CASE REPORT

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Epithelioid sarcoma of femoral nerve – A case report

Epiteloidni sarkom femoralnog nerva

Dejan Ivanov*†, Mirjana Živojinov†‡, Milan Ranisavljević†§

Clinical Center of Vojvodina, *Clinic for Abdominal, Endocrine Surgery and Transplantation, †Department for Pathology, Novi Sad, Serbia; ‡Oncology Institute of Vojvodina, Sremska Kamenica, Serbia; University of Novi Sad, §Faculty of Medicine, Novi Sad, Serbia

Abstract

Introduction. Epithelioid sarcoma is a slow-growing malignant soft tissue tumor and occurs approximately in 1% of all soft tissue malignant tumors. This case report describes epithelioid sarcoma in femoral nerve and as we know it is the first described case of epithelioid sarcoma at this anatomical localisation. **Case report.** A 44-years-old female patient presented with strong pain in her left leg. On magnetic resonance imaging (MRI), tumor was presented as a node 8 cm in the diameter at left femoral nerve between ileopsoas and iliacus muscle, without infiltration of muscle fascia and tendons. Four enlarged lymph nodes described in left iliac fosa, were suspected on secondary tumor deposits. We preformed radical surgical excision of the tumor and femoral nerve transection with local lymp hnode disection. Histopathological analysis revealed epithelioid sarcoma, and tumor stained highly positive for anti-pan cytokeratin antibody (AE1/AE3), vimentin, cancer antigen 125 (CA125), anti-cytokeratin antibody (MNF116), hematopoietic progenitor cell antigen (CD34) and epithelial membrane antigen (EMA) markers. After the operation patient recived radiotherapy without chemotherapy. Six months postoperatively, there was no evidence of local relapses or distant metastases. **Conclusion.** Initial wide surgical resection and adjuvant radiotherapy is beneficial in treatment of epitheloid sarcoma.

Key words:
sarcoma; soft tissue neoplasms; femoral nerve; diagnostic techniques and procedures; surgical procedures, operative; radiotherapy.

Apstrakt

Uvod. Epiteloidni sarkom je redak, sporo rastući maligni tumor mekog tkiva i pronalazi se u oko 1% svih mezenhimnih tumora. Prikazujemo slučaj epiteloidnog sarkoma femoralnog nerva, koji je prema našim saznanjima, prvi ovakav opisan slučaj. **Prikaz bolesnika.** Bolesnica stara 44 godine, primljena je na pregled sa jakim bolovima u levoj nozi. Na snimku magnetne rezonance tumor se prezentovao kao nodus dimenzija 8 cm na levom femoralnom nervu, između bočnog slabinskog i ilijačnog mišića, bez infiltracije mišićnih fascija i tetiva. Četiri uvećana limfna čvora, opisana u ilijačnoj jami, bila su suspektna na prisustvo sekundarnih depozita. Urađena je radikalna hirurška ekscizija tumora i transekcija nerva sa lokalnom limfadenektomijom. Patohistološka analiza potvrdila je dijagnozu epiteloidnog sarkoma, a tumor je pokazivao izrazitu imunohistohemijsku pozitivnost na anti-pan citokeratin antitelo (AE1/AE3), vimentin, karcinomski antigen 125 (CA125), anti-citokeratin antitelo (MNF116), antigen progenitorskih ćelija hematopoeze (CD34) i epitelni membranski antigen (EMA). Nakon operacije bolesnica je primila radioterapiju bez hemioterapije. Šest meseci nakon operativnog lečenja kod bolesnice nije bilo lokalnog recidiva i udaljenih metastaza. **Zaključak.** Inicijalna široka resekcija i adjuvantna radioterapija je ključna u tretmanu epiteloidnog sarkoma.

Ključne reči:
sarkomi; meka tkiva; n. femoralis; dijagnostičke tehnike i procedure; hirurgija, operativne procedure; radioterapija.

Introduction

Epithelioid sarcoma (ES) is an uncommon malignant soft tissue tumor. It is a slow-growing tumor and occurs approximately in 1% of all soft tissue malignant tumors. It is mostly found in young adult male patients.Proximal and distal type of ES was described. Proximal type is located in subcutaneous tissues, muscular fascia or tendon sheaths of the upper extremities. Distal type has the same anatomical localisation but on distal limbs. Rare cases have been reported in the pelvis, vulva, penis, and spine [1–5].

