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# Primary sclerosing epithelioid fibrosarcoma presenting as lombosciatic syndrome: Case report and literature review

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## ABSTRACT

**INTRODUCTION AND IMPORTANCE:** Sclerosing epithelioid fibrosarcoma (SEF) is a rare variant of low grade fibrosarcoma, with specific histological and immunohistochemical features. SEF is a difficult to diagnose. The prognosis is poor with a 40% mortality rate.

**CASE PRESENTATION:** We report a case of 45-year-old female patient who presented to our department with a history of right sciatalgia evolving for three months. On physical examination, a firmly not well-defined mass was found in the right gluteal region. The histological diagnosis revealed a SEF.

**CLINICAL DISCUSSION:** SEF appears to be a slowly growing tumor often present for several months or years before diagnosis. The 3-month delay of our diagnosis shows the difficulty arising from the inconclusive clinical of this tumor.

**CONCLUSION:** SEF of the gluteal region can induce sciatalgia. The diagnosis should be made as early as possible in order to improve the prognosis.

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## 1. Introduction

Sclerosing epithelioid fibrosarcoma (SEF) is a low-grade variant of fibrosarcoma. It was first described by Meis-Kindblom [1] in 1995 as an uncommon and aggressive sarcoma. It typically takes several months from the first onset of symptoms to diagnosis. The metastasis may be common to find on presentation. To our knowledge, only a case of low-grade fibromyxoid sarcoma that presented with sciatalgia has previously been reported [2]. We here describe a case of the right gluteal region SEF with the treatment details.

This study is reported in line with the SCARE checklist [3].

## 2. Case presentation

A 45-year-old female patient with no past surgical history presented to our department with a history of an L5 sciatalgia without sensory-motor deficit evolving for three months. She didn't use any kind of drugs. Her family members were healthy. On physical examination, a firm ill-defined mass approximately 11 × 7 cm was found in the right gluteal region. Magnetic resonance imag-

ing showed a solid mass with a necrotic center in the right gluteal region extended to the great sciatic notch but respecting the great sciatic nerve and the initial part of the right gluteal artery (Fig. 1). Contrast computed tomography (CT) of chest, abdomen and pelvis revealed a secondary right Fowler lung nodule (Fig. 1).

Biopsy was done, which showed diagnosis to be Sclerosing Epithelioid Sarcoma (SEF).

Surgery was performed under general anesthesia and intubation. It was carried out by the orthopedic surgery team of Habib Bougatfa Hospital. A surgeon of the team have an experience in oncologic surgery.

The patient was placed in the left lateral position. A direct approach to the tumor was performed taking the path of the biopsy. A compartmental resection was carried out removing the three gluteus muscles and part of the tensor muscle Fascia lata (Fig. 2). The resected specimen was of 17 × 13 × 9 cm and was submitted for pathological examination (Fig. 2). The patient was given postoperative analgesic treatment and then was referred for chemotherapy. No immediate complication was noted.

The histological study shows a malignant mesenchymal proliferation of heterogeneous cellularity with densely cellular areas and other sparsely cellular myxoids. This proliferation is arranged in nests and rows of epithelioid cells of small sizes, with sparse eosinophilic cytoplasm and vesicular nucleus ovoid and round. These cells take up an alveolar arrangement in places. The stroma is abundant, made of sometimes thick collagen vessels. Large areas of tumor necrosis are observed. The limits of surgical resection

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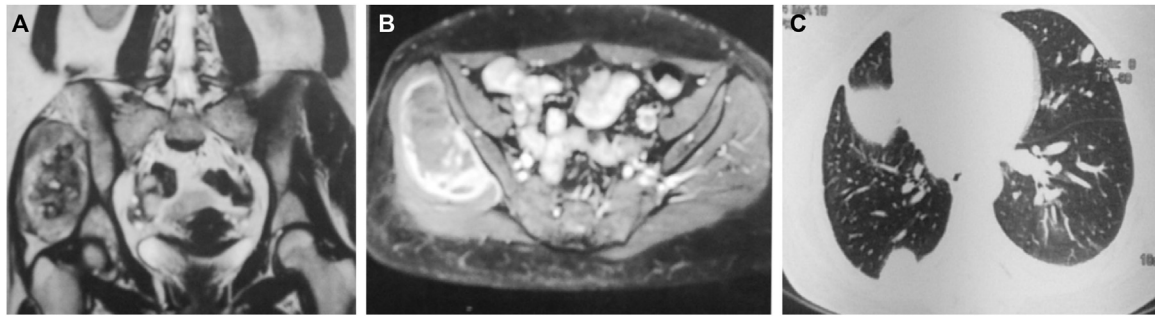


