

Case Reports & Case Series

Epithelioid sarcoma involving sciatic nerve with lung metastasis and local recurrence: A case report



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ABSTRACT

Involvement of a deep peripheral nerve by epithelioid sarcoma (ES) is extremely rare, especially in sciatic nerve. It is a highly aggressive malignant tumor with poor prognosis and comparatively difficult to detect and confirm diagnosis in early stage. We report a rare case of epithelioid sarcoma involved the sciatic nerve which presented with chronic sciatic pain and treated with radically wide resection, but lung metastasis and local recurrence have occurred shortly after surgical resection and resulting in the patient's death in six months after initial presentation.

1. Introduction

Epithelioid sarcoma (ES) is a rare soft tissue sarcoma, first characterized as a distinct clinicopathologic entity by Enzinger in 1970, which presents in the two main clinicopathologic settings [1]. Deep-seated ES may spread along the neurovascular bundles with potential vascular and lymphatic invasion and metastasis. As deeper tumors increase in size, they may compress adjacent nerves and produce pain or neurologic symptoms which may be misdiagnosed in the early stage. To the best of our knowledge, there are only two cases that have been reported of an epithelioid sarcoma involving sciatic nerve [2,3].

2. Case report

A 31-year-old male was referred to our department with diagnosis of "Right sciatic nerve mass" from local hospital. He had a 2-year history of lower back pain which aggravated over the past 2 month and radiated to posterior and lateral area of the right leg and foot. He was diagnosed of "Lumber disc herniation" and received conservative treatment accordingly without any significant improvement. A CT scan was performed and revealed approximately 3.5×4.2 cm solitary mass involving the proximal portion of sciatic nerve. [Fig. 1A] Open biopsy was done [Fig. 1B] and pathological study revealed "epithelioid sarcoma with sciatic nerve involvement" at local hospital.

Neurological exam revealed normal sensation of the right leg but patient was reluctant to cooperate with movement exam and electromyogram was not conducted due to previous history of operation. Chest

X-ray showed not involvement of the lung. Briefly, operation was performed under general anesthesia, complete and extended excision of the tumor include skin and subcutaneous soft tissues with 10 cm in length and 5 cm in width around tumor location was made meticulously by protecting the adjacent nerves or vessels under microscope with $\times 4$ loupe magnification [Fig. 1C]. Sciatic nerve gap was reconstructed by bilateral sural nerve grafting to reconstruction of common peroneal nerve [Fig. 1D]. The postoperative pathological diagnosis of tumor was confirmed on the basis of light microscopy pathological examination and immunohistochemical staining [Fig. 2].

Radiotherapy was started after complete wound healing in twenty days after operation. Patient suffered intermittent hemoptysis in two month after operation and pulmonary CT scan revealed "occupied mass" [Fig. 3A]. Biopsy was done pathological study indicated epithelioid sarcoma (metastasis from primary lesion) [Fig. 3B]. Patient was treated in respiratory department for long period but respiratory failure have occurred and aggravated with local recurrence of tumor in four month after initial operation [Fig. 3C].

3. Discussion

Epithelioid sarcoma is defined as malignant mesenchymal neoplasm that exhibits a predominantly epithelial phenotype and it is rarely arises primarily from peripheral nerves. When it does occur, it may be difficult to differentiate from benign peripheral nerve sheath tumors [4]. To the best of our knowledge, there are only two cases have been report of an epithelioid sarcoma arising from the epineurial sheath of the sciatic