Correspondence to: Milan Ranisavljević, Clinical Center of Vojvodina, Clinic for Abdominal, Endocrine Surgery and Transplantation, Somborska 8a, 21000 Novi Sad, Serbia. E-mail: ranke82@gmail.com

This case report describes ES in femoral nerve and it is the first case of ES at this anatomical location on the basis of avaible literature data.

Case report

A 44-years-old female patient was first examinated by physiatrist, because of strong pain in her left leg. After unsuccessful physiatrist treatments, her doctor decided to perform magnetic resonance imaging (MRI) of small pelvis and lumbal spine.

On MRI tumor was presented like a node of 8 cm in diameter at the left femoral nerve between ileopsoas and iliacus muscle, without infiltration of muscle fascia and tendons. Four enlarged lymph nodes were described in left iliac fosa, with a suspicion of secondary tumor deposits. After additional diagnostic procedures (ultrasonography – US, blood analysis), she was referred to general surgeon with presumed diagnosis of left femoral nerve tumor – retroperitoneal tumor. In her medical history there were no chronic diseases. She had no personal or family history of cancer.

After standard preoperative preparation, the patient underwent operative treatment. Intraoperatively, we found a small amount of cloudy ascites intraperitoneally. After section of peritoneum in left iliac fosa we found the tumor mass. Tumor was 8 x 4 cm in diameter and it was located inside femoral nerve with partial infiltration of ileopsoas muscle fascia. Four enlarged lymph nodes clinically suspected on metastasis were found near the tumor.

After surgical preparation of the left femoral nerve and tumor deliberation from nearby structures we performed dissection of the nearby lymph nodes which we sent to histopatological fast frosen anaylsis (HP *ex tempore*). *Ex tempore* analysis revealed malignant mesenhimal tumor.

After consultation with neurosurgeon we decided to make nerve transection, proximal and distal from the tumor mass and partial resection of the left ileopsoas muscle with fascia. Surgical free margines on femoral nerve were 2 cm from the tumor (Figure 1). We did not identify any postoperative surgical complications, except complications in left femoral nerve transection (quadriceps muscle weakness and wasting, loss of knee jerk and numbness along the medial side of the thigh and anteromedial side of the calf).

Definitive HP analysis revealed epithelioid sarcoma with high mitotic feature with free tumor margins (minimally 0.3 cm). HP analysis also revealed tumor metastasis in lymph nodes (3/4) with lymphovascular invasion. The tumor stained highly positive for anti-pan cytokeratin antibody (AE1/AE3), vimentin, cancer antigen 125 (CA125), anti-cytokeratin antibody (MNF116), hematopoietic progenitor cell antigen (CD34) and epithelial membrane antigen (EMA) markers. Calretinin, transformation-related protein 63 (p63), Wilms tumor protein (WT1), cytokeratin 5/6, chromogranin, MART1, cytokeratin 20, factor VIII, CD68 were negatively stained and CEA (Figure 2) was moderately stained. We did not perform staining for integrase interactor 1 (INI1) and SWI/SNF-related matrix-associated actin-dependent regulator of chromatin subfamily B member 1(SAMARCB$_1$). Additional imaging procedures, positron emission tomography – computed tomography (PET–CT), that we performed after the surgery, did not reveal any distant metastasis.

The case was then reviewed at tumor board and after analysis of HP and imaging findings, it was decided that the patient should recived conformal radiotherapy without adjuvant chemotherapy. The patient was followed with close surveillance. Since then, the patient was seen every month. Six months postoperatively, there were no evidence of local relapses and distant metastases.

