Case Report

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대한영상의학회지

Extraskeletal Ewing Sarcoma of the Chest Wall Manifesting as a Palpable Breast Mass: Ultrasonography, CT, and MRI Findings

만져지는 유방 종괴로 발현된 가슴벽 골외 유잉씨 육종의 초음파, 전산화단층촬영 및 자기공명영상 소견

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Ewing sarcomas constitute a group of small, round, blue cell tumors of the bone and soft tissue. Extraskeletal Ewing sarcoma (EES) is a rare malignant neoplasm that arises from soft tissues, and it usually affects children and young adults. EES of the thoracopulmonary region commonly presents with a palpable mass or pain. Although rarely reported, EES affecting the anterior chest wall may present as a breast mass. We report a case of EES arising from the chest wall and manifesting as a palpable breast mass in a 22-year-old woman. The large mass was initially misdiagnosed as a breast origin mass on ultrasonography, but subsequent CT and MRI showed that the mass originated from the chest wall. Radiologists should be aware of the imaging findings of EES, and they should understand that chest wall lesions may be clinically confused as breast lesions.

Index terms Chest Wall; Computed Tomography, X-Ray; Ewing Sarcoma; Magnetic Resonance Imaging; Ultrasonography

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INTRODUCTION

Ewing sarcoma family of tumors is a group of malignant, small, and round blue cell tumors of bone and soft tissue, including skeletal Ewing sarcoma, extraskeletal Ewing sarcoma (EES), peripheral primitive neuroectodermal tumor (PNET), and Askin tumor (thoracopulmonary EES or PNET) (1, 2). The prevalence of EES is only about 15–20% of that of skeletal Ewing sarcoma (1). This rare malignant neoplasm usually affects young

adults and children (3). The chest wall is one of the most common sites, and EES usually manifests as a large tumor involving the chest wall and pleura with an eccentric growth pattern (2, 4). We report the imaging features of a 22-year-old woman with EES arising in the chest wall manifesting as a palpable breast mass and initially diagnosed as a breast tissue mass.

CASE REPORT

A 22-year-old woman presented with a palpable mass in the right breast that had been present for 3 months. On physical examination, the right breast was asymmetrically enlarged with a firm and non-tender palpable mass.

Ultrasonography (US) of the breast revealed a 14-cm, oval, complex cystic, and solid mass involving the entire right breast and exceeding the US field of view. The superficial margin of the mass was circumscribed, but the deep margin was not visible on US because of the large size. The complex cystic and solid mass showed heterogeneous internal echogenicity and multiloculated cystic components were mainly located in the superficial area of the mass (Fig. 1A, left). Color Doppler US revealed internal vascularity in the solid portion of the mass (Fig. 1A, right). There was no evidence of lymphadenopathy in both axillae. Initially, the mass was thought to be of breast origin because it was difficult to distinguish whether the origin was intramammary or extramammary on US. We suspected that the mass was malignant according to the Breast Imaging Reporting and Data System (BI-RADS, category 4B), because it was rapidly growing, palpable lump with huge size and the mass showed internal heterogeneous echogenicity with complex cystic and solid portion and internal vascularity.

