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(CASE REPORT)

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Proximal-type epithelioid sarcoma: A new case report

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Abstract

Epithelioid sarcoma (ES) is a rare high-grade soft tissue sarcoma, first described by Enzinger in 1970, classically presenting as a subcutaneous or deep dermal mass in the distal extremities of adolescents and young adults. In 1997, Guillou et al. described the proximal-type epithelioid sarcoma, which is found mostly in the pelvic, perineal regions, and genital tracts of young to middle-aged adults. Epithelioid sarcoma is characterized by a proliferation of epithelioid-like cells with rhabdoid features and the absence of a granuloma-like pattern. We present, in this paper, a case of proximal-type epithelioid sarcoma of abdominal wall, with an aggressive clinical course, including distant metastasis and death one month after diagnosis. In this particular case, the location is infrequent, and it had not previously been described.

Keywords: Epithelioid Sarcoma; Proximal-type; Oncology; Case report

1. Introduction

Proximal-type epithelioid sarcoma (PES) is a rare high-grade soft tissue sarcoma [1], (less than 1% of all soft tissue sarcomas), classically presenting as a subcutaneous or deep dermal mass, with slow-growing and a high potential for distant metastasis in adolescents and young adults [2]. These tumors arise in proximal locations (axilla, genital area, trunk ...). Late diagnosis and treatment are associated with a poorer prognosis[3]. Owing to its rarity, aggressive behavior, the complexity of its clinical and histopathological diagnosis, and its high recurrence potential, we report a new case of a 60 years old man presented with proximal-type epithelioid sarcoma of his anterior abdominal wall.

1.1. Patient and observation

A 60 years old man presented with a three-months history of three slowly growing masses in his anterior abdominal wall. He had been smoking for 26 years and reported no other significant personal or family medical history.

The largest abdominal mass appeared to be budding, ulcerated, and bleeding on contact, measuring 10 cm in larger diameter (Figure 1).

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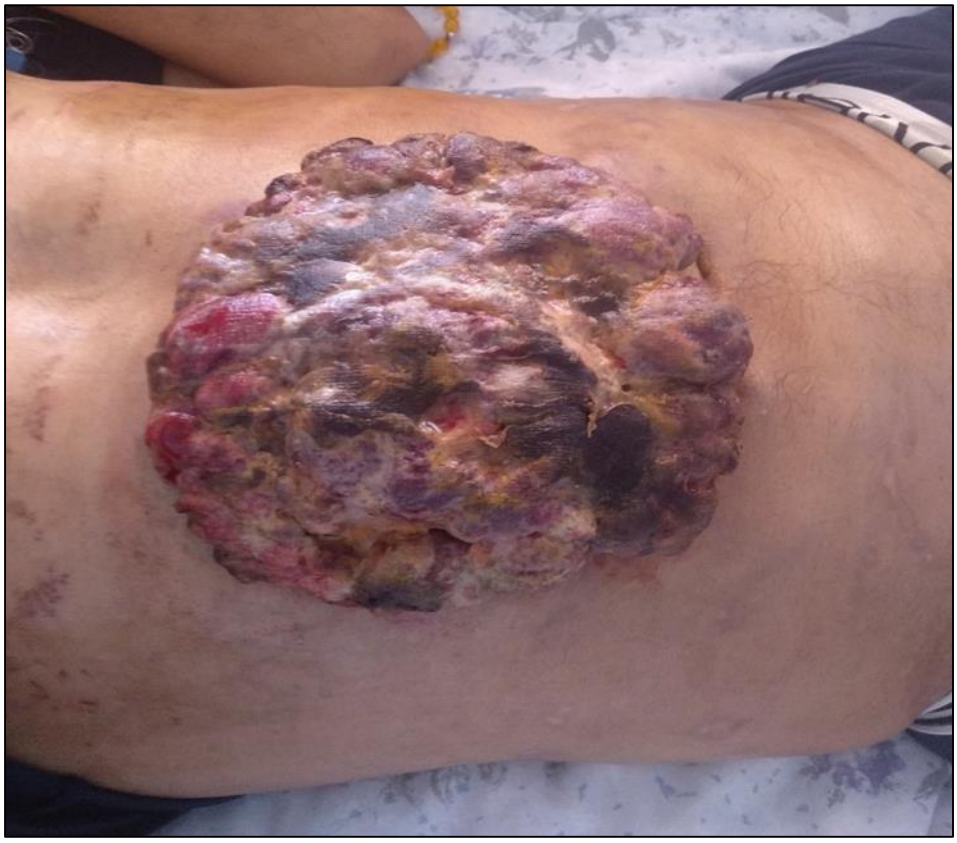

