



Retroperitoneal Extraskkeletal Osteosarcoma without Calcification Mimicking Pancreas Tumor: CT Imaging of a Case Report

췌장종양으로 오인된 육안적 석회화를 동반하지 않은 후복막강 골외성 골육종: CT 소견에 대한 증례 보고

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Retroperitoneal extraskkeletal osteosarcoma is a rare tumor. Typical imaging findings include a soft tissue mass accompanied with internal calcifications. The authors encountered a case involving a 44-year-old woman with a large retroperitoneal extraskkeletal osteosarcoma, without calcification, that mimicked a pancreatic tumor. The present report highlights computed tomography features of retroperitoneal extraskkeletal osteosarcoma, followed by a brief literature review. It is challenging for radiologists to diagnose retroperitoneal masses. However, in patients who present with large retroperitoneal masses, combined with clinical information including the elevation of serum alkaline phosphatase levels, retroperitoneal extraskkeletal osteosarcoma should be considered in the differential diagnosis, even if the mass does not exhibit a gross calcification on the imaging.

Index terms

Retroperitoneal Space
Osteosarcoma
Ossification
Tomography, X-ray Computed

INTRODUCTION

Retroperitoneal extraskkeletal osteosarcoma is rare malignant mesenchymal tumor with an osteoid component without contact to bony structure(s) (1). On computed tomography (CT), soft tissue tumor(s) show various levels of ossified matrix; therefore, it is crucial to detect calcification to narrow the differential diagnosis. However, approximately 50% of extraskkeletal osteosarcomas do not exhibit gross calcification on imaging studies. Therefore, it is difficult for radiologists to diagnose extraskkeletal osteosarcoma arising in the retroperitoneal space without calcification on CT scan. We report a case of retroperito-

neal extraskkeletal osteosarcoma, without calcification on CT scan, mimicking a pancreatic tumor, and describe its imaging features with ancillary clinical information to facilitate diagnosis.

CASE REPORT

A 44-year-old woman presented with a 10-month history of a palpable mass in the abdomen. Laboratory examinations were unremarkable except for elevation of alkaline phosphatase level [159 IU (normal range 40–130 IU)]. To evaluate her symptoms, abdominal CT was performed with contrast enhancement, which revealed a large (approximately 16.5 cm) soft tissue mass

