CLINICAL IMAGE

Giant retroperitoneal undifferentiated pleomorphic sarcoma

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A 70-year-old man was referred to an oncology clinic for a consultation due to a suspicion of a left kidney tumor. He occasionally experienced back pain that he attributed to having fallen off a ladder several months before. A computed tomography (CT) scan showed an irregular retroperitoneal tumor measuring 24 × 18.2 × 25 cm (anteroposterior × transverse × craniocaudal). The tumor was well-demarcated, contained multiple cystic areas, had poor contrast enhancement, and was covered by thickened peritoneum and fluid. It displaced the descending colon anteriorly; the aorta, vena cava, and mesentery to the right; and the left kidney, pancreas, spleen, stomach, and the left dome of the diaphragm upwards, causing a sliding hiatal hernia containing the cardia (FIGURE 1A–1D). Core needle biopsy results indicated malignant sarcoma.

The patient was referred for radical tumor resection with en bloc partial left colectomy and nephrectomy. The postoperative pathological examination indicated an undifferentiated pleomorphic sarcoma: T4N0M0, G2 (according to Unicancer: total score, 5; differentiation, 3; mitotic rate, 1; necrosis, 1), and R0 (microscopically margin-negative resection). The tumor had 2 distinct patterns. The dominant one was a pleomorphic pattern with diffused atypical mitotic figures and multiple giant polynuclear cells with a mixed inflammatory infiltration and lymphatic nodule formation (FIGURE 1E). The remaining part had a spindle-cell pattern with focal high atypia without inflammation (FIGURE 1F); it tested positive on CD34, Bcl-2, and CD10 staining (FIGURE 1G and 1H).

The patient received adjuvant chemotherapy with ifosfamide. One year after the surgery, a follow-up positron emission tomography-CT scan showed no signs of tumor recurrence or dissemination.

Soft tissue sarcomas are heterogeneous and relatively uncommon neoplasms, typically more aggressive than carcinomas, yet they very rarely metastasize to the lymph nodes. Sarcomas of the retroperitoneal space constitute 15% of all cases and are associated with a worse prognosis than those occurring in more common locations involving the extremities. They are commonly diagnosed in an advanced stage due to the nonspecific initial symptoms and the large capacity of the retroperitoneum and abdomen allowing the tumor to grow asymptomatically for a long time.[1-3]

Initial management includes staging using imaging techniques and core needle or open biopsy. CT is the recommended modality, except for the lesser pelvis, where magnetic resonance imaging may provide more details. The differential diagnosis should include malignant neoplasms of the kidney, pancreas, and adrenal gland, as well as germ cell tumors, lymphomas, and metastases of testicular neoplasms.[1-5]

Radical tumor resection remains the recommended treatment option. The tumor is usually not contained within the fascia and may easily infiltrate the surrounding structures. This often requires multiorgan resection, including the kidney and/or adrenal gland (50%), large bowel (20%), distal pancreas (15%), and/or spleen (10%). Sometimes, resection of organs not macroscopically involved should be considered, as the microscopic infiltration may be as high as 50%. Nonradical resection (R1/2), high histologic grades (G2–3), and aggressive tumor subtypes (other than well-differentiated liposarcoma) are associated with worse prognosis. Distant metastases are rare; however, there is a tendency for local recurrence. Indications for radiation therapy and/or adjuvant chemotherapy should be discussed individually within a multidisciplinary team of specialists.[1-3]

