Case Report

Malignant Peripheral Nerve Sheath Tumor of Breast in Patient without Von Recklinghausen's Neurofibromatosis: A Case Report

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Malignant peripheral nerve sheath tumor (MPNST) of the breast without Von Recklinghausen's neurofibromatosis (VRN) is extremely rare. The authors report a 19 year-old woman who presented with a 12 month history of a painless mass of the left breast. Tissue biopsy was performed. The histologic diagnosis was made with immunohistochemical study in which the tumor showed positivity of vimentin, S-100 protein, neuron-specific protein(NSE), neurofilament protein(NF) and glial fibrillary acidic protein(GFAP). The patient was referred for radiation therapy after simple mastectomy.

Keywords: *Breast tumor, Malignant peripheral nerve sheath tumors, von Recklinghausen's neurofibromatosis*

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Malignant peripheral nerve sheath tumors (MPNST) account for approximately 5-10% of all soft tissue sarcomas and about half of them occur in association with Von Recklinnghausen's neurofibromatosis (VRN). The incidence of MPNST in patients with VRN is around 4%, compared to an incidence of 0.001% in the general clinical population[1]. The common sites of MPNST are trunk (46%), extremities (34%), head and neck (19%)[2]. MPNST of the breast with and without VRN is a rare tumor and only a few cases have been reported[3,4].

Case Report

A 19 year-old woman presented with a 12 month history of a large painless mass in the upper outer quadrant of her left breast. The mass was slow growing but there were no other breast-related symptoms. She noticed minimal bloody fluid from left nipple in the last 3 months.

On physical examination, she had a 7 cm in diameter,movable, painless, cystic mass in the upper outer quadrant of her left breast. No axillary lymph node enlargement or sign of neurofibromatosis were detected. The ultrasonography of the breast demonstrated a large hypoechoic mass with a well defined border in the upper and lower parts of the left breast. The mass contained cystic area with septation and a solid part. Chest radiogram and ultrasonography of the abdomen showed no evidence of metastatic disease.

Fine needle aspiration (FNA) was performed and 25 ml. of serosanguinous fluid was aspirated from the mass. Cytological examination found histiocytes and neutrophils but no ductal cell. The mass was shrunk after FNA however grew back to the same size within 2 weeks. The patient underwent an incisional biopsy and operative finding revealed a multiloculated cyst which contained bloody discharge and hypervascular, fliable soft tissues. No invasion to the skin and chest wall was found.

The pathological examination showed spindle shape cell tumor composing necrosis, pleomorphism and a few mitotic figures. Immunohistochemical study revealed tumor expressed vimentin, S-100 protein, NSE, NF and GFAP consistent with malignant peripheral nerve sheath tumor (Fig. 1-3)[8,9].

The patient underwent simple mastectomy under general anaesthesia. The pathological examina-

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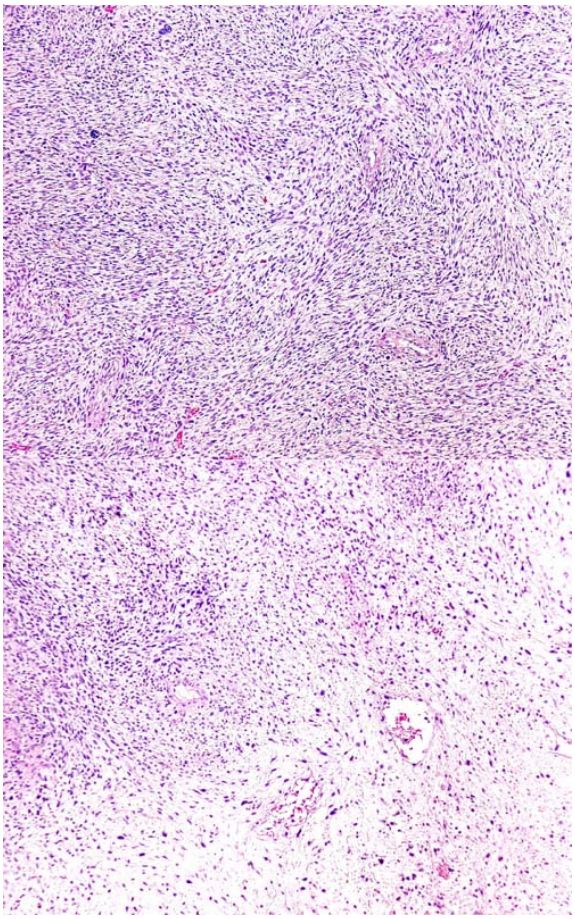

Fig. 1 Malignant peripheral nerve sheat tumor of breast shows dense hypercellular fascicles (upper) alternate with hypocellular area (lower) (100, both)

