Case Report

Eur Arch Med Res 2018; 34 (3): 200-2

A Rare Tumor, a Rare Localization: A Malignant Small Round Cell Tumor in the Thigh

Özlem Çolak[1], Ayça Ergan Şahin[1], Özay Özkaya Mutlu[1], Tülay Sayılgan[2]

[1]Clinic of Plastic, Reconstructive and Esthetics Surgery, University of Health Sciences Okmeydanı Training and Research Hospital, İstanbul, Turkey
[2]Clinic of Pathology, University of Health Sciences Okmeydanı Training and Research Hospital, İstanbul, Turkey

Abstract

Small round cell malignant tumors are the general term given to tumors that are small, round, and usually develop from undifferentiated cells. Tumors included in this group are: Ewing sarcoma (peripheral neuroectodermal tumor), primitive neuroectodermal tumor, rhabdomyosarcoma, synovial sarcoma, non-Hodgkin lymphoma, cutaneous neuroendocrine carcinoma (Merkel cell carcinoma), retinoblastoma, peripheral neuroblastoma, hepatoblastoma, and nephroblastoma or Wilms' tumor. In histopathological examinations, positive markers, such as cytokeratin, epithelial membrane antigen, desmin, vimentin, neuron-specific enolase, and CD15, can be found. The patient consulted our clinic with a diagnosis of neuroectodermal small round cell malignant tumor after an excisional biopsy was performed at an external center following a complaint of a mass in the thigh. A 40-year-old female patient with a complaint of a mass in the right femoral posterior region for over 1 year had excisional biopsy performed at an external center with a diagnosis of malignant tumor infiltration. Biopsy material was evaluated by our hospital's pathology clinic. The result was found to be small round cell malignant tumor. The patient was admitted to our clinic for re-excision. Re-excision was made from the previous incision line on the posterolateral of the right thigh with a 2 mm surgical border. Small round cell tumors are a heterogeneous group of tumors composed of malignant neoplasia. Even though they are often observed in childhood, some primitive neuroectodermal variants may be seen in adults. Owing to their low surveys, early diagnosis and aggressive treatment are significant in these cases.

Keywords: Malignant small round cell tumor, neuroectodermal tumor, primitive tumor

Cite this article as:
Çolak Ö, Ergan Şahin A, Özkaya Mutlu Ö, Sayılgan T. A Rare Tumor, a Rare Localization: A Malignant Small Round Cell Tumor in the Thigh. Eur Arch Med Res 2018; 34 (3): 200-2

Corresponding Author:
Ayça Ergan Şahin
E-mail
draycaergan@gmail.com

Received: 06.06.2017
Accepted: 09.11.2017
DOI:10.5152/eamr.2018.46503

INTRODUCTION

Malignant small round cell tumor is a general term for a collection of tumors, which usually develop from small, round, and undifferentiated cells and have a high nucleus/cytoplasm ratio. The tumors included in this group are Ewing's sarcoma (peripheral neuroectodermal tumor), primitive neuroectodermal tumor (PNET), rhabdomyosarcoma, synovial sarcoma, non-Hodgkin's lymphoma, cutaneous neuroendocrine carcinoma (Merkel cell carcinoma), retinoblastoma, peripheral neuroblastoma, hepatoblastoma, and nephroblastoma (Wilm's tumor). Other tumors in the group of small round cell tumors are small cell osteosarcoma, undifferentiated hepatoblastoma, granulocytic sarcoma, and intraabdominal desmoplastic small round cell tumor (1-6). Sarcomas are the most common small round cell tumors that spread cutaneously. Although they can primarily be seen as cutaneous lesions, they may also be seen as metastatic lesions (5).

Differential diagnosis of malignant small round cell tumors is difficult because of their primitive character. Positive markers, such as cytokeratin, epithelial membrane antigen (EMA), desmin, vimentin, neuron specific enolase (NSE), and CD15, can be found in histopathological examinations (2). In this case report, the treatment process of a patient in whom an excisional biopsy was

performed in an external center due to a mass on the thigh and whose pathology result was malignant small round cell tumor originating from the endoscopic neuroectoderm is described. PNETs were first described by Stout (7) in 1918 and were classified under the Ewing sarcoma family. Batsakis et al. (8) divided PNETs into three groups: those originating from the central, autonomous, and peripheral nervous systems. Particularly, tumors which are originated from peripheral nervous system are rare (7).

CASE PRESENTATION

A 40-year-old female patient was admitted to our clinic when the result of an excisional biopsy performed at an external center due to a mass in the posterior part of the right thigh showed a malignant tumor infiltration. The biopsy material was evaluated by the pathology clinic of our hospital Figures (1-4). The patient was hospitalized in our clinic for re-excision when the result was malignant small round cell tumor.

In the physical examination, a 6 cm-, oblique-, and hyperpigmented-incision scar was observed between the posterolateral proximal onethird and middle onethird of the right thigh. No palpable mass was observed in the thigh. No mass was seen in the bilateral inguinal and popliteal regions. No pathology was found in the abdominal computed tomography (CT), gluteal and pelvic region magnetic resonance imaging, and positron emission tomography (PET)-CT scans of the patient. The patient was operated under general anesthesia. Re-excision was performed through the previous incision line with a 2-cm surgical margin in the posterolateral region of the right thigh, and the muscle fascia on the base was also included in the excision material. The resulting defects were primarily repaired. No problems occurred in the early postoperative follow-up of the patient.

