

## Epithelioid Sarcoma of the Big Toe: A Case Report

Ghorbel I<sup>1</sup>, M'Chirgui M<sup>1,2\*</sup>, Abid A<sup>1</sup>, Karra A<sup>1</sup>, Ennouri K<sup>1</sup>

<sup>1</sup>Orthopedic Surgery and Trauma Department, Hospital Maamouri, Nabeul 8000, Tunisia

<sup>2</sup>Department of Plastic Surgery, Faculty of Medicine Sousse, Sousse, Tunisia

**Rec:** October 18, 2017; **Acc:** January 10, 2018; **Pub:** January 17, 2018

\***Corresponding author:** Mayssa M'Chirgui, Department of Plastic Surgery, Faculty of Medicine Sousse, Sousse, Tunisia, Tel: +216 50 251 437; E-mail: [elmchirgui.mayssa@hotmail.fr](mailto:elmchirgui.mayssa@hotmail.fr)

### Abstract

**Background:** Epithelioid sarcoma is a rare form of soft tissue sarcoma. It appears as a subcutaneous mass at the extremities of adolescents and young adults. The notion of trauma is often reported. The location at the foot is exceptional. Through a case of epithelioid sarcoma of the big toe, we will detail the clinical and histological characteristics of this tumor, its main differential diagnosis and the therapeutic procedures.

**Case presentation:** We report a case of an epithelioid sarcoma of distal location in an 18-year-old man, who consults initially for a chronic fistula in the right big toe. He has undergone surgical excision. One year later he consulted for a revival of the inflammatory and infectious phenomena evolving towards fistulization despite a well conducted treatment. Surgery, fustilectomy and bacteriological samples for specific germs and histopathological examination were performed. The results of the pathologic examination and the immunohistochemical (IHC) were in favor of a distal-type epithelial sarcoma (grade 1 according to FNCLCC). The decision was to perform an extra-compartmental surgery associated with a lymph node biopsy.

**Conclusion:** Epithelioid sarcoma is a rare tumor, its location at the foot is unusual, and it must be considered in case of nodular or ulcerated lesion that does not yield under medical treatment in a young adult male.

**Keywords:** Epithelioid sarcoma; Soft tissue sarcoma; Case study; Malignant tumor; Surgical treatment

### Background

Epithelioid sarcoma is rare and occurs in only 1% to 2% of soft tissue sarcomas. It was described for the first time by Enzinger in 1970 [1]. It is a lymphophilic tumor with a high metastatic potential that mainly affects the adolescent and young adult male.

It manifests itself clinically in the form of superficial nodules of the extremities (hand and forearm). However, the diagnosis is often confused with annular ulcers or granulomas, or even palmar fibromatosis [2-5]. CT-scan or Magnetic Resonance Imaging (MRI) centered on the suspect area may be useful in the local extension report [6]. Immunohistochemical (IHC) examination confirms the diagnosis.

Surgical Treatment is conservative at an early stage of the disease mainly due to the harmless clinical appearance and small size of the tumor. At an advanced stage the treatment becomes more radical and consists mainly of a large local excision or an amputation [7].

Patients should be under control at regular intervals for early detection of recurrence or metastasis. Therefore, the follow-up intervals are chosen according to the histological grade, the size of the initial lesion and the site of the tumor [6].

The prognosis is also pejorative as the seat is proximal or axial, deep, sizes >5 cm. Its mitotic activity is important as there are foci of necrosis and vascular emboli [8].

Through a case of epithelioid sarcoma of the big toe, which is a rare localization, we develop the clinical and histological features of this rare pathological entity, its main differential diagnosis and its therapeutic procedure.

### Case Presentation

We present the case of an 18-year-old man, who consults for a chronic fistula in the right big toe. The history of his illness dates back to 2012 when the patient was victim of a TA by the inclusion of an iron burr at his right big toe, evolving towards an infected wound. A surgical excision of the necrotic and infected tissues and an ablation of the foreign body were performed. The immediate sequences were simple with healing of the lesions and resumption of current activities.

However one year later it was noticed a revival of the inflammatory and infectious phenomena evolving towards the fistulisation to the skin in spite of a well conducted local treatment and an antibiotic treatment by general route. The standard radiological assessment did not show any anomalies (Figure 1).

The indication of a surgical resumption was made and a fustilectomy and bacteriological samples in search of specific germ and histopathological examination were carried out.