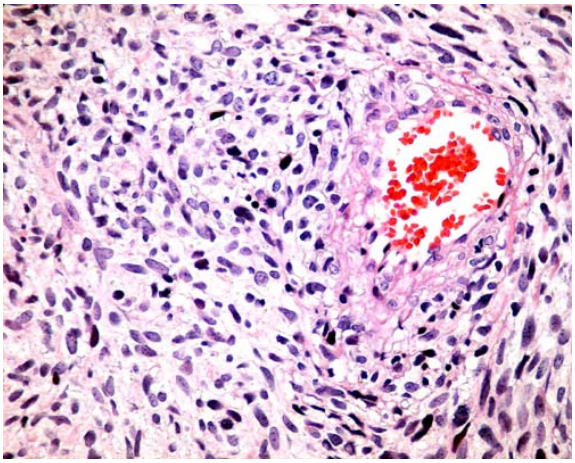

Fig. 2 Malignant peripheral nerve sheath tumor with pleomorphic areas (400)

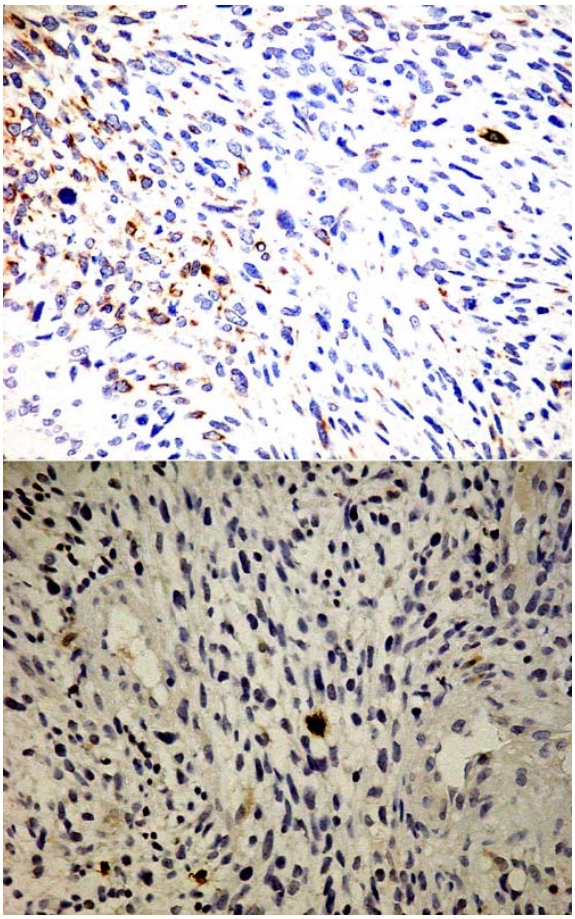

Fig. 3 Immunohistochemistry stains GFAP (upper) and S100 (lower)

tion revealed a free surgical margin. The patient was referred for postoperative radiation therapy.

Discussion

Although previously known as neurofibrosarcoma, neurogenic sarcoma, malignant neurilemmoma, and malignant schwannoma, MPNST is the currently preferred term for this group of tumors because whether they arise from schwann cells or from peripheral fibroblasts is not clearly understood[5].

MPNST is often associated with VRN[2]. Without the evidence of VRN (multiple caf au lait spots, cutaneous neurofibromas, evidence of visceral or central von Recklinghausen's disease, a positive family history of phakomatosis), the clinical diagnosis of MPNST of the breast is difficult. The radiologic pattern and role of cytology in preoperation diagnosis are still uncertain[4]. Histological examination is the most important tool for diagnosis. Currently, electron

microscopy and immunohistochemistry enable better diagnosis in those questionable patients[5].

Because of the long history of the mass, the cystic consistency, and radiological pattern, the initial diagnosis of the presented case was fibrocystic disease and FNA was not helpful. Final diagnosis was made from the immunohistochemical profile of the tumor cells.

Treatment of MPNST should include aggressive surgery with wide surgical margins combined with adjuvant radiation therapy[5,6]. The role of chemotherapy is unclear[5,7].

The MPNST is an extremely rare tumor, and large tumor size, tumor location, the presence of VRN, and total resection are the most important prognostic factors[2].

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เนื้องอกปลอกประสาทชนิดร้ายของเต้านมในผู้ป่วยหญิงที่ไม่มีโรคเท้าแสนปม: รายงานผู้ป่วย 1 ราย

ไชยยุทธ ธนไพศาล, สุพินดา คูณมี, สริธร ศิริธันยาภรณ์

เนื้องอกปลอกประสาทชนิดร้ายของเต้านมในผู้ป่วยหญิงที่ไม่มีโรคเท้าแสนปมพบได้น้อยมาก ผู้เขียนรายงานผู้ป่วย 1 ราย ซึ่งมาพบแพทย์ด้วยปัญหามีก้อนที่เต้านมซ้ายมา 12 เดือน แพทย์ได้ทำการผ่าตัดเพื่อนำชิ้นเนื้อมาตรวจทางพยาธิวิทยา การตรวจโดยวิธี immunohistochemistry ให้ผลบวกต่อ ไวเมนติน, โปรตีน เอส-100, neuron-specific protein (NSE), neurofilament protein (NF) และ glial fibrillary acidic protein (GFAP) ซึ่งเข้าได้กับเนื้องอกดังกล่าว การรักษาทำโดยการผ่าตัดเต้านมออกทั้งหมด และ ฉายรังสีภายหลังการผ่าตัด