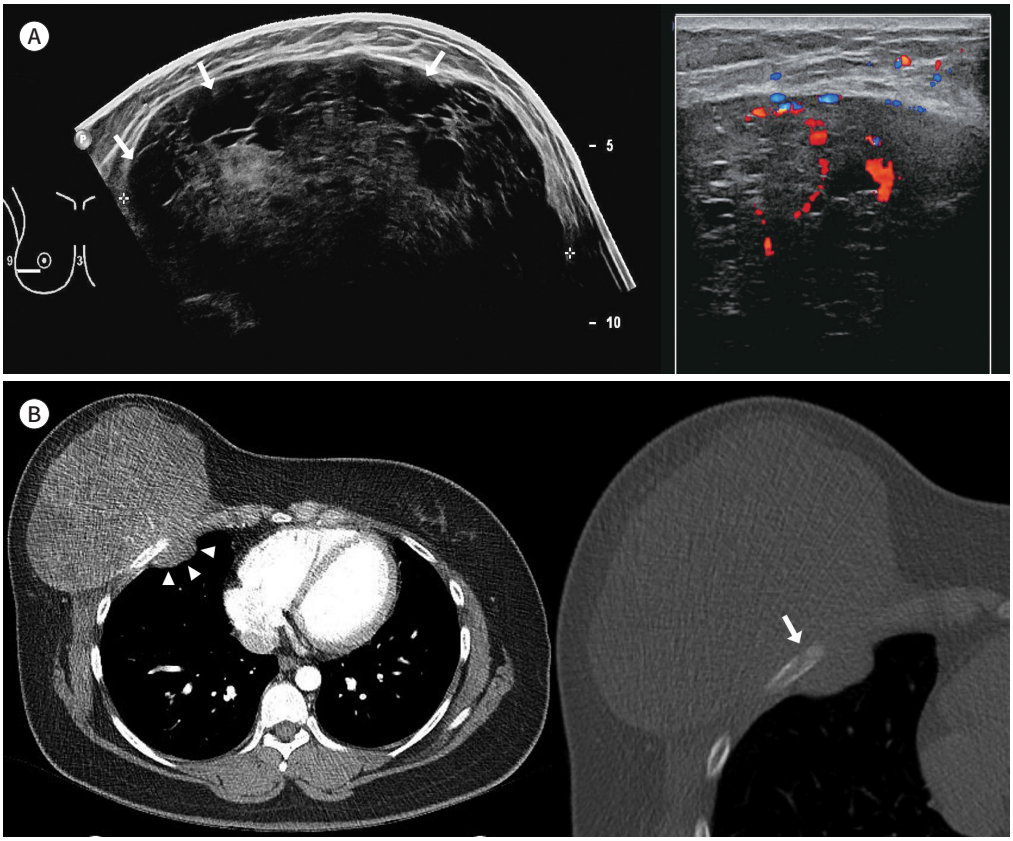
Additional contrast-enhanced chest CT was performed to evaluate the extent of the lesion. Chest CT revealed a 14-cm, multilobulated, and heterogeneously enhancing mass with multiple cystic components, which originated from the right chest wall. There was no calcification within the mass. The mass showed focal extension to the intrathoracic area and destruction of the adjacent right fifth rib (Fig. 1B).

US-guided core needle biopsy was performed, and the histopathologic examination revealed a diagnosis of Ewing sarcoma. Six cycles of neo-adjuvant chemotherapy were administered, and additional US, CT, and MRI were performed to evaluated the response to chemotherapy and operability. The mass decreased in size from 14 to 12.5 cm in maximum diameter, and there were many cystic or necrotic changes in the mass on US and CT images after neo-adjuvant chemotherapy.

MRI showed an oval-shaped, circumscribed, and heterogeneous mass containing multiloculated fluid collections, suggesting necrosis within the mass, which originated from the right pectoralis major muscle. The solid portion of the mass showed intermediate signal intensity with dark internal septa on the T2-weighted image (T2WI). On dynamic contrast enhancement, the mass showed heterogeneous enhancement. The breast parenchyma was compressed superficially by the mass, without invasion. The mass extended to the thoracic cage with bony destruction of the fifth rib (Fig. 1C).

The patient underwent surgical excision of the mass after six cycles of neo-adjuvant chemotherapy. The round mass originating from the right pectoralis muscle was observed intra-

Fig. 1. A 22-year-old woman with Ewing sarcoma arising from the chest wall and presenting as a palpable mass in her right breast.
A. Ultrasonography image reveals a large, oval, complex cystic, and solid mass. The mass shows heterogeneous internal echogenicity, and multiloculated cystic components are mainly located within the superficial area of the mass (arrows) (left image). There is internal vascularity within the solid portion of the mass on color Doppler ultrasonography (right image).
B. A contrast-enhanced chest CT axial image with a mediastinal window setting shows a multilobulated and heterogeneously enhancing mass with multiple cystic components in the right chest wall. The mass shows a focal extension to the intrathoracic area (arrowheads) (left image). Axial CT scan with bone window setting demonstrates the destruction of the adjacent right fifth rib (arrow) (right image).

