Extraskeletal osteosarcoma is a rare neoplasm that accounts for 2-4% of all osteosarcomas and 1-2% of all soft tissue sarcomas. The most common sites of involvement are the lower extremities (47%), upper extremities (20%), retroperitoneum (17%) and otherwise trunk (10%) [1-6]. We describe a case of primary retroperitoneal extraskeletal osteosarcoma, and discuss the radiological differential diagnosis. To the best of our knowledge, both the computed tomography (CT) and magnetic resonance (MR) imaging of a primary retroperitoneal extraskeletal osteosarcoma has only been reported in few literatures [1, 6].

**CASE REPORT**

A 53-year-old man presented with a gradually enlarging palpable mass in the right lower abdomen for more than six years. He had no other discomfort or any associated symptoms such as nausea, vomiting, abdominal pain, change of bowel habit, or body weight loss. He had a history of gastric ulcer and received hernioplasty for left inguinal hernia 15 years ago.

Results of laboratory examinations were unremarkable, except slightly elevated serum level of alkaline phosphatase.
phosphatase, which was 154 U/L (normal range, 10-100 U/L). Abdominal radiography revealed a densely ossified mass centered in the low abdomen (Fig. 1). Abdominal CT images revealed a large, well-demarcated mass with dense ossifications situated in the right lower abdomen below the level of aortic bifurcation (Fig. 2). The mass was in close contact posteriorly with the right psoas muscle, right iliac vessels, right ureter, and anterior margin of the lumbosacral junction and anteriorly abutted against the abdominal wall. The mass was heavily ossified in the center and spared in the periphery. On MR images, the central part of the mass was predominantly low signal intensity on both T1- and T2-weighted images, consistent with ossification. The peripheral zone of the mass showed isointense on T1-weighted image and hyperintense on T2-weighted image, and was composed of solid and multiple interspersed cystic components. The solid component enhanced avidly after administration of contrast material (Fig. 3).

Surgical resection was performed and confirmed retroperitoneal origin of the tumor without attachment to the bone. Grossly, the tumor was hard in consistency. Microscopic examination revealed deposition of a network of irregular, eosinophilic, glassy osteoid with interspersed cellular stroma composed of spindle cells with pleomorphic nuclei (Fig. 4). The pathological diagnosis was extraskeletal osteosarcoma.

**DISCUSSION**

Extraskeletal osteosarcomas typically affect adults in the 5th to 7th decades of life, in contrast to osteosarcomas of bone that affect younger individuals. Men are two times more frequently affected than women [1, 5]. Most patients present with a gradually enlarging firm mass with or without pain [1, 3, 4]. The elevation of serum alkaline phosphatase level is a common finding in osteosarcomas. Elevated serum alkaline phosphatase levels have also been reported in retroperitoneal extraskeletal osteosarcomas [6].

Retroperitoneal extraskeletal osteosarcoma has a variable amount of mineralization, with intense and amorphous ossifications [3, 7-9]. The nonmineralized soft tissue has attenuation consistent with that of muscle on CT scans and nonspecific intermediate signal intensity on T1-weighted MR images, which is similar to our case. After administration of contrast material, osteosarcomas are commonly hypervascular; however, hypovascular tumors have also been reported [10]. Hypovascularity of the malignant osteoid can explain the faint enhancement of the tumor after administration of contrast material [6].

The differential diagnoses for a retroperitoneal mass with ossification or calcification include both benign and malignant entities. The benign nontumoral conditions are myositis ossificans, foreign body granuloma, and post-traumatic calcified hematoma, and the patients usually have a history of trauma or surgery. Myositis ossificans has a characteristic radiologic finding of gradual ossification of the lesion from the periphery toward the center (zoning phenomenon) [6, 11, 12]. The ossification pattern of extraskeletal osteosarcoma has the densest ossification in the center of the lesion and the least ossification at the periphery (reverse zoning phenomenon) [11]. The benign retroperitoneal neoplasms that may calcify are mature teratoma, hemangioma, ganglioneuroma, schwannoma,
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and paraganglioma. Mature teratoma typically has a heterogeneous internal structure containing fat, fluid, and calcification. Hemangioma is hyperintense on T2-weighted images and may contain phleboliths [6]. Tumors of neural origin are commonly heterogeneous and hyperintense on T2-weighted images [1, 6]. The malignant retroperitoneal tumors that may calcify or ossify include undifferentiated pleomorphic sarcoma (formerly termed malignant fibrous histiocytoma), dedifferentiated liposarcoma, malignant mesenchymoma, extraskeletal mesenchymal chondrosarcoma, and extraskeletal osteosarcoma. Undifferentiated pleomorphic sarcoma is typically a large, heterogeneous, deep-seated tumor with progressive or rapid enlargement, and contains fibrous areas admixed with zones of necrosis, hemorrhage or myxoid change [6]. Dedifferentiated liposarcoma is usually a large painless mass with coexisting both fatty and non-fatty solid components, and can have bulky calcified area [6]. Malignant mesenchymoma is a sarcoma that exhibits two or more lines of specialized differentiation, and can have prominent calcification/ossification or fatty component [13, 14]. Extraskeletal mesenchymal chondrosarcoma is characterized by a biomorphic pattern composed of highly undifferentiated small round cells and islands of well differentiated hyaline cartilage, and shows chondroid-type calcifications and foci of low signal intensity within enhancing lobules [15].

**Figure 3.** Axial T1-weighted (a) and axial T2-weighted fat-suppressed (b) MR images showed that central part (arrow) of the mass was predominantly hypointense with radiating pattern on both T1- and T2-weighted images, consistent with ossification. The peripheral area was composed of solid and cystic components. The solid component (asterisk) was isointense on T1-weighted image and hyperintense on T2-weighted image. Axial (c) and sagittal (d) T1-weighted fat-suppressed MR images after administration of contrast material showed avid contrast enhancement of the solid part (asterisk) of the periphery of the tumor.
In summary, extraskeletal osteosarcoma is a rare soft tissue sarcoma with distinct presentation from osteogenic osteosarcoma. To date, MR imaging of primary retroperitoneal extraskeletal osteosarcoma has only been reported in few literatures [1, 6]. We describe both CT and MR imaging of a densely ossified retroperitoneal mass which was proved to be primary extraskeletal osteosarcoma by surgical resection. If an adult patient has a large densely calcified retroperitoneal mass associated with elevated serum alkaline phosphatase level, extraskeletal osteosarcoma should be considered in one of the differential diagnosis.

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