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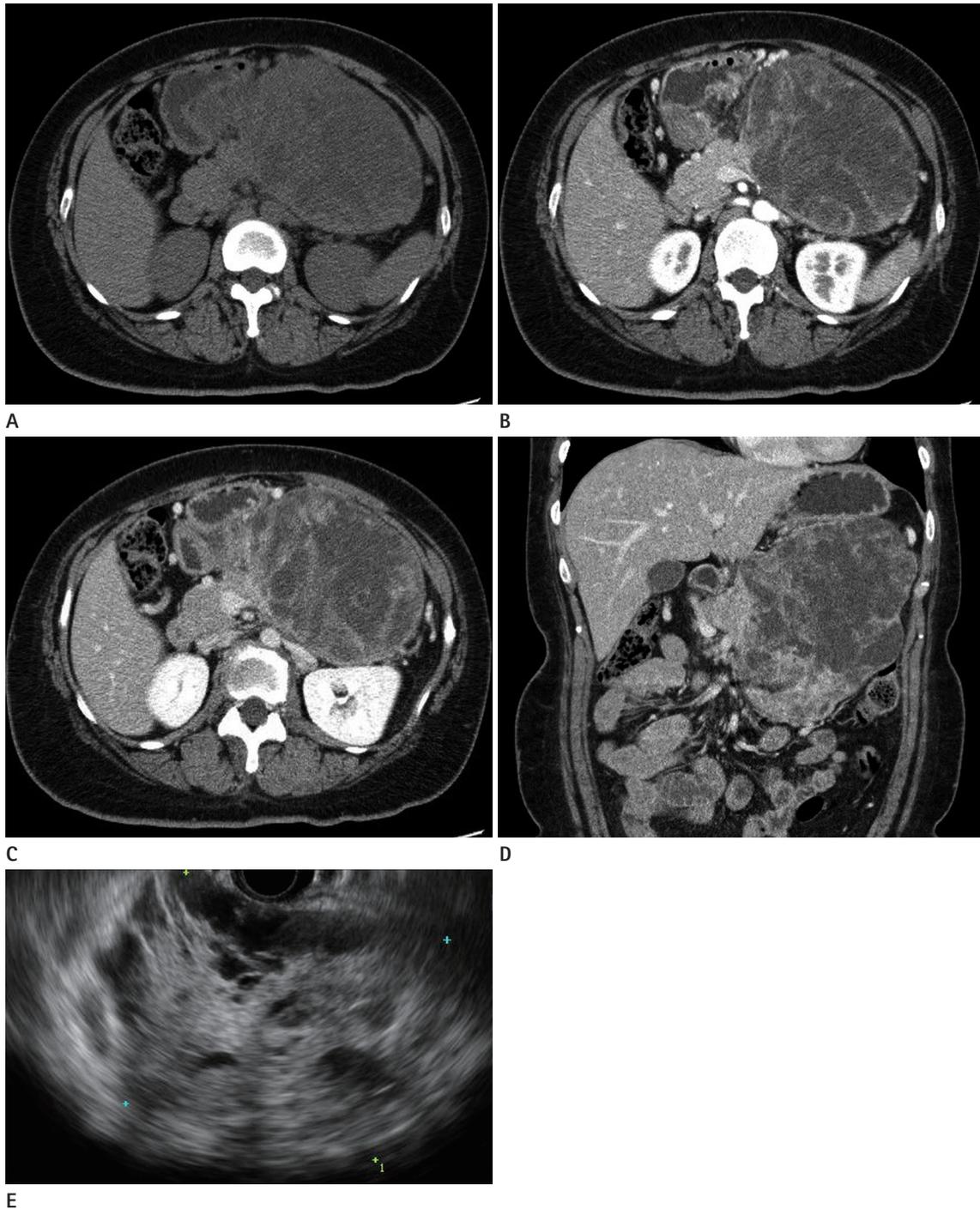
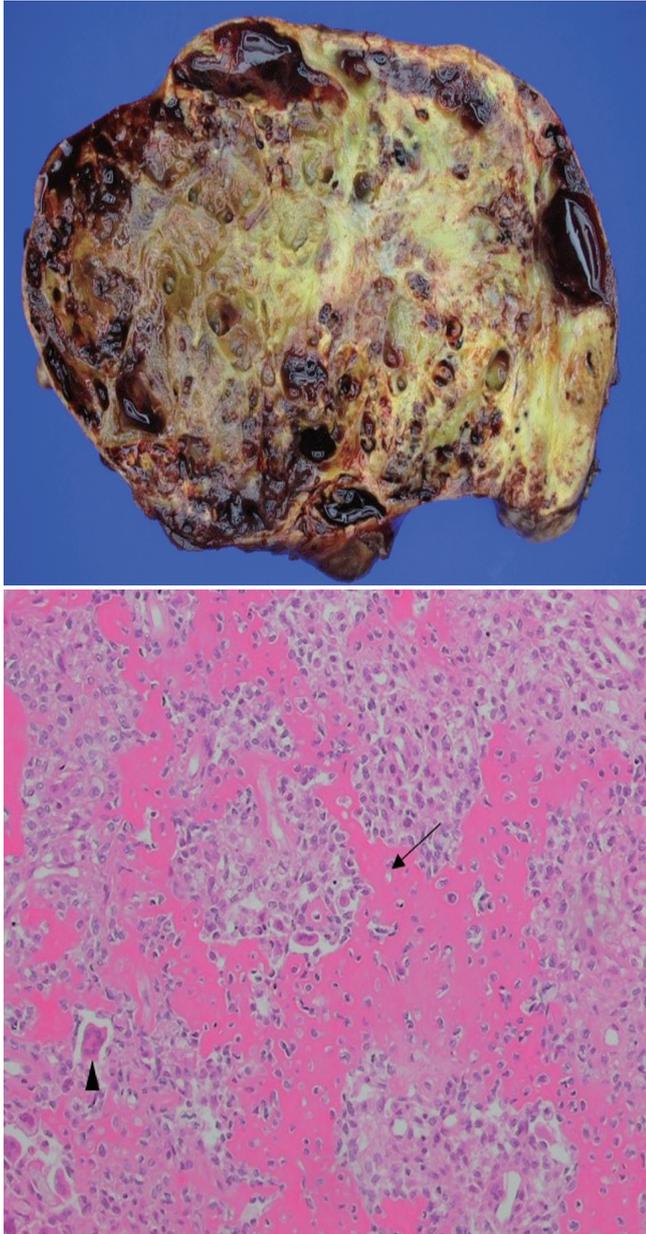