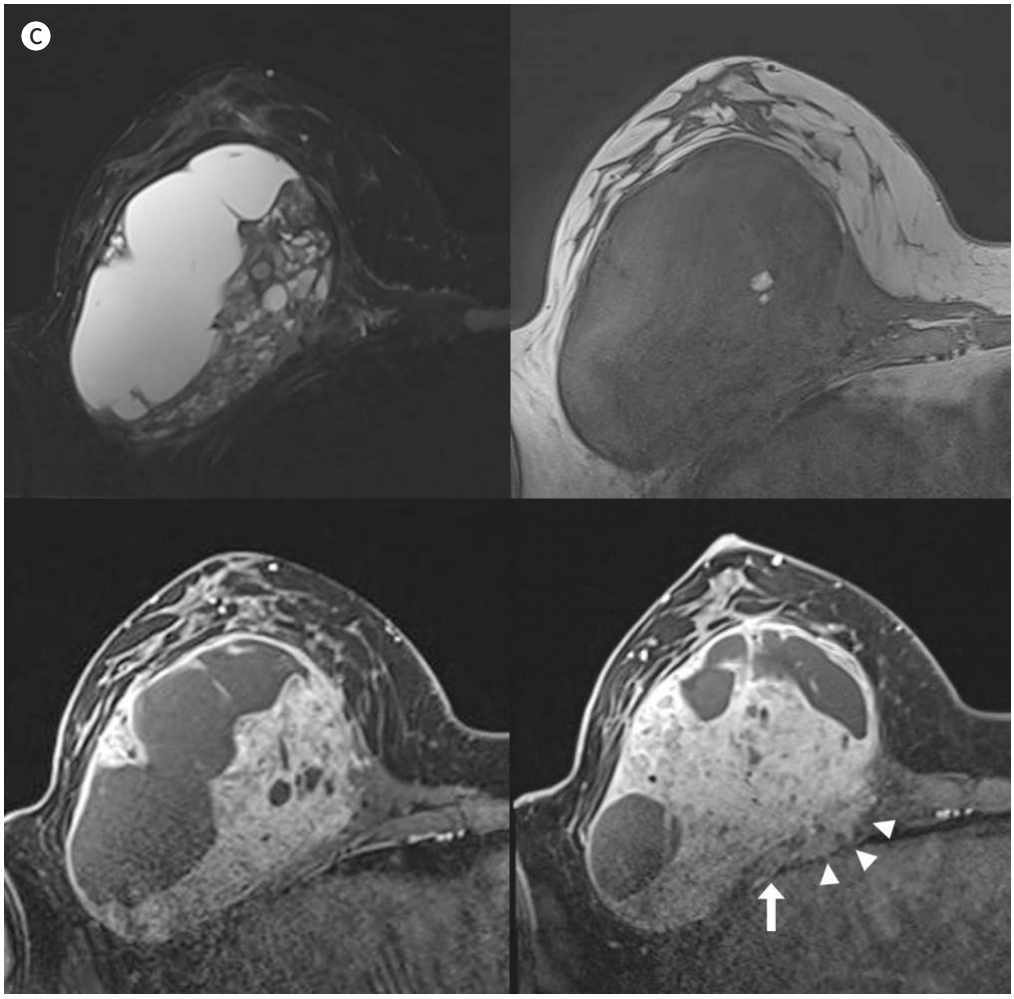


operatively and was easily distinguishable from the breast parenchyma. The mass had invaded into the thoracic cage through the fourth intercostal space, but the parietal pleura was not involved. The mass was excised with partial resection of the right fourth and fifth ribs.

On gross examination, the excised mass measured 12.5 cm × 12.0 cm × 10.0 cm in size. On section, the tumor was grayish-brown and soft, with hemorrhage and cystic areas (Fig. 1D, upper left). The tumor showed direct invasion into the right fifth rib. Histologically, the tumor was composed of uniform, small round cells (Fig. 1D, upper right). The tumor cells had finely granular chromatin, inconspicuous nucleoli, and scanty cytoplasm. The tumor cells were arranged in lobular or sheets pattern. The mitotic rates were 31 per 10 high power fields. On immunohistochemical staining, the tumor cells showed diffuse membranous positivity for CD99 (Fig. 1D, lower left) and diffuse nuclear positivity for NKX2.2 (Fig. 1D, lower right). Histologic findings and immunohistochemical results were consistent with EES.

The patient underwent additional three cycles of chemotherapy after surgery. After 6

Fig. 1. A 22-year-old woman with Ewing sarcoma arising from the chest wall and presenting as a palpable mass in her right breast.
C. MRI, after the sixth cycle of neoadjuvant chemotherapy, reveals an oval, circumscribed, and heterogeneous mass in the right pectoralis major muscle. The mass shows multiloculated, heterogeneously iso- to high signal intensity fluid collections of various phase hemorrhages on a T2-weighted image (upper left panel) and an intermediate signal intensity (isointense to adjacent muscle) with high signal intensity foci representing a hemorrhage on a T1-weighted image (upper right panel) show heterogeneous enhancement within the solid portion of the mass and a focal extension to the thoracic cage (arrowheads) with rib destruction (arrow).