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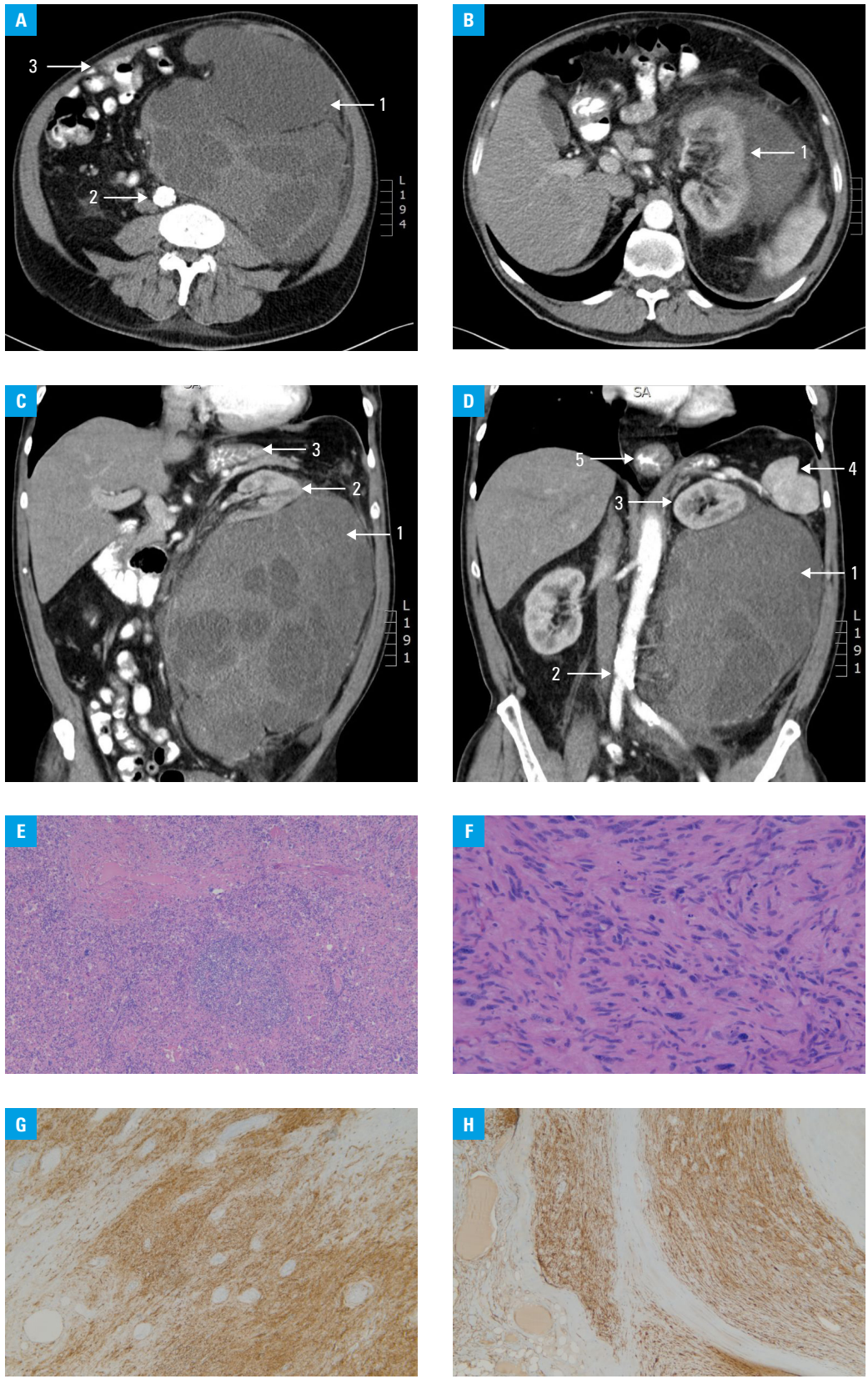

FIGURE 1 Macroscopic and microscopic images of a retroperitoneal sarcoma; **A**, **B** – computed tomography (CT) imaging (arterial phase, transverse view) showing the tumor (1) displacing the aorta (2) and intestines (3) to the right side of the abdomen (**A**), and dislocating the left kidney (1) to the midline and upwards, moving it to an unusual transverse position (**B**); **C**, **D** – CT imaging (arterial phase, coronal view) showing the tumor (1) displacing the left kidney (2) and stomach (3) upwards and towards the midline (**C**), and the tumor (1) displacing the aorta (2), left kidney (3), spleen (4), and stomach, causing a hiatal hernia (5) (**D**); **E**, **F** – histopathological analysis showing a pleomorphic pattern with inflammatory infiltration and poorly-formed lymphatic nodules in the center (**E**; hematoxylin and eosin staining, magnification × 40), and a spindle-cell pattern (**F**; hematoxylin and eosin staining, magnification × 200); **G**, **H** – histopathological analysis showing a positive reaction in the spindle-cell portion of the tumor (**G** – immunohistochemical [CD34] staining, magnification × 100; **H** – immunohistochemical [Bcl-2] staining, magnification × 40)

ARTICLE INFORMATION

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