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CASE REPORT

Epithelioid Sarcoma of the Sciatic Nerve Perineural Sheath: a Mimic of Nerve Sheath Tumor

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We report herein a rare case of epithelioid sarcoma, in a 39-year old lady involving the sciatic nerve. Clinically and radiologically it stimulated a nerve sheath tumor. Involvement of a nerve by an epithelioid sarcoma is extremely uncommon. To the best of our knowledge, this is the first case of an epithelioid sarcoma involving the sciatic nerve and needs documentation. (Pathology Oncology Research Vol 8, No 2, 148–150, 2002)

Keywords: epithelioid sarcoma, sciatic nerve, soft tissue tumor, sarcoma

Introduction

Epithelioid sarcoma (ES) was first described as a distinct entity by Enzinger in 1970.[1] However, the first case possibly of this tumor was described by Laskowski[2] who regarded its origin as aponeurotic and skin structures. The tumor is more prevalent in adolescents and young adults between 10 to 35 years and frequently involves the hand, fingers, wrist, forearm, buttock, thigh, ankle and lower legs. Rarely the trunk and head and neck regions are involved.[3] Most of these tumors present as a superficial firm nodule or ulcer and rarely as deep seated lesions. Involvement of deep nerves is extremely uncommon. We report a rare case of this tumor which was involving the sciatic nerve.

Case Report

A 39-year-old lady presented with complaints of pain at the back of thigh and progressively increasing numbness of the left sole of foot for the last two and a half years. This pain was not related to coughing, sneering or movements of spine. Power of the left ankle and knee ranged from 3/5 to 5/5. MRI scan of the thigh showed a localised globular enlargement of the sciatic nerve at the inferior margin of the gluteus maximus muscle just before the nerve entered the thigh. This enlargement measured 3.2x1.7x2 cm. It was hyperintense on T2W images and showed intense enhancement on contrast injection (*Figure 1*). With a clinical diagnosis of schwannoma, left sciatic nerve exploration and gross total excision of the tumor was done. This was subjected for histopathological examination.

Pathological Examination

Specimen measured 3x2x2 cm in dimension. Tissue was routinely fixed in 10% buffered neutral fomalin and was paraffin embedded. Five micron thick sections were cut and stained with Hematoxylin and Eosin (H&E) stain. Immunohistochemical staining was done by using antibodies against cytokeratin (CK dil 1:50), epithelial membrane antigen (EMA dil 1:50), S-100 protein (dil 1:100), HMB-45 (dil 1:50), neuron specific enolase (dil 1:100), panactin (dil 1:50), desmin (dil 1:50) and factor VIII (dil 1:50). All antibodies were obtained from M/s Dako Patt, Denmark.

Immunohistochemical staining

Microscopic examination of the sections showed a nodular arrangement of the tumor cells with areas of necrosis in the centre of these nodules (zonal necro-

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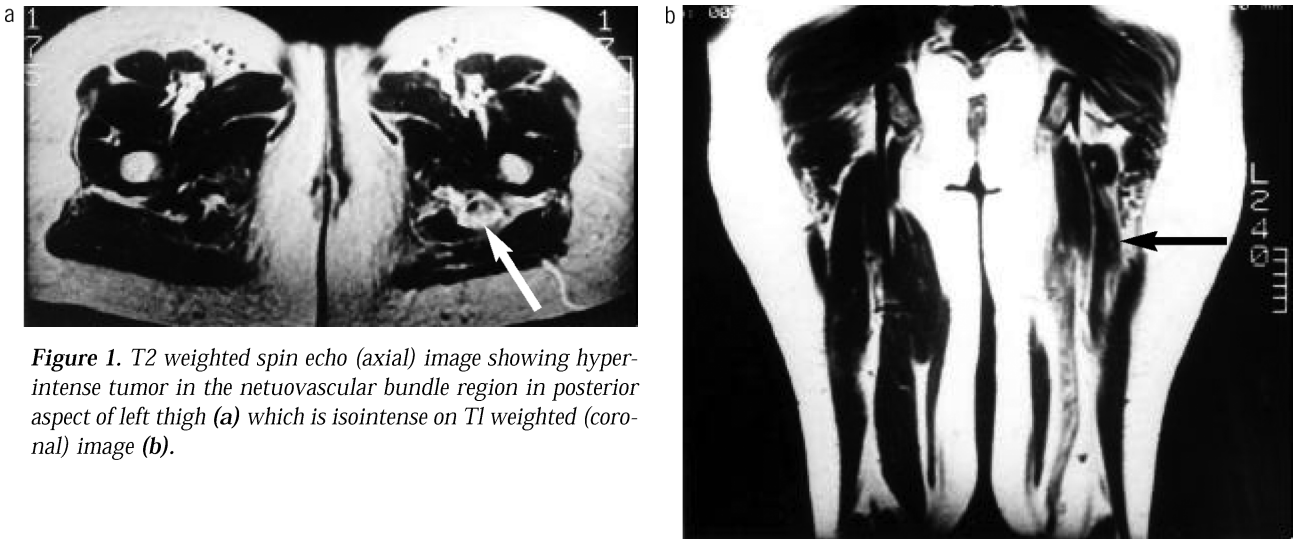