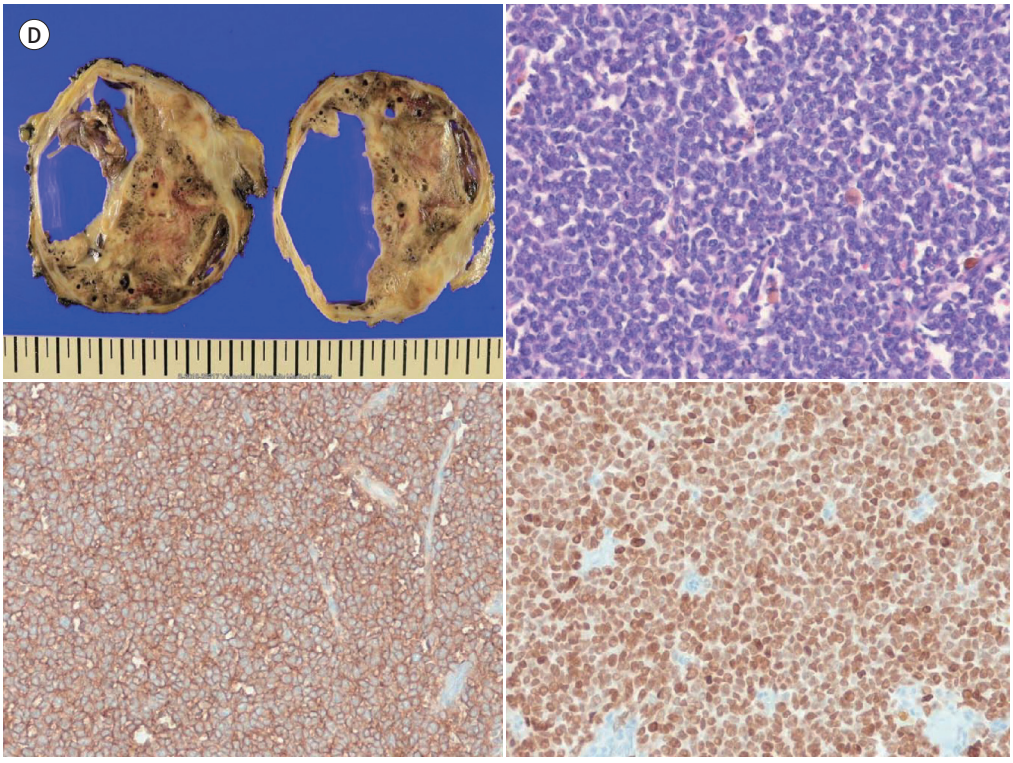
months from surgery, the patient had no recurrence or metastasis.

DISCUSSION

Chest wall tumors account for approximately 5% of all thoracic malignancies. More than half of all chest wall tumors are malignant, including primary malignancies and secondary invasion or metastasis from thoracic tumors. Ewing sarcoma is the third most common cause of primary chest wall malignancy and the most common primary malignant chest wall tumor in children and young adults (5).

Ewing sarcoma is a highly malignant, small round blue cell tumor, which was first de-

Fig. 1. A 22-year-old woman with Ewing sarcoma arising from the chest wall and presenting as a palpable mass in her right breast.
D. Gross and microscopic findings. The tumor is well-circumscribed, grayish brown, and soft, with focal hemorrhaging and a cystic area (upper left panel). The tumor is composed of sheets of uniform, small, round cells (hematoxylin and eosin stain, × 200) (upper right panel). The tumor cells show diffuse membranous positivity for CD 99 (lower left panel) and diffuse nuclear positivity (lower right panel) (immunohistochemical stain, × 200).



scribed in 1921 by Ewing (6). Skeletal Ewing sarcoma, EES, PNET, and Askin tumor (thoracopulmonary EES or PNET) have histologically similar appearances and are cytogenetically closely related to a common karyotype abnormality involving chromosomes 11 and 22. Thus, they are now thought to belong to the same tumor family, referred to as Ewing sarcoma family of tumors (1, 2).

EES is quite rare compared to skeletal Ewing sarcoma and usually affects children and young adults, between the age of 20 months and 30 years (1). The most common sites for EES include the paravertebral region, lower extremities, chest wall, retroperitoneum, pelvis and hip, and upper extremities (4). As in our case, EES arising in the chest wall usually manifests as a large, rapid-growing mass with or without pain (3). Our patient presented with complaints of a palpable breast mass, raising a suspicion of a breast origin lesion. However, the tumor was located in the chest wall, especially in the pectoralis major muscle, and had invaded the adjacent soft tissue and bony structures. The normal breast parenchyma was compressed by the huge mass without invasion. The chest wall mass, especially involving the anterior chest wall, can be confused with breast mass as clinical features.