Fig. 1. A/B Magnetic resonance imaging of pelvis showing a mass in the right gluteal region. C Computed tomography (CT) of chest a secondary right Fowler lung nodule.

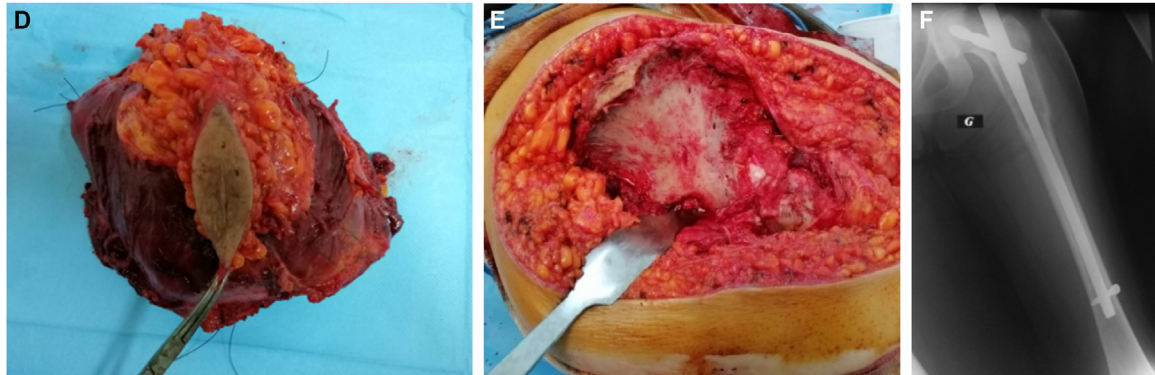


Fig. 2. D Intraoperative photograph showing the right gluteal region after resection. E Specimen photograph. F X-ray of a proximal femoral metastasis treated with nail.

on the side of the iliac spine antero-superior, greater trochanter, and antero-inferior iliac spine were healthy. The limit of deep surgical resection is infra-millimeter. Immunohistochemical pattern included slightly positive for EMA while negative for PSA, CLA, CD34 and CK. The pathology examination was consistent with sclerosing epithelioid fibrosarcoma (SEF).

One year later, the patient was operated for pulmonary metastases (lower right lobectomy) and she refused chemotherapy. She was admitted a year later for local recurrence with pleuropulmonary and bone metastasis. She had nailing of the left femur (Fig. 2). The patient died due to generally late metastasis.

### 3. Discussion

According to our knowledge and our research in the literature only one case had been reported a patient with low-grade fibromyxoid sarcoma in the gluteal region presenting as sciatic nerve pain [4]. The specificity of the present case is that the sciatalgia was caused by the compression of the big tumoral mass.

Sclerosing epithelioid fibrosarcoma (SEF) is a rare soft tissue tumor with similar morphological and molecular characteristics to low-grade fibromyxoid sarcoma (LGFMS). Although SEF is histologically classified as a low-grade sarcoma, it is clinically aggressive. SEF was first described by Meis-Kindblom et al. in 1995 [1] as a tumor involving mainly deep musculature, associated with fascia or periosteum, that microscopically simulated an infiltrating carcinoma. It's composed of epithelial tumor cells arranged in nests embedded in a hyalinized fibrous stroma. The sclerosing epithelioid fibrosarcoma is a clinicopathologically distinct low grade fibrosarcoma that is closely related to low grade fibromyxoid sarcoma [4,5] and hyalinising spindle cell tumour with giant rosettes [6,7]. These three tumours belong to the same family of so called "fibrosing fibrosarcomas". The average age range was 45 years, with a slight male predominance [1]. The most frequent sites of involvement are deep soft tissues of lower extremities or limb girdle followed

by trunk, upper extremities, and head and neck region, although primary SEF of unexpected locations such as the base of the penis [8], bone [9], and kidney [10] and pelvis were also described as case reports or small series [2,11,14]. Primary SEF in visceral organs is exceedingly rare, with only a single case reports in the liver [13], the lower gastrointestinal tract [12], the ovary [15], and the pancreas [11].