Figure 1. *T2 weighted spin echo (axial) image showing hyperintense tumor in the netuovascular bundle region in posterior aspect of left thigh* ***(a)*** *which is isointense on Tl weighted (coronal) image* ***(b).***

sis). Nodules showed irregular, undulating borders. Cells had an epitheloid appearance and were large, round, polygonal with abundant dense eosinophilic cytoplasm (*Figure 2*). Nuclei were vesicular with small nucleoli. Nerve bundles were seen to be invested by the tumor cells.

Immunohistochemical staining showed positivity for cytokeratin (CK), epithelial membrane antigen (EMA), vimentin (*Figure 3*) and focal positivity for S-100 protein but was negative for HMB-45, glial fibrillary acidic protein (GFAP), neuron specific enolase (NSE), smooth muscle actin (SMA), panactin, desmin and factor VIII.

Based on histopathological features and positivity for CK, EMA and vimentin a diagnosis of epithelioid sarcoma was made. The possibility of a metastatic carcinoma was not considered in view of a long clinical history of 3 years and absence of primary tumor at any other sites.

Follow up

Post-operatively the patient was given local radiotherapy in the dose 6000 rads over a period of 6 weeks. Follow up at one year revealed no recurrence or metastasis.

Discussion

Epithelioid sarcoma is a tumor of adult age and has predilection for the distal portions of the extremties. It pursues a slow but relentless clinical course.[4-7] This tumor occurs both in superficial and deep locations. The superficial lesions are situated in the subcutis and present as firm nodules which are raised above the surface and frequently become ulcerated. The deep seated lesions are firmly attached to tendon, tendon sheaths or fascial structures. They are larger, less well defined and vary in size from 5 mm to 15 cm or more. Rarely they

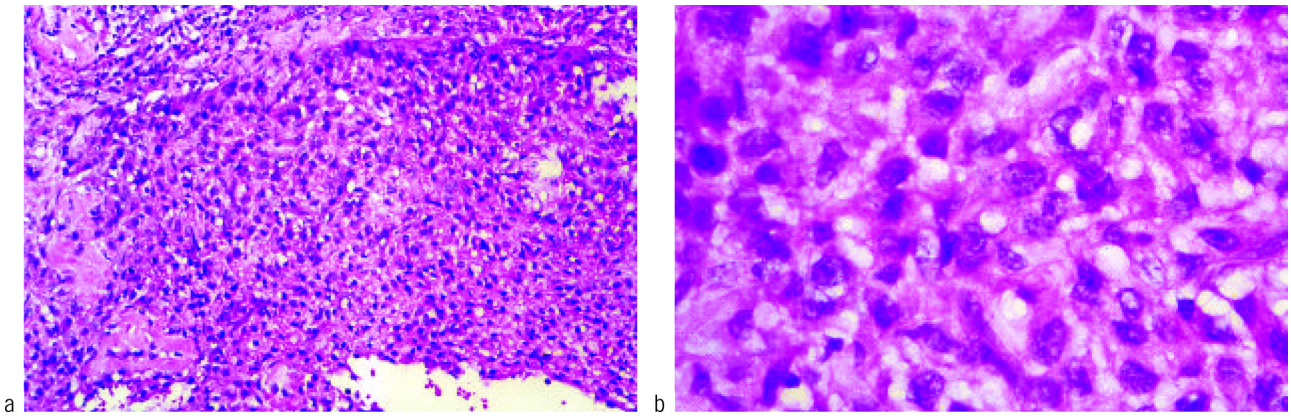

Figure 2. *Photomicrographs showing a nodular aggregate of tumor cells consisting of sheets of polygonal cells (H&E x 40). Higher magnification demonstrating moderate amount of eosinophilic cytoplasm with indistinct cellular outlines (H&E x 200).*

grow along the neurovascular bundles and cause pain or tenderness.

Pressure on the nerves or growth along the neurovascular bundles is not uncommon but presentation of the tumor primarily as nerve sheath tumor is extremely rare.