The imaging features of EES are nonspecific. On US, they are often hypoechoic, but can have anechoic areas, representing necrosis or hemorrhage (1, 7). Breast US can be performed

initially for the evaluation of the chest wall lesion because it is easily accessible and there is no risk of radiation exposure. However, US with a high-frequency linear transducer has limits in evaluating the deep structures; sometimes, it is difficult to distinguish between the breast lesion and chest wall lesion (8). In our case, the mass was thought to be of breast origin on initial US. We considered the possibility of giant juvenile fibroadenoma or phyllodes tumor based on the patient's young age and the huge size of the mass. This highlights the importance of knowing that not all breast masses arise from the breast itself, and if the lesions are suspected to arise from the chest wall on US, additional studies such as CT or MRI are necessary to determine the exact site of origin.

CT is a useful modality to evaluate chest wall neoplasms with revealing the presence of lesion, site and tissue origin, morphologic features, internal components such as fat and mineralization. Also, tumor vascularity can be identified with administration of contrast material (5). EES arising in the chest wall is frequently associated with rib destruction and pleural effusion. In this case, adjacent rib was destructed by the mass, but there was no intratumoral calcification or pleural effusion on CT images.

Although the MRI features are non-specific, MRI is the optimal imaging modality for delineating the extent of the lesion and assessing its relationship with the surrounding structures because of the superior soft tissue contrast (7). MRI often demonstrates a large heterogeneous mass with predominant intermediate signal intensity (iso- or hyperintense to muscle) on T1WIs and high signal intensity on T2WI. Areas of hemorrhage or necrosis can be seen as regions of high signal intensity on both T1- and T2WI. The lesions usually demonstrate intense heterogeneous enhancement after the administration of intravenous contrast material (1, 2, 4, 7, 9).

Differential diagnoses of chest wall tumors in children and young adults include primary (Ewing sarcoma, rhabdomyosarcoma, osteosarcoma, neuroblastoma) or secondary malignancies (metastasis, lymphoma/leukemia). As the imaging features of these chest wall malignancies are non-specific, it is difficult to distinguish between these unusual tumors (5, 7).

Ewing sarcoma family of tumors is treated by combination therapy including neoadjuvant chemotherapy and surgical resection. Adjuvant chemotherapy and supplementary radiation therapy could be considered (1, 2).

In summary, we report a case of Ewing sarcoma arising in the anterior chest wall (pectoralis major muscle), presenting as a palpable breast mass, in a young woman. US demonstrated a huge, complex cystic, and solid mass. CT and MRI revealed a multilobulated, heterogeneous mass with multiple cystic components and heterogeneous enhancement, which was finally diagnosed as Ewing sarcoma. Radiologists should be aware of the imaging findings of EES and understand that chest wall lesions may be clinically confused as breast lesions.

Author Contributions

Conceptualization, K.M., L.S.E.; data curation, K.M., C.J.H.; formal analysis, K.M., L.S.E.; funding acquisition, L.S.E.; investigation, K.M., C.J.H.; methodology, K.M., L.S.E.; project administration, L.S.E.; resources, K.M., C.J.H.; software, K.M., L.S.E.; supervision, L.S.E.; validation, K.M., L.S.E.; visualization, all authors; writing—original draft, K.M.; and writing—review & editing, L.S.E., C.J.H.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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유잉육종계열의 종양은 뼈와 연부조직에 발생하는 악성 소원형청색세포종양이다. 골격외 유잉씨 육종은 드문 악성 종양으로 연부조직에 발생한 유잉육종의 한 형태이며, 소아와 젊은 성인에서 호발한다. 흉폐부위에 발생한 골격외 유잉씨 육종은 임상적으로 만져지는 종괴나 통증으로 나타난다. 골격외 유잉씨 육종이 앞가슴벽을 침범한 경우에는 유방 종괴로 나타날 수 있으나, 이러한 보고는 드물다. 저자들은 22세 여성에서 유방 종괴로 나타난 앞가슴벽에 발생한 유잉씨 육종의 증례를 보고한다. 초기의 초음파에서 이 거대 종괴는 유방에서 발생한 종괴로 오인되었으나, 추가적인 전산화단층촬영 및 자기공명영상에서 종괴는 흉벽에서 기원하였음을 알 수 있었다. 영상의학과 의사는 골격외 유잉씨 육종의 영상 소견을 알고, 흉벽의 병변이 임상적으로 유방 병변으로 오인될 수 있음을 이해하는 것이 중요하다.

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