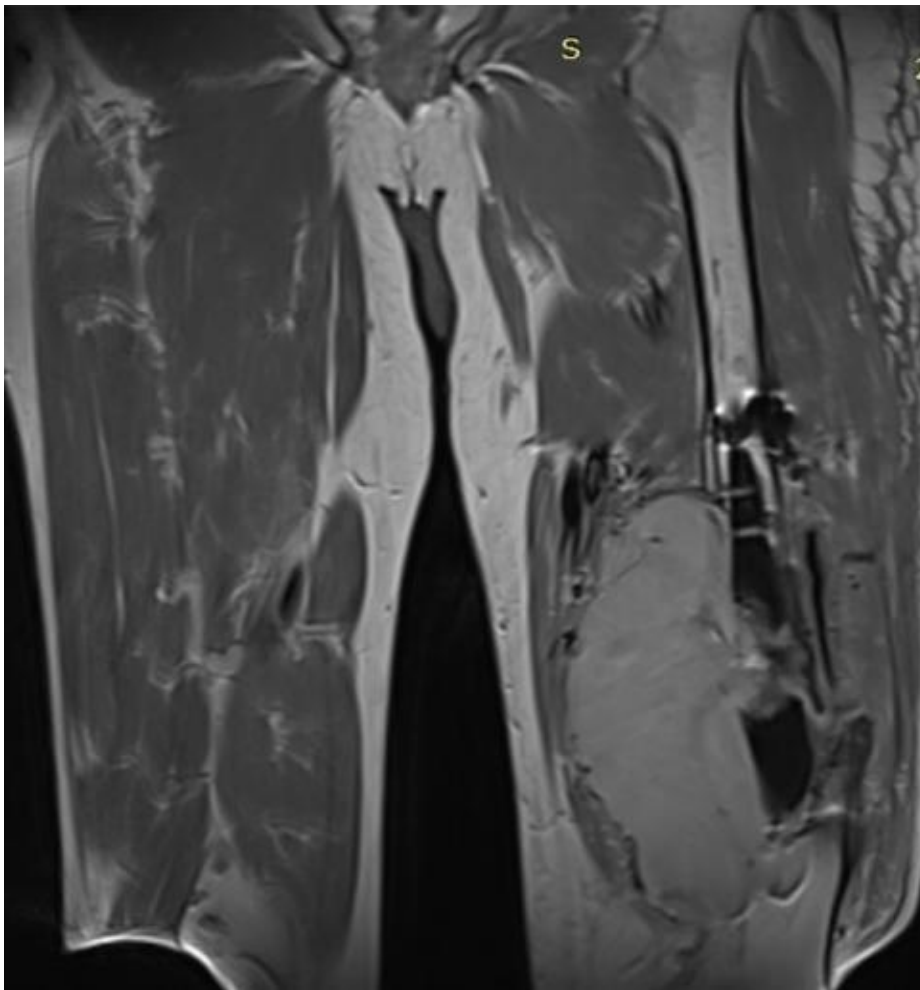


Figure 1: T2 sagittal section of a tumor mass in the middle third of the left thigh.

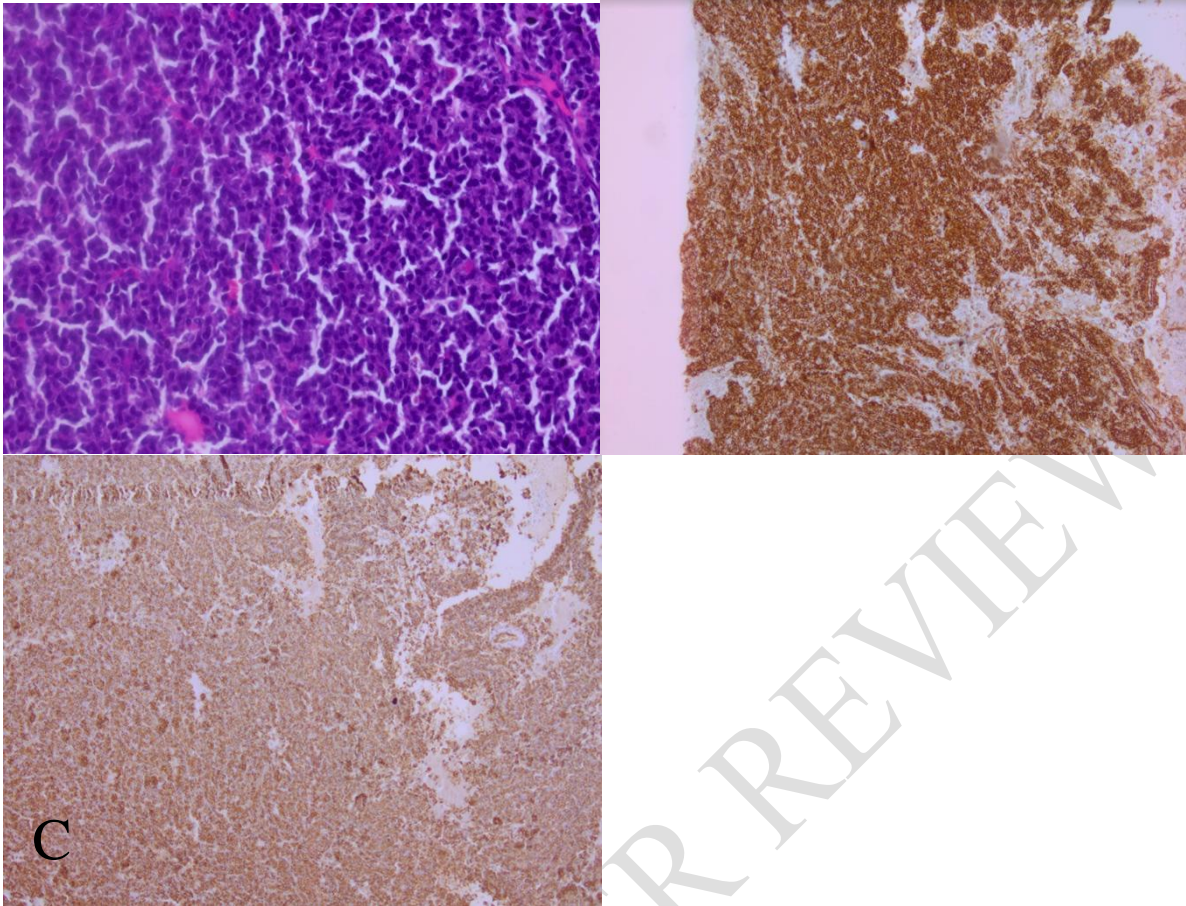


Figure 2: A: Histological appearance of plasmacytoma (HES: 10×40): well differentiated, monomorphic, plasma cell-like cells. B: Immunohistochemical appearance (IHC: 10×4): strong, intense, diffuse membrane expression of CD138. C: Immunohistochemical appearance (IHC: 10× 4): Plasma cells also express monotypic IgG Kappa.

