CASE REPORT

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Primary adrenal leiomyosarcoma with inferior vena cava thrombosis

Abstract We report a primary adrenal leiomyosarcoma in a 59-year-old man. Computed tomography demonstrated a poorly enhanced mass, measuring 10 cm, between the left kidney and the normal left adrenal gland, with tumor thrombus in the inferior vena cava (IVC). The patient underwent left radical nephroadrenalectomy with IVC thrombectomy. The histological diagnosis was adrenal leiomyosarcoma. Adrenal leiomyosarcomas are extremely rare. Only seven cases have been reported previously in the English-language literature.

Key words Adrenal gland · Leiomyosarcoma · IVC thrombus

Introduction

Leiomyosarcoma is a rare soft-tissue tumor and may arise from any structure or organ containing smooth muscle. It usually arises from the myometrium, respiratory tract, and retroperitoneal structures. We report a primary adrenal leiomyosarcoma in a 59-year-old man. Adrenal leiomyosarcomas are extremely rare; only seven cases have been reported previously in the English-language literature. In this report, we present an additional case and review the cases reported in the literature.

Case report

The patient, a 59-year-old man, first came to Tokyo Kosei Nenkin Hospital on July 23, 2001, complaining of left low abdominal and back pain. Ultrasound scan of the abdomen revealed a large mass superior to the left kidney. Results of routine laboratory studies were normal, including full blood count and urea and electrolytes. All the hormonal data were within normal limits.

Computed tomography (CT) demonstrated a poorly enhanced mass, measuring 10 × 9 × 5 cm, between the left kidney and the normal left adrenal gland. The filling defect extended into the inferior vena cava (IVC), which suggested a tumor thrombus (Fig. 1). To rule out adrenal pheochromocytoma with tumor thrombus, a metaiodobenzylguanidine (MIBG) scintigram was performed; the results were negative. Angiographically, the mass was demonstrated to be hypovascular. Arteriographically, irregular tumor vessels ran from the renal artery through the mass lesion. IVC venography showed tumor thrombus extending below the diaphragm. Preoperative lung CT showed no evidence of lung metastasis. A bone scintigram was not performed, but there was no radiological evidence of any skeletal abnormality with regard to chest or abdomen.

Based on a clinical diagnosis of nonfunctional adrenal tumor, the patient underwent left radical nephroadrenalectomy with IVC thrombectomy, on August 23, 2001. The histological diagnosis was adrenal leiomyosarcoma with massive vena cava and renal vein tumor thrombosis.

The postoperative course was unfortunate. A month after the surgery, the patient had developed a dull pain in the neck. Neurological examination revealed muscle weakness and sensory disturbance of the upper limb. At that time, multiple bone metastases, including at the cervical spine (C6), as well as liver metastasis, had been presented. To palliate the bone pain and upper limb paresis, the patient was treated with 33 Gy of external beam radiation to the cervical spine, but his paresis rapidly progressed and the pain was not reduced at all. Two months after the surgery, a tumor thrombus was again present in the IVC. The cervi-
Cal metastasis showed rapid progress and the patient showed complete tetraparesis 4 months after the surgery. The patient died of the disease 6 months after surgery.

Pathology

The surgical specimen weighed 590g along with the resected kidney and perinephric fat. The tumor measured $10 \times 6.5 \times 5.5$ cm and had a firm consistency and grayish-white appearance, and there were areas of hemorrhage and necrosis. Identifiable bright yellow adrenal cortical tissue was noted stretched over the mass (Fig. 2). The tumor showed tight adhesion to the adrenal gland.

Histologically, the tumor consisted of spindle-shaped cells with a fibrillary appearance. Some of the tumor cells showed nuclear pleomorphism and several mitotic figures (Fig. 3). The tumor cells were immunoreactive for smooth muscle actin (SMA; Fig. 4), as well as desmin and vimentin, showing that the tumor was derived from smooth muscle cells. Immunostainings for neuron-specific enolase (NSE), S-100, chromogranin, synaptophysin, and CD34 were negative. The histological diagnosis was leiomyosarcoma.

Microscopically, the tumor cells showed tight attachment to the normal adrenal tissue. Staining for elastic van Gieson (EVG), which stains elastic fibers of vessels, showed the renal vein tumor thrombus to be involved with the renal vein wall. Although the exact origin could not be determined, the tumor, presumably, originated from the adrenal vein.