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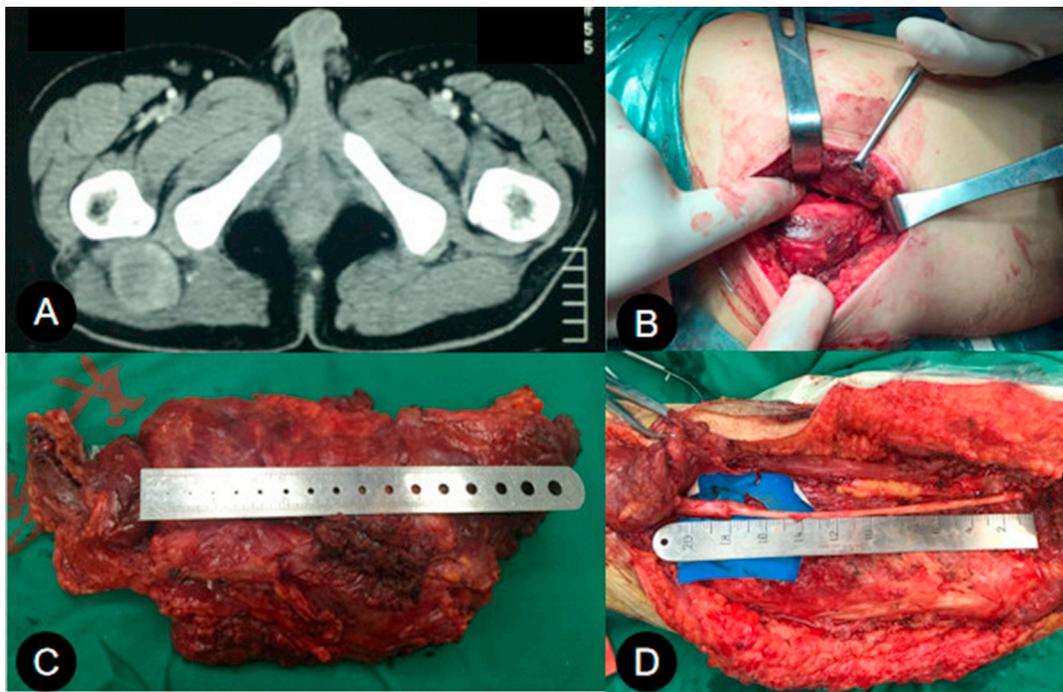


Fig. 1. A CT scan showed solitary mass involving right sciatic nerve. B Intraoperative finding demonstrated a solitary mass involvement of sciatic nerve. C Wide radical resection of the tumor and surrounding tissues and sample was sent for pathological study. D Bilateral sural nerve grafted to bridge approximately 18 cm gap.

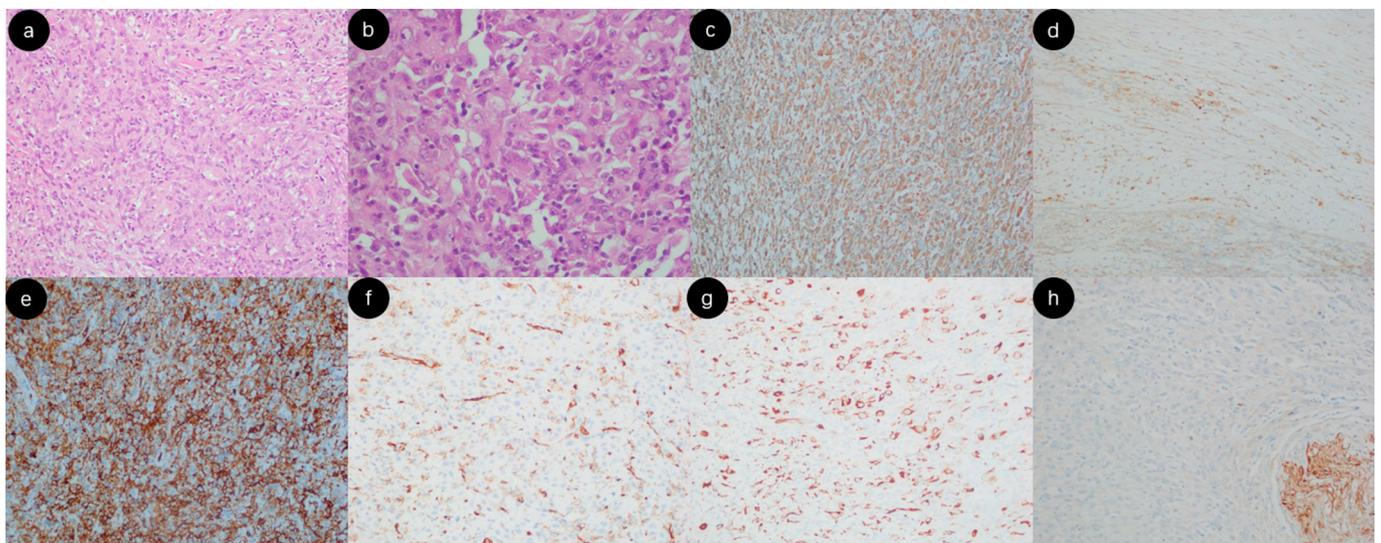


Fig. 2. Histopathological findings and Hematoxylin and eosin stain (H&E) results of the tumor (a) $\times 200$ magnification (b) $\times 400$ magnification; (c) Positive staining for vimentin ($\times 200$ magnification); (d) Positive staining for epithelial membrane antigen ($\times 200$ magnification); (e) Positive staining for CD34 ($\times 200$ magnification); (f) Positive staining for CD31 ($\times 200$ magnification); (g) Positive staining for CK-14 ($\times 200$ magnification); (h) Negative staining for S-100 ($\times 200$ magnification).