SEF is a difficult to diagnose because of its rarity and its epithelioid appearance, closely mimicking carcinomas. It is characterised histologically by a proliferation of uniform, small, slightly angulated, round to oval epithelioid cells with sparse, often clear cytoplasm arranged in nests and cords, associated with prominent hyaline sclerosis of the stroma. There may be myxoid areas with cyst formation, foci of hyaline cartilage, calcification, and metaplastic bone.

Neoplastic cells are positive for vimentin, bcl-2, mucin-4 (MUC4), weakly and focally for epithelial membrane antigen (EMA), and are negative for broad spectrum cytokeratins, smooth muscle actin, desmin, CD34, S-100 protein, HMB45 and melan-A [16–18]. Doyle et al. identified MUC4 as a highly sensitive and specific immunohistochemical marker for low-grade fibromyxoid sarcoma [16].

FUS gene rearrangement as detected by fluorescence in situ hybridization, as well as FUS-CREB3L2/FUS-CREB3L1 chimeric fusion genes detected by reverse transcription polymerase chain reaction, are other reliable approaches for diagnosing low-grade fibromyxoid sarcoma [19].

The differential diagnosis of low-grade fibromyxoid sarcoma includes nodular fasciitis, myxoma, desmoid fibromatosis, perineurioma, neurofibroma, dermatofibrosarcoma protuberans, ossifying fibromyxoid tumor, and low-grade myxofibrosarcoma.

Our diagnosis of Sclerosing epithelioid fibrosarcoma (SEF) was based on all our histochemical findings.

Patients that were metastatic at diagnosis tended to be those with large deep-seated tumours located centrally [20]. The asymp-

omatic nature of SEF coupled with the large primary tumour size suggested a long-standing malignancy and is in keeping with the possibility of SEF slowly developing the ability to metastasize over-time in these patients.

One or more local recurrences occur approximately in 50% of cases, with metastatic spread being reported in more than 40% of cases, most often affecting the pleura, lungs, bone and central nervous system [1,21]. The local recurrence rate is about 50%, with an average delay of 3.5 years. The factors of poor prognosis are as follows:

- occurrence in humans
- a proximal site
- large tumor size
- the presence of tumor recurrence
- metastases [1,22]

In our case, the patient was dead due to metastasis despite surgery resection and chemotherapy which started with an important delay.

Surgical resection remains the standard treatment for Sclerosing epithelioid fibrosarcoma (SEF), with wide surgical resection being the most effective strategy [21,24,25]. The benefit of radiotherapy remains in doubt and is performed only in patients at risk for recurrence or metastasis such as margin positivity, tumor location, and tumor size [24]. The efficiency of adjuvant therapy in the control of SEF is not yet demonstrated [23]. Chemotherapy is reserved for patients whose tumors recur locally or spread to distant sites [25,26]. Regular and long-term follow-up is recommended since recurrences or distant metastases can be seen late [8].

#### 4. Conclusion

SEF is histologically low-grade sarcoma but clinically aggressive tumor. SEF of the gluteal region can induced clinically a sciatic nerve pain. As the rates of recurrence and metastasis are high, long-term follow-up is necessary. The diagnosis must be established as early possible in order to improve the prognosis.

#### Declaration of Competing Interest

This article has no conflict of interest with any parties.

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#### Ethical approval

The study type is exempt from ethical approval.

#### Consent

The patient is dead. We have the authorization of the chef of department.

#### Author contribution

Writing the paper, Study Concept: Khezami karim.  
Data collection, Study Concept: Bennour Mohamed Amine, Ahmed Gharbi, Chabaane Mohamed, Nouri Habib.  
Supervision: Mohamed Amine Bennour, Nouri Habib.

#### Registration of research studies

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#### Guarantor

Dr Khezami Karim.

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