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Answers to Medical Quiz

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A1.

	Benign Types	Malignant Types
Mesodermal Tumors	Lipoma Leiomyoma Rhabdomyoma Myxoma Fibroma Lymphangioma Hemangioma	Liposarcoma Leiomyosarcoma Rhabdomyosarcoma Myosarcoma Fibrosarcoma Malignant Mesenchymoma
Neuorgenous Tumors	Ganglioneuroma Pheochromocytoma Neurofibroma	Neuroblastoma (below age 6) Malignant Schwannoma
Remnant Tumors	Teratoma (in children) epithelial cysts adenoma	Teratocarcinoma Malignant chordoma Carcinoma
Others	Urinoma Abscess	Lymphoma Extraosseous Ewing's sarcoma Renal tumor

A2. MRI along and magnetic resonance angiography can be done to further delineate the mass and assess its origin and vascularity.

A3. She needs biopsy to determine the histological nature of the mass, which will help in planning the initial management, whether it will be chemoradiotherapy to downstage the tumor followed by surgical resection or surgery with further adjunctive therapies for metastasis. However, she will have to be staged exactly if the mass proves to be a tumor.

DISCUSSION

This patient had a positive biopsy for extraosseous Ewing's sarcoma with invasion

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into the left kidney. She underwent surgical resection, which was mainly a debulking and palliative operation since the tumor was extensive in the retroperitioneal area and pulmonary metastasis was documented.

Ewing's family of tumors (EFTs) include: 1.Ewing's tumor of bone (ETB or Ewing's sarcoma of bone), 2. extraosseus Ewing's (EOE), 3. primitive neuroectodermal tumors (PNET or peripheral primitive neuroepithelioma), and 4. Askin's tumor (PNET of the chest wall)

Studies using immuno-histochemical markers, cytogenetics, molecular genetics, and tissue culture indicate that these tumors are all derived from the same primordial stem cell.

EFTs occur most frequently in the second decade of life and account for 4% of childhood and adolescent malignancies. The incidence in boys is slightly higher than girls (ratio of 1.1:1). ETB is estimated to be 60% of the EFTs. The most common sites of EOE are: trunk (32%), extremity (26%), head and neck (18%), retroperitoneum (16%), and all other sites (8%).

Major prognostic factors include the site, tumor volume, and the presence of metastases.
Extra-skeletal Ewing sarcoma (EES) is a rare soft tissue tumor that is morphologically indistinguishable from the more common Ewing sarcoma of bone. It must be distinguished from other small, blue round cell tumors (SBRCTs). The most frequent sites of occurrence are the chest wall, lower extremities, and paravertebral region. Less frequently, the tumor occurs in the pelvis and hip region, the retroperitoneum, and the upper extremities. It is usually found in young adults (younger than 30 years) and has a slight predominance in male patients.

Diagnostic scans (chest x-ray, chest CT, magnetic resonance imaging (MRI), or CT of the primary) should be performed prior to any procedure that requires anesthesia because anesthesia-induced abnormalities in the chest may be difficult to assess. Other studies such as bone scan and bone marrow biopsy may be obtained pre-operatively if the index of suspicion is high.

The successful treatment of patients with tumors of the Ewing's family (EFTs) requires the use of multi-drug chemotherapy, in addition to radiation therapy and surgical therapy to the primary tumor. Many patients with metastatic disease at diagnosis respond well to the therapy; however, in most cases the disease is only partially controlled or it recurs after complete remission. Patients with only lung metastases have a better event-free survival than those with metastases to bone and/or bone marrow. Usually, they have an unfavorable prognosis with a survival rate of less than fifty percent

REFERENCE

1. Otto H. Wegener. Whole Body Computed Tomography. 2nd edn.1993:462-78.
Wrong and not mentioned in the text