Prat J et a1[4] reported a series of 22 cases of epithelioid sarcomas, of which 3 patients presented with pain, swelling and difficulty in moving the fingers. In these 3 cases, besides the involvement of tendons of flexor digitorum profundus, superficialis and interosseous muscles, tumors were encircling the median nerve also. In the same study one case which was located in the axilla was invading the brachial plexus. Bryan et al[8] reported a series of 13 cases of epithelioid sarcoma, of which 2 were deep seated but none showed neural involvement. In another series of 51 cases of epithelioid sarcomas, 26 were deep seated close to deep fascia or tendon sheath or both but none of them showed neural involvement[6]. In the present case under discussion, the tumor was encircling the sciatic nerve and was mimicking a neural tumor. The tumor was probably arising from the perineural sheath.

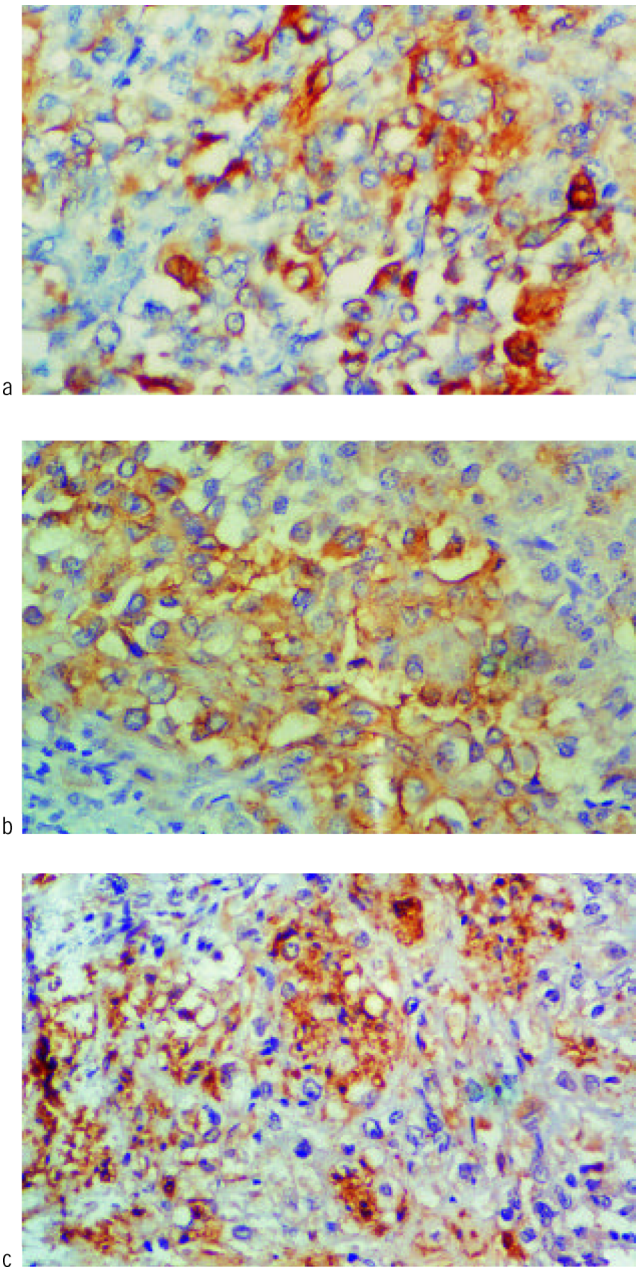

Figure 3. *Photomicrographs showing positivity for cytokeratin* ***(a)****, epithelial membrane antigen* ***(b)*** *and vimentin* ***(c)*** *(X200 each).*

Prognosis of this tumor is favourable with 5 and 10 year survival of 70% and 50% respectively[6]. In a large series of 202 cases from AFIP,[5] 70% experienced recurrences. Metastasis does occur to regional lymph nodes, lungs and less frequently to central nervous system, soft tissue and scalp.

Treatment of choice is surgical resection, however, soft tissue sarcoma involving the sciatic nerve poses a challenge. Recommended guidelines for any soft tissue sarcoma involving sciatic nerve are that, if tumor is less than 10 cms and involvement of sciatic nerve is minimal, then every endeavour should be made to preserve the nerve.[9] Tumor can be dissected from the nerve sheath, more so if the tumor is not of neural origin. If margins are positive for microscopic tumors, post-operative radiotherapy or brachy therapy is recommended. Sciatic nerve should be sacrificed only if the tumor is very large and it is far superior than limb amputation. Chemotherapy should be reserved for recurrent or metastatic disease. Although rare, the possibility of this tumor should be considered in the differential diagnosis of the sarcomas involving the sciatic nerve.

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