Figure 1 Epithelioid sarcoma of the anterior abdominal wall

Budding, ulcerated, and bleeding tumor of the anterior abdominal wall measuring 10cm*6cm

A first biopsy was performed and was in favor of a benign lesion (pseudoangiomatous histiocytofibroma), the patient then underwent a simple excision, and the specimen was sent for pathologic evaluation, the result was in favor of epithelioid sarcoma. A thoraco-abdomino-pelvic CT scan was performed and showed the presence of two subcutaneous masses measuring 90*55 mm and 25*12 mm on the anterior abdominal wall with intraperitoneal tissue nodule located at the left flank, measuring 75 * 55 mm, multiple nodules of the anterior and right lateral abdominal wall and subcutaneous tissue of variable sizes and left upper lung lobe nodule, measuring 35 * 32 * 28 mm evoking a peritoneal parietal and lung metastasis.

The tumor was considered to be inoperable, so the decision was to start chemotherapy with ifosfamide plus doxorubicin, but the patient died two weeks later.

2. Discussion

Epithelioid sarcoma (ES) was initially described in 1970 by Enzinger[1]. It is a rare malignant mesenchymatous tumor more frequently found in young patients from 23 to 40 years old[4]. As a result of its diverse clinical scenario, diagnosis is generally late. ES is usually divided in proximal and distal presentation, with predominant topography on distal zones such as upper extremities, mainly fingers, hands, and wrists[5]. The proximal-type or the axial-type variant of this undifferentiated soft tissue sarcoma was only recently described[6]. The proximal-type variant is found mostly in the pelvis, perineum, thigh, and genital tract in adults[7]. This variant is more often deep-seated, has distinct histological features, metastasizes earlier, and behaves more aggressively than the usual distal ES[6,7].

The microscopic appearance of ES ranges from spindle cells to large polygonal cells with an acidophilic cytoplasm[8]. Diagnosis can be confirmed with immunochemical staining positive for epithelial markers such as epithelial membrane antigen and cytokeratin, CD34, and a mesenchymatous marker (vimentin). Finally, a cytogenetic analysis was performed in some small series, identifying genetic variations in the long arm of chromosome 22[9].

ES is characterized by its high recurrence rate, with up to 77% of local recurrences reported, and a high percentage of node and lung metastasis (36%-44%)[10]. Five years and ten-year survival are 65.3% and from 25% to 50% respectively [11]. However, the meantime, from recurrence to death in patients older than 36 years, stands at 5.6 ± 4.5 months and in younger patients at 15.2 ± 17.2 months [5]. Furthermore, the specific treatment for this pathology has not been established by international consensus; where a distal type ES tends to avoid amputation, a local recurrence is treated with local excision plus radiotherapy [9]. Nevertheless, in tumors with unfavorable factors such as proximal type or size greater than 5 cm, a systemic treatment plus surgical intervention should be evaluated [12].

ES is a rare variant of malignant sarcoma, with very aggressive behavior, which will only benefit from a prompt diagnosis and an intensive, multidisciplinary treatment. In this particular case, the location is infrequent, and it had not previously been described. Thus, the importance of reporting this case to increase awareness of this rare disease, since it seems only an early diagnosis with definite surgical treatment that can improve prognosis.

3. Conclusion

Epithelioid sarcoma is identified as one of the rarest types of sarcomas. This tumor has an aggressive clinical course with high recurrence and metastasis rates, especially the proximal type. This case suggests that ES can be derived from the soft tissue of the abdominal wall, and future research is still needed for this rare malignancy and clinical presentation.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare no competing interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors'.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

Authors' contributions

All the authors contribute in the diagnosis and treatment of the patient. All the authors contribute in the write-up of the manuscript and approved the final version.

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