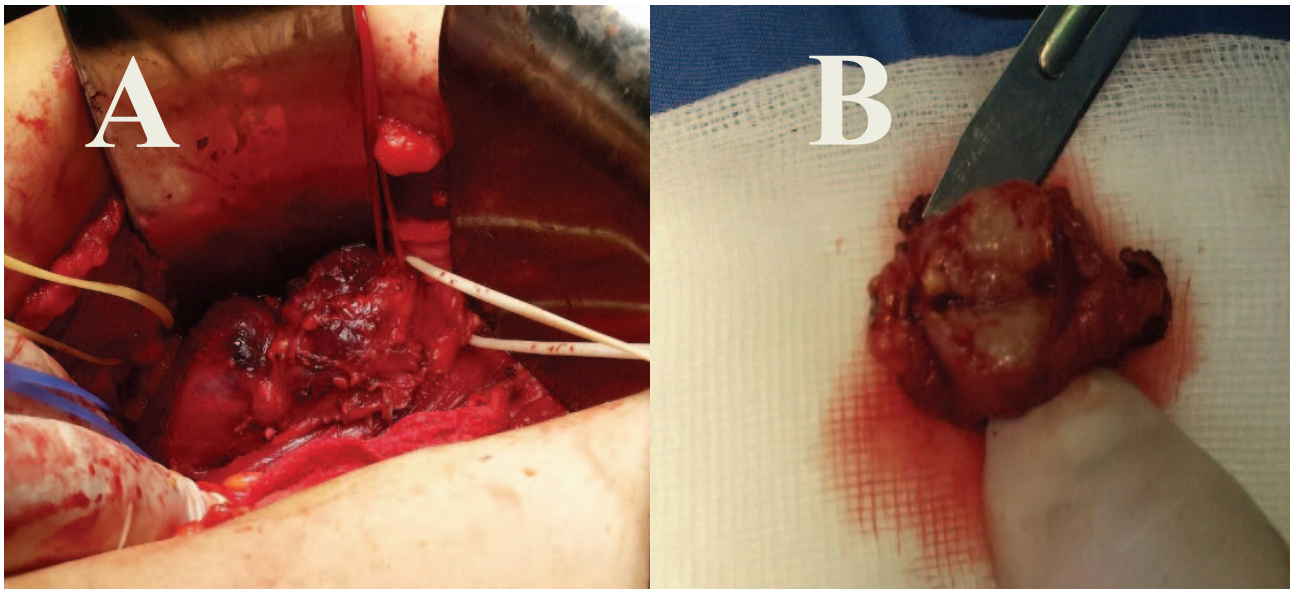

Fig. 1 – Intraoperative findings and tumor after excision.
A) White and blue string mark free margins of left femoral nerve; B) Tumor after excision.