The results of the pathologic examination and the IHC

**Citation:** Ghorbel I, M'Chirgui M, Abid A, Karra A, Ennouri K. Epithelioid Sarcoma of the Big Toe: A Case Report. Case Rep J. 2018;2(1):006



**Figure 1:** Radiographic aspect of the right foot.



**Figure 2:** Preoperative aspect of lesion at the right hallux.

were in favor of a distal-type epithelial sarcoma (grade 1 according to FNCLCC).

The patient was summoned to complete the assessment of the extension by a thoraco-abdomino-pelvic CT scan which showed the presence of centimetric adenopathy at the inguinal ganglionic areas of inflammatory appearance. The rest of the exam is without anomalies. The decision was to perform an extra-compartmental surgery associated with a lymph node biopsy, which was performed with simple surgical procedures (Figures 2-4).

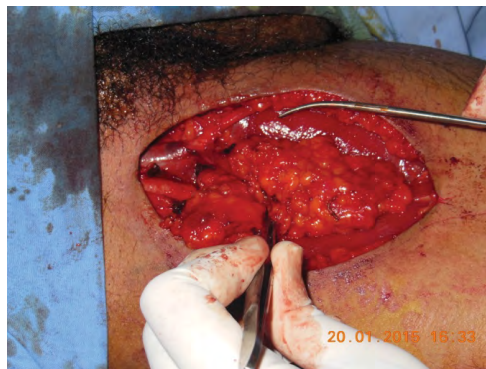
The anatomopathological examination of the surgical specimen showed the limits of healthy resection and absence of lymph node metastasis.



**Figure 3:** Per operative drawing.



**Figure 4:** Piece of surgical resection.



**Figure 5:** Per operative aspect of lymph node dissection.

At the 3-month clinical examination, the result shows the presence of adenopathies in the inguinal region. The computed tomography appearance revealed the same appearance at the beginning.

In view of the lymphophilic character of the tumor and the persistence of the ADP, it was decided to complete the inguinal clearing which proved negative (Figure 5 and 6). At the 2-year follow-up, there was no local recurrence (Figure 7).



**Figure 6:** Aspect after femoral lymph node dissection.



**Figure 7:** Clinical appearance of amputation stump at last clinical control.

## Discussion

Epithelioid sarcoma was described for the first time by Laskowski in 1961 [9]; Enzinger was the first to attempt to establish that it is a separate entity [1], it accounts for only 1% to 2% of all soft tissue sarcomas during the first two decades. It presents clinically in the form of an indurated, painless, ulcerated, superficial cutaneous or subcutaneous nodule that is more rarely adherent deep tissue or tendons and aponeuroses. This tumor affects essentially the upper limbs (60% to 70%). It is also seen in the legs, buttocks, thigh and knee, and more rarely in the feet [2-5].

It is often confused with benign pathologies such as granuloma (rheumatoid, annular, infectious), fibromatosis (duputren, peyronie), a histiocytoma, or malignant

such as other carcinomas, including ulcerative malpighien, epithelioid angiosarcoma and melanoma, hence an immunohistochemical examination is required before any inflammatory lesion that does not yield under medical treatment [5].

MRI is the preferred radiological examination before biopsy, although it does not show specific signs of epithelioid sarcoma, it allows determining the limits of the tumor and makes it possible to differentiate tumor recurrence from postoperative changes [10].

Under the microscope, this tumor is characterized by a granuloma-like pattern aspect: clusters of fusiform and epithelioid cells circumscribe areas of central hyalinization and necrosis.

Fibrous histiocytoma-like and angiomatoid subtypes have also been reported as less common histologic variants [11,12]. Immunohistochemically, these tumors demonstrate nonspecific cytoplasmic EMA-reactivity [13]. There is consistent staining for CA-125, and some have suggested using it as a serum marker to monitor for metastasis. S100 is typically negative (distinguishing it from malignant peripheral nerve sheath tumor), as are endothelial markers (distinguishing it from epithelioid angiosarcoma), and CK5/6 (distinguishing it from squamous cell carcinoma). CD34 is expressed in 50% to 60% of epithelioid sarcomas, but is negative in carcinomas, helping to distinguish the two. P63 is also a useful marker that is present in virtually all squamous cell carcinomas, but absent in epithelioid sarcomas [5].

Proximal or axial seat, deep, size >5 cm, mitotic activity, necrosis and the presence of vascular emboli are associated with a pejorative prognosis [8].

