Non-rhabdomyosarcoma soft tissue sarcoma - NRSTS

Clinical History:

A 7-year-old girl presented with a large abdominal mass. The mass had been increasing in size over the past month and was associated with abdominal discomfort and weight loss.

Imaging Findings:

Ultrasound of the abdomen showed a large, ill-defined, mixed solid and cystic vascular mass lesion (Fig. 1a-c). On MR imaging, the lesion was demonstrated to have mass effect on adjacent organs. STIR sequence did not show any fat suppression, suggesting that the contents were fluid (Fig. 2a). Coronal (Fig. 2a) and axial (Fig. 2b) T2w images confirmed the mass to be predominantly cystic. The mass was seen displacing adjacent bowel loops and the mesenteric vessels. The stomach was displaced anteriorly, the spleen superiorly and the pancreas to the right side. The location of the mass was presumed in the retroperitoneum. It was seen in close proximity to the left renal artery. The left renal vein was not identified and was possibly compressed or encased by the mass. Severe left hydronephrosis was noted secondary to mass effect on the proximal left ureter (Fig. 2f).

Discussion:

Nonrhabdomyosarcoma soft tissue sarcomas (NRSTs) are a heterogeneous group of mesenchymal cell neoplasms that account for about 4% of childhood cancers [1, 4]. Soft tissue sarcomas are rare paediatric tumours classified by the WHO according to histologic cell types [2]. 40% are rhabdomyosarcomas and 60% are NRSTS [2]. Paediatric NRSTs behave differently than the adult varieties, hence causing this dilemma [1].

The pathogenesis is due to the lack of tumour suppressing genes; p53 and retinoblastoma suppressing gene, resulting in limitless replication, invasion and metastatic growth [1]. Four categories of biological potential are included: benign, intermediate–locally aggressive, intermediate–rarely metastasizing, and malignant [1].

The types of soft tissue sarcoma based on histology include [2]: Alveolar soft part sarcoma, angiosarcoma, clear cell sarcoma, desmoid tumour, desmoplastic small round cell tumour, epithelioid sarcoma, extraskeletal chondrosarcoma, extraskeletal osteosarcoma, fibrosarcoma, haemangiopericytoma, leiomyosarcoma, liposarcoma, malignant fibrous histiocytoma, malignant schwannoma and synovial sarcoma.

Ultrasound is the first radiological investigation in the work-up of paediatric abdominal masses. It is inexpensive,
readily available, does not involve radiation and seldom requires sedation. Ultrasound identifies the organ of origin as well as