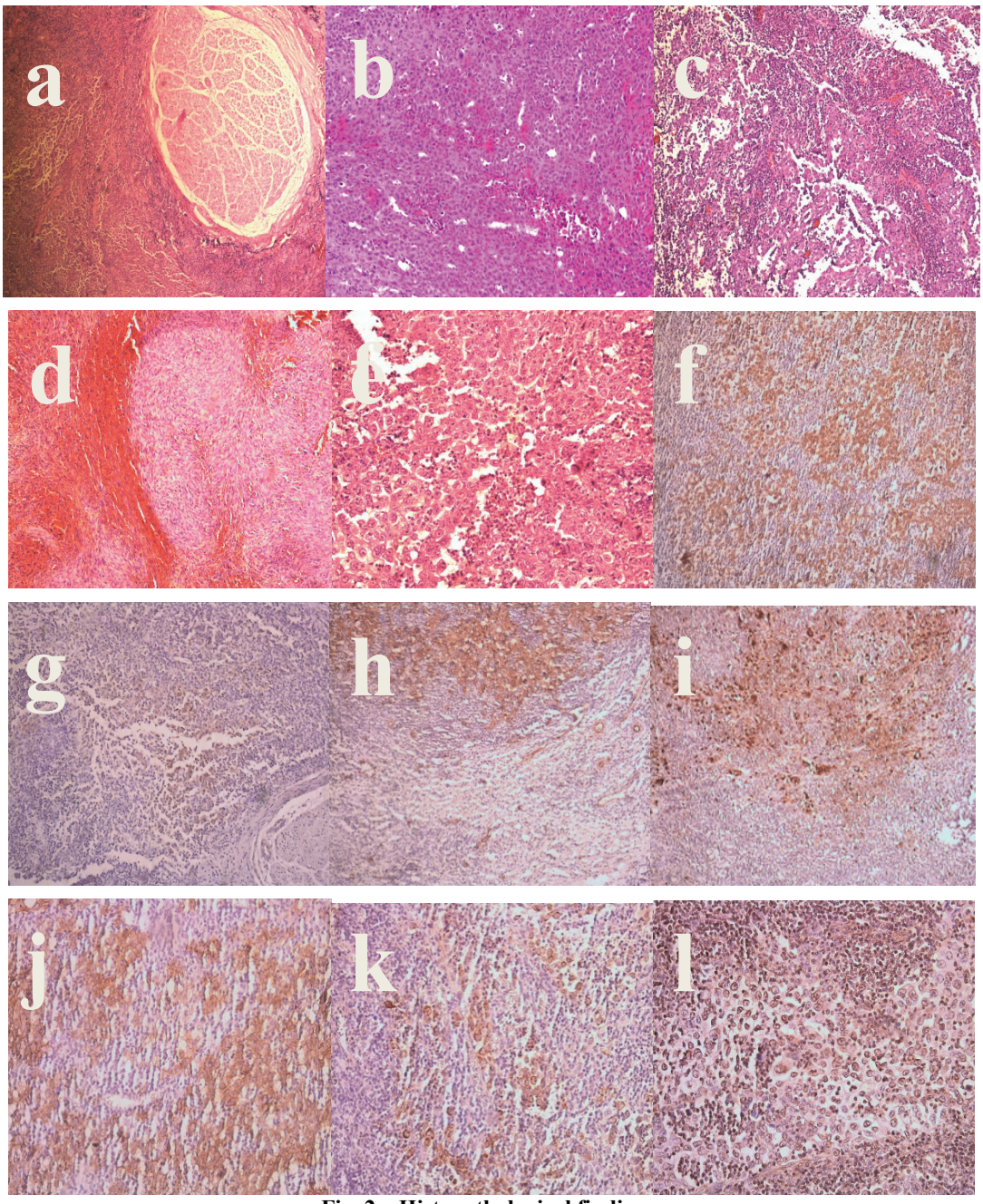

Fig. 2 – Histopathological findings.
a) Hematoxylin and eosin stain (H&E), 5 x 10; b) H&E, 10 x 10; c) H&E, 10 x 10; d) H&E, 10 x 10; e) H&E, 20 x 10; f) Positive staining for AE1/AE3 10 x 10; g) Positive staining for CA125 10 x 10; h) Positive staining for CD34 10 x 10; i) Moderate staining for CEA 10 x 10; j) Positive staining for EMA 20 x 10; k) Positive staining for MNF116 20 x 10; l) Positive staining for vimentin 20 x 10.

Discussion

ES is an aggresisive and rare soft tissue tumor, which was first described by Enzinger [6] in 1970, even with currently available therapy. After reviewing the avaible literature data, we did not find any case of ES at femoral nerve, and, as far as we know, it was the first case of ES at this anatomical location.

Differential diagnosis of epithelioid sarcoma from other forms of cancer is challenging and require various immunohistochemical stain analysis [7].

The prognosis of epithelioid sarcoma is very poor, but multimodal treatment is clearly important to raise the average survival rate of patients. The treatment of choice for ES is wide surgical resection and adjuvant chemoradiotherapy with ifosamide and adriamycin, and in some cases gemcitabine/docetaxel for an aggressive recurrent tumor [7–12]. The role and choice of adjuvant hemiotherapy still remains unclear because it was rare disease [9]. Patients who have had adjuvant radiation therapy show lower recurrence rates [9–11].

It was reported that five years of overal survival (OS) for ES after surgery and chemoirradiaton was 32–78% [8]. OS depends of tumor size, local recurrence (proximal location has poorer prognosis then distal), lymph node involvement, number of mitoses, tumor necrosis, hemorrhage and vascular invasion [1, 8–11]. ES have a high rate of distant metastases especially in lungs and lymph nodes, ranging from 40% to 57% [1, 2, 7, 10, 11].

Development of target antibody therapy might provide alternative therapeutic options. Thway et al. [12] found that inactivation of SMARCB1/IN1 (tumor suppressor gene) played a crucial role in tumorogenesis of ES. Immunochemical studies revealed that 85% to 93% of cases have inactivated SMARCB1/IN1 and target therapy restoring SMARCB1/IN1 gene function could provide new therapy in ES treatment.

There is still a question about prophylactic lymph node dissection for the ES patients. Curent data suggest that lymph node dissection is indicated only in cases when lymph node involvement was observed by preoperative diagnostic imaging (MRI, PET-CT). After HP conformation that nodes are positive, we can perform serial lymph node dissection and adjuvant radiation therapy [10].

Conclusion

Our case suggests that early diagnosis of ES with radical wide resection can improve prognosis. Novel strategies are needed to improve the survival of patients with these highly aggressive sarcomas.

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