Fig. 1. Retroperitoneal extraskelatal osteosarcom without calcification mimicking pancreas tumor in a 44-year-old woman.
A–D. Abdominopelvic computed tomography with contrast enhancement.
A. No gross radiopaque calcification in the mass is apparent in the pre-contrast image.
B, C, D. On postcontrast scan, the mass is located in the left retroperitoneal space and obscures the pancreas body and tail. Persistent heterogeneous enhancement is apparent, especially in the peripheral portion. The central portion of the mass is non-enhancing and low density.
E. Endoscopic ultrasonography revealing a mass > 7 cm in size, with a heterogeneous echotexture composed of solid and cystic portions. The pancreas body to tail portion is not visible.

with lobulated contours in left side of the retroperitoneum (Fig. 1). There was no definite radiopaque lesion suggesting calcification or fat attenuation within the mass on the precontrast scan (Fig. 1A). On the arterial phase scan, the mass abutted the body of the pancreas and obscured the pancreas body and tail. Obliteration of the splenic vein was also noted on the portal ve-



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Fig. 1. Retroperitoneal extraskeletal osteosarcoma without calcification mimicking pancreas tumor in a 44-year-old woman.
F. Gross cross-section specimen (upper) revealing a cystic and solid tumor. On microscopy (lower, hematoxylin and eosin, $\times 100$), infiltration of the neoplastic spindle or plump cells with neoplastic bone formation is apparent (arrow), accompanied by non-neoplastic osteoclast-type giant cells (arrowhead).

nous phase (Fig. 1C, D). The enhancement pattern of the mass was heterogeneous on the arterial and portal phases (Fig. 1B, C). The peripheral portion of the mass exhibited persistent enhancement, while the central portion exhibited a low-density lesion without contrast enhancement, suggesting cystic or necrotic change(s) (Fig. 1C, D). It was assumed that the origin of the mass was in the pancreas and made impression with solid and cystic masses of the pancreas, such as acinar cell carcinoma and solid pseudopapillary neoplasm. On endoscopic ultrasonography, the echotexture of the mass was heterogeneous and mixed with solid and cystic components (Fig. 1E). Endoscopic ultrasonography also suggested that the mass did not originate from the stomach, similar to a gastrointestinal stromal tumor, because layers of the gastric wall were maintained.

Surgical resection was the chosen treatment option. The mass adhered to the stomach and appeared to originate from the pancreas. Grossly, the tumor was a cystic and solid mass with lobulated contours (Fig. 1F). Engorged vessels in the tumor were noted. After dissection of the adhesion between the mass, stomach, and celiac trunk, the mass was resected with the distal pancreas. Microscopic examination revealed deposition of a network of irregular, eosinophilic, glassy osteoids with interspersed cellular stroma composed of spindle or plump cells (Fig. 1F). The pathological diagnosis was extraskeletal osteosarcoma. Positron emission tomography CT performed before surgery revealed no other hot uptake lesions except for the retroperitoneal mass. It was therefore concluded that the retroperitoneal extraskeletal osteosarcoma was the primary lesion.

DISCUSSION

Extraskeletal osteosarcoma is rare malignant mesenchymal tumor consisting of osteoid tumor cells without bony involvement (1, 2). It accounts for approximately 4% of all osteosarcomas and 1% of all soft tissue sarcomas (1, 2). Primary osteosarcoma of the bone occurs mostly in the first decade of life; however, extraskeletal osteosarcomas occur in adults 50 to 70 years of age (1, 3). The most common sites of tumor occurrence are the lower extremities (47%), upper extremities (20%), retroperitoneum (17%), and trunk/other (10%) (1-4); men are more frequently affected than women (1). Although elevation of serum alkaline phosphatase levels is a common finding in osteosarcomas, in-