DISCUSSION

Malignant small round cell tumors are a heterogeneous group of neoplasms, and they are often observed in children or young

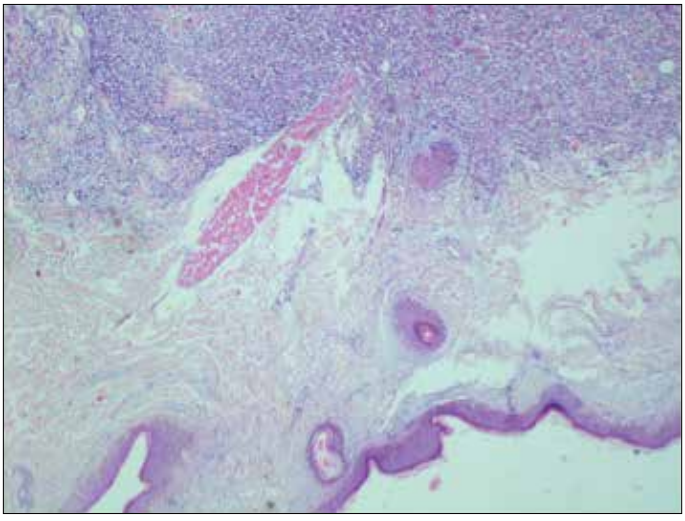

Figure 1. Infiltration of malignant small round cell tumor cells under the skin (hematoxylin–eosin staining, 40X)

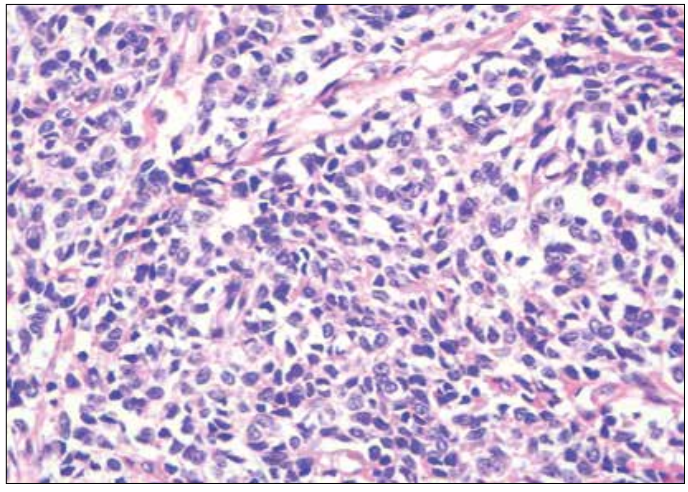

Figure 3. Malignant cells that have a uniform appearance, spread in a diffuse pattern, and have a hyperchromatic nucleus and narrow cytoplasm (hematoxylin–eosin staining, 400X)

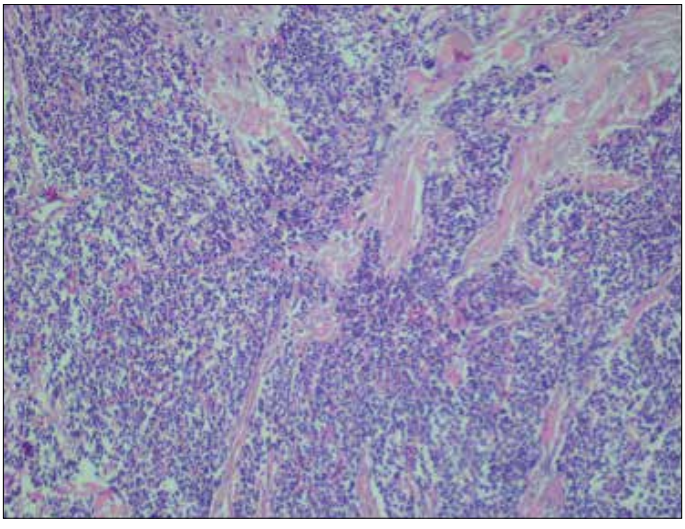

Figure 2. Infiltration of malignant small round tumor cells (hematoxylin–eosin staining, 100X)