nerve [2,3]. ES is a highly aggressive tumor with high local recurrence rate as well as local or distal metastasis. In the largest case series to date, Chase and Enzinger reported a crude local recurrence rate of 77% despite the fact that nearly half of the patients underwent limb amputation [5].

Epithelioid sarcoma involving sciatic nerve usually present with symptoms mimicking lumbar disc herniation or sciatica which may prolong its clinical course and late consideration of tumor existence

on sciatic nerve. Our presented case is typical example of late diagnosis with poor prognosis, patient suffered long history of lower back pain and treated as “lumbar disc herniation” based on MRI result of “lumbar vertebra” which didn’t reveal the “true” lesion in sciatic nerve region. It is recommended to consider peripheral nerve diagnoses in patients with pain complaints not readily explained by more common spinal etiologies considering its rarity and poor survival rates.

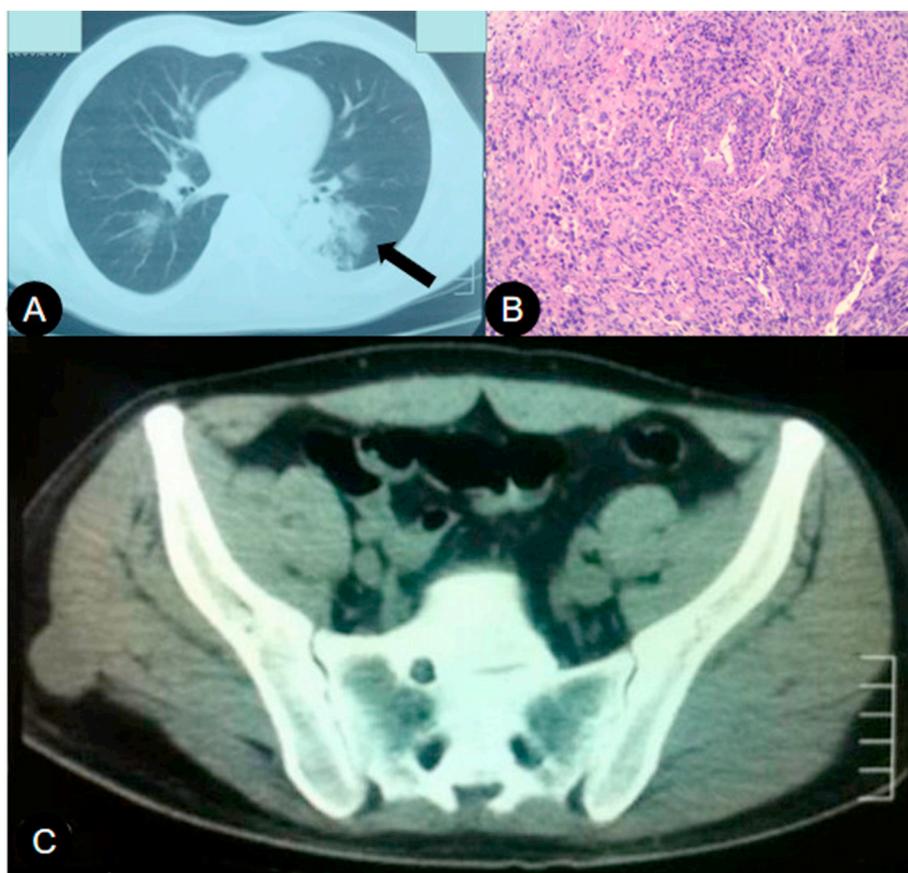


Fig. 3. A Pulmonary CT scan showed occupied mass two month after initial operation. B Histopathological is consisted with originated malignant epithelioid features. C CT scan demonstrated mass recurrence in four month after initial operation.

Declaration of Competing Interest

The authors declare that they have no conflicts of interest.

Acknowledgements

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