cluding retroperitoneal extraskeletal osteosarcomas, in other conditions also exhibiting retroperitoneal mass(es), it is not prone to elevation (3). Pathogenesis is unclear; however, a few risk factors have been reported including previous radiation exposure or trauma. Other studies have proposed that myositis ossificans may be a precursor of extraskeletal osteosarcoma (2, 5). Retroperitoneal extraskeletal osteosarcoma appears as various amorphous shapes or intense calcifications (4). On CT scan, matrix ossification or calcification could be present in approximately one-half of primary tumors; moreover, calcification may represent tumor progression (1). Extraskeletal osteosarcoma has been reported to exhibit a pseudocapsule and no contact with adjacent bony structures on CT (1). CT reveals a soft tissue mass often displacing fat planes, with variable degrees of matrix mineralization. Nonmineralized soft tissue exhibits attenuation consistent with that of muscle on CT. On post-contrast CT, extraskeletal osteosarcomas are usually heterogeneously enhanced depending on the degree of necrosis and hemorrhage. The tumors can directly invade adjacent structures (1-5).

The radiological differential diagnosis of retroperitoneal extraskeletal osteosarcoma mimicking pancreatic tumor includes pancreatic solid and cystic masses, such as solid pseudopapillary tumor, invasive intraductal papillary mucinous neoplasm, and acinar cell carcinoma (6, 7). Solid pseudopapillary tumors have smooth margins, with various levels of solid and cystic components, and occurs mostly in young women (6, 7). Invasive intraductal papillary mucinous neoplasm, especially the branched duct type, also appears as a solid and cystic mass but does not exhibit calcification and, on magnetic resonance cholangiopancreatography reveals communication with the pancreatic duct (6). Acinar cell carcinoma is a rare epithelial pancreatic tumor that is typically well-defined and homogeneously hypovascular; however, when it is large, it may exhibit various levels of cystic portions due to necrosis (7). Nevertheless, it is difficult to predict the specific pathological type of the retroperitoneal masses on imaging studies (8). On the other hand, as reported, elevation of serum alkaline phosphatase level could be helpful in the differential diagnosis of extraskeletal osteosarcoma (3).

The treatment of extraskeletal osteosarcoma is variable, including radical excision only, adjuvant or neoadjuvant chemotherapy with excision, and radiotherapy (9). The prognosis for

extraskeletal osteosarcoma is not favorable, with a reported 5-year survival rate of approximately 60% due to metastasis and recurrence (approximately 50%) (9). The most important factors in the prognosis of extraskeletal osteosarcoma are the size and location of the tumor. Smaller tumor sizes (< 5 cm) and more superficial locations, such as the extremities or trunk, rather than the retroperitoneal or intra-abdominal space, make complete resection possible (9).

We reported a case of extraskeletal osteosarcoma in the retroperitoneal space without calcification mimicking a pancreatic tumor on CT. Because of its varying morphology on imaging studies, it is difficult to predict a diagnosis from imaging before pathological confirmation. Additionally, tumor invasion of the pancreas was a confusing factor in determining the origin of the mass. We suggest that if a large retroperitoneal heterogeneous mass is encountered, even without calcification on CT, in addition to careful review of clinical characteristic, such as elevation of serum alkaline phosphatase levels, retroperitoneal extraskeletal osteosarcoma should be considered in the differential diagnosis.

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췌장종양으로 오인된 육안적 석회화를 동반하지 않은 후복막강 골외성 골육종: CT 소견에 대한 증례 보고

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후복막강에서 발생한 골외성 골육종은 매우 드문 종양이다. 전형적인 골외성 골육종의 영상의학적 소견은 연부조직 음영의 종괴에 동반된 병변 내부의 석회화로 알려져 있다. 우리는 44세 여성에서 췌장 종양으로 오인된 석회화를 동반하지 않은 큰 후복막강 골외성 골육종을 보고하고 전산화단층촬영에서의 영상의학적 소견을 이전 문헌과 고찰하고자 한다. 영상 의학과 의사에게 있어서 후복막강에서 발생한 종양의 진단은 쉽지 않은 일이다. 하지만 후복막강에 큰 종괴가 있을 때 종괴 내부에 육안적으로 확인되는 석회화가 없더라도 골외성 골육종 역시 감별 진단으로서 고려되어야 할 필요가 있으며, 특히 환자의 혈청 알카라인 포스파타제 수치의 증가와 같은 임상 양상이 도움이 될 수 있다.

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