The surgical treatment is consisting of a wide resection or an amputation if there are several recurrences or if there is no significant loss of function. However, this does not seem to aid in the control of local metastases [7,14], this risk is of the order of 34% to 77% at 5 years, 80% at 10 years; these metastases are often multiple along the fascia, regional tendon or nerve and ganglionic sheaths.

They are mostly early but can be tardy and seen until 25 years after the diagnosis of the primary tumor [8].

Remote metastases are also frequent (45% to 8% of cases) and mostly affect lung, ganglia, CNS, skin and soft tissue. These metastases occur mostly within 2 years.

With regard to the risk of lymphatic metastases, some have proposed a sentinel node biopsy and regional lymph node dissection, but documentation of the results is insufficient and further research is needed [15].

Overall survival is 50% to 70% at 10 years [8]. This risk can be lowered by early diagnosis and wide excision at an early stage of the disease [16].

## Conclusion

Epithelioid sarcoma is a rare tumor that requires complex, cumbersome, multidisciplinary management that can only be assured within a reference center.

## References

1. Fisher C. Epithelioid sarcoma of Enzinger. *Adv Anat Pathol*. 2006;13(3):114-21.
2. Fletcher CD, Unni KK, Mertens F; WHO, IARC. Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press; 2002.
3. Weiss SW, Goldblum JR, Enzinger FM. Enzinger and Weiss's soft tissue tumors. 4th ed. St Louis: Mosby; 2001.
4. Bryan RS, Soule EH, Dobyns JH, Pritchard DJ, Linscheid RL. Primary epithelioid sarcoma of the hand and forearm. A review of thirteen cases. *J Bone Joint Surg Am*. 1974;56(3):458-65.
5. Miettinen M, Fanburg-Smith JC, Virolainen M, Shmookler BM, Fetsch JF. Epithelioid sarcoma: an immunohistochemical analysis of 112 classical and variant cases and a discussion of the differential diagnosis. *Hum Pathol*. 1999;30(8):934-42.
6. Delloye C, Schubert T. [The management of soft tissue sarcomas in orthopedic surgery]. *Newsletter* 27. King Albert II Institute; 2015.
7. Enzinger FM. Epithelioid sarcoma. A sarcoma simulating a granuloma or a carcinoma. *Cancer*. 1970;26(5):1029-41.
8. de Visscher SA, van Ginkel RJ, Wobbles T, Veth RP, Ten Heuvel SE, Suurmeijer AJ, et al. Epithelioid sarcoma: Still an only surgically curable disease. *Cancer*. 2006;107(3):606-12.
9. Laskowski J. Sarcoma aponeuroticum. *Nowotwory*. 1961;11:61-7.
10. Herr MJ, Harmsen WS, Amadio PC, Scully SP. Epithelioid sarcoma of the hand. *Clin Orthop Relat Res*. 2005;431:193-200.
11. Hasegawa T, Matsuno Y, Shimoda T, Umeda T, Yokoyama R, Hirohashi S. Proximal-type epithelioid sarcoma: a clinicopathologic study of 20 cases. *Mod Pathol*. 2001;14(7):655-63.
12. von Hochstetter AR, Meyer VE, Grant JW, Honegger HP, Schreiber A. Epithelioid sarcoma mimicking angiosarcoma: the value of immunohistochemistry in the differential diagnosis. *Virchows Arch A Pathol Anat Histopathol*. 1991;418(3):271-8.
13. Mirra JM, Kessler S, Bhuta S, Eckardt J. The fibroma-like variant of epithelioid sarcoma: a fibrohistiocytic/myoid cell lesion often confused with benign and malignant spindle cell tumors. *Cancer*. 1992;69(6):1382-95.
14. Swanson PE, Scheithauer BW, Manivel JC, Wick MR. Epithelial membrane antigen reactivity in mesenchymal neoplasms: an immunohistochemical study of 306 soft tissue sarcomas. *Surg Pathol*. 1989;2:313-22.
15. Chase DR, Enzinger FM. Epithelioid sarcoma: Diagnosis, prognostic indicators and treatment. *Am J Surg Pathol*. 1985;9(4):241-63.
16. Blazer DG, Sabel MS, Sondak VK. Is there a role for sentinel lymph node biopsy in the management of sarcoma? *Surg Oncol*. 2003;12(3):201-6.