# **Clinical Orthopaedics and Trauma Care**

Case Report

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## Exophytic Soft Tissue Sarcoma Versus Cutaneous Carcinoma. A Challenge in Orthopedic Oncology. Case Report

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#### **Abstract**

Soft tissue sarcomas (SPB) are heterogeneous tumors of mesenchymal origin that include more than 100 different subtypes. The great histological variability and its infrequency make diagnosis a challenge, as shown by the series of 1463 patients with sarcomas by Ray-Coquard et al., carried out in 2016 with a lack of concordance of 43%.

**Keywords:** auxetic structure; cross chiral structure; variable stiffness; musculoskeletal assistive structure

### Introduction

Soft tissue sarcomas (SPB) are heterogeneous tumors of mesenchymal origin that include more than 100 different subtypes (1,2,3). The great histological variability and its infrequency make diagnosis a challenge, as shown by the series of 1463 patients with sarcomas by Ray-Coquard et al. carried out in 2016 with a lack of concordance of 43% (4).

Merkel cell carcinoma (MCC) is an aggressive neuroendocrine carcinoma of cutaneous origin with a high rate of metastasis and an overall 5-year survival of close to 50% (5). This lesion is predominantly located in photo-exposed areas and has an average size of 2-4 cm (6).

The case that we are going to report is that of a MCC referred to our institution with a presumptive diagnosis of exophytic soft tissue sarcoma in the gluteal region with a diameter greater than 13cm.

### Case presentation:

Our patient is a 66-year-old man referred from another institution, who consulted the orthopedic oncology clinic due to a tumor in the left gluteal region of approximately 2 years of evolution, large-volume exophytic, stony consistency and purple in color, slightly painful on palpation and with rapid growth in recent months (images 1 and 2).

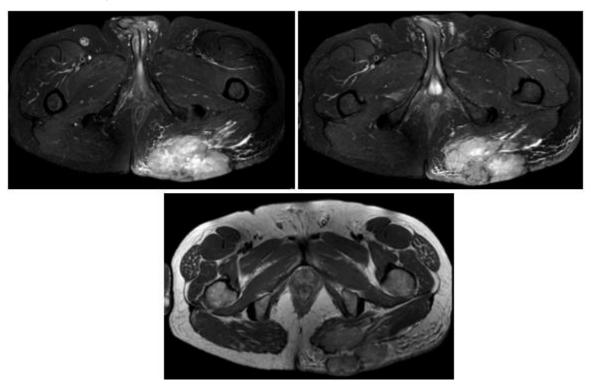




Figure 1 and 2: admission image: exophytic purple stony tumor measuring 130 x 66 x 104 mm (according to CT)

#### Imaging studies were initially performed:

**Resonance**: Polylobulated mass of heterogeneous signal intensity that involves the subcutaneous cellular tissue (TSC) and belly of the gluteus maximus muscle in its medial sector. It presents post-contrast enhancement and restricts diffusion (atypical appearance). Its measurements are 121x69x83mm. No lymph nodes are observed (images 3 - 5).



**Figure 3, 4 and 5:** MRI of the pelvis with and without contrast in which a polylobulated mass of heterogeneous signal intensity is observed that involves the subcutaneous cellular tissue (TSC)

*CT chest*: Both lung fields without nodular lesions, of primary or secondary atypical appearance, or areas of consolidation of the parenchyma.

*CT abdomen and pelvis*: abdominal cavity without particularities. Polylobulated mass of soft tissue density involving TSC and left gluteus maximus muscle of primary atypical appearance. It presents heterogeneous enhancement after contrast. Its measurements are 130x66x104 mm.

Suspecting a soft tissue tumor with extension to the skin, an incisional biopsy was performed for a pathological study, the analysis of which reported: infiltrating atypical neoplasm with neuroendocrine features, linkable to MCC (images 6 and 7).

The histopathological analysis together with the MRI images provided the key to interpreting that said tumor did not appear to have a soft

tissue origin (with subsequent cutaneous invasion), but suggested that it correspond to an originally cutaneous lesion, progressing over time with deep underlying infiltration. These imaging characteristics, together with the histological characteristics (neuroendocrine features), led to the suspicion of MCC. This histopathological diagnosis was confirmed by routine techniques and immunohistochemical markings (- cytokeratin 7: (-) negative, - cytokeratin 20: (+) paranuclear positive, - Pancytokeratin: (+) positive, - CD3: (-) negative, - CD 20: (-) negative, - Racemase: (-) negative, - Prostate Specific Antigen: (-) negative, - CD 99: (-) negative, - Desmin: (-), - Mygenin:

Based on these findings, the interpretation of a deep tumor that compromises the skin barrier was changed to a skin tumor that invades the gluteal region. With this new diagnostic suspicion, the specific markers () were made that allowed the diagnosis to be reached.

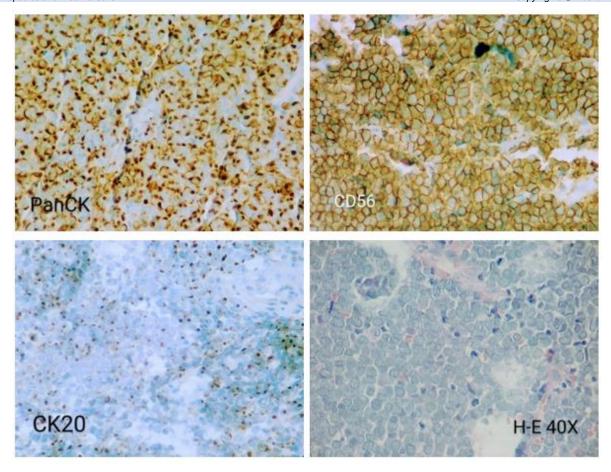


Figure 6, 7, 8 and 9: marking of the histopathological sections and their immunohistochemical markings.

After the interdisciplinary meeting, it was decided to carry out chemotherapy treatment (4 cycles of doxorubicin, cyclophosphamide, vincristine) and the subsequent excision of the tumor.



**Figure 10:** post-chemotherapy clinical image. Image 9 preoperative (x days post-chemo).

The excision of the tumor presented insufficient margins since it was in contact with the rectum. After this, with and despite various attempts to improve survival, the patient died 18 months after the first consultation.

### **Discussion**

In the international literature we have found only one case with the same characteristics and location, as reported by Dr. Raelina Howell (7) in her article it is necessary. Merkel tumors are painless tumors

found in photo-exposed areas (for example, the head), neck and extremities) and with an average size it is usually 2-6 cm. Therefore, we can deduce that a non-painful tumor in unusual areas can trigger different behavior or a delay in the consultation.

The low frequency of soft tissue sarcomas and their great heterogeneity associated with a tumor, in this case, atypical and advanced MCC represent a diagnostic challenge and delay their effective treatment.

We want to emphasize that a fluid dialogue between specialties is essential to interpret low-prevalence pathologies.

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