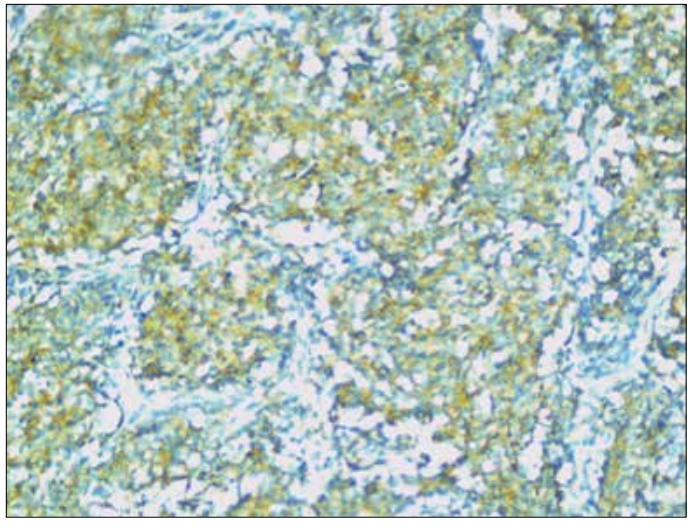

Figure 4. Immunohistochemical diffuse neuron specific enolase (NSE) staining in tumor cells (NSE, 400X)

adult males (9). It has a predisposition of involving the skeletal system and soft tissues. This tumor group is composed of cytomorphologically similar neoplasms with different origins. Ewing sarcoma, PNET, rhabdomyosarcoma, synovial sarcoma, neuroblastoma, lymphomas, and desmoplastic small round cell tumors are the most common tumors of this group (3). In the differential diagnosis, minor differences are effective in terms of the patient's age, the location of tumor, and structural and cytoplasmic properties. Immunohistochemical staining and chromosomal studies are useful in classifying the tumors (4).

Malignant small round cell tumors may occur in specific regions of the body, such as the abdomen, gonads, head and neck, upper thorax, and internal viscera. Some malignant small round cell tumors are very rare, but they can occur in different parts of the body. Involvement of the extremities is very rare.

An exact protocol has not been established for the treatment of these tumors. Surgery, chemotherapy, radiotherapy, and combined treatments can be applied according to the size and localization of the tumor. Due to the aggressive clinical pattern, an accurate diagnosis is important for the application of correct treatment and for not applying unnecessary procedures in patients. Both early surgical excision and extensive surgical excision (performing tumor excision with a 2–3 cm tumor-free margin) due to local recurrence after resection are the important points to be considered during the treatment (11). Adjuvant chemotherapy regimens containing agents, such as vincristine, doxorubicin, and cyclophosphamide, and/or adjuvant radiotherapy are also recommended in patients who are diagnosed at an early stage (9-11).

CONCLUSION

Although the prognosis is poor, care should be taken in terms of making the definite diagnosis and applying the correct treatment in cases of rare and atypical tumors. The best treatment is early surgical excision and adjuvant chemoradiotherapy in selected patients.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – Ö.Ç.; Design – Ö.Ç., A.E.Ş.; Supervision – Ö.Ö.; Resources – A.E.Ş.; Materials – Ö.Ö.; Data Collection and/or Processing – A.E.Ş.; Analysis and/or Interpretation – T.S.; Literature Search – A.E.Ş.; Writing Manuscript – Ö.Ç., A.E.Ş., Ö.Ö.; Critical Review – Ö.Ö.; Other – T.S.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Rajwanshi A, Srinivas R, Upasana G. Malignant small round celltumors. J Cytol 2009; 26: 1-10. [CrossRef]
2. Akalın T, Kandiloğlu G. Desmoplastik küçük yuvarlak hücreli tümör (üç olgu sunumu). Türk Patoloji Dergisi 2000; 16: 103-5.
3. Lee JH, Seo JH, Park KH, Jun BC. A Case of Malignant Small Round Cell Tumor of Temporal Bone with Facial Paralysis. Korean J Audiol 2012; 16: 145-7. [CrossRef]
4. Reshma V, Rao K, Priya NS, Umadevi HS, Mohsin G, Hosthor S. Small Round Cell Tumor of the Head and Neck Region: A Review. International Journal of Oral and Maxillofacial Pathology 2013; 4: 24-33.
5. Machado I, Traves V, Cruz J, Llombart B, Navarro S, Lombart-Bosch A. Superficial small round-cell tumors with special reference to the Ewing's sarcoma family of tumors and the spectrum of differential diagnosis. Semin Diagn Pathol 2013; 30: 85-94. [CrossRef]
6. Devoe K, Weidner N. Immunohistochemistry of small round-cell tumors. Semin Diagn Pathol 2000; 17: 216-24.
7. Stout AP. A tumour of the ulnar nerve. Proc NY Pathol Soc 1918; 18: 2-11.
8. Batsakis JG, Mackay B, el-Naggar AK. Ewing's sarcoma and peripheral primitive neuroectodermal tumour: an interim report. Ann Otol Rhinol Laryngol 1996; 105: 838-43. [CrossRef]
9. Soma S, Shetty SK, Bhat S. PNET of the Abdominal Wall: A Rare Presentation. J Clin Diagn Res 2015; 9: XD01-XD02. [CrossRef]
10. Thomas BP, Sasi K, Pallapati SCR, Mathew A, Sreekanth R, Thomas M. Malignant tumours of the hand and wrist. Indian J Plast Surg 2011; 44: 337-47. [CrossRef]
11. Grassetti L, Torresetti M, Brancorsini D, Rubini C, Lazzeri D, Di Benedetto G. A peculiar case of large primary cutaneous Ewing's sarcoma of the foot: Case report and review of the literature. Int J Surg Case Rep 2015; 15: 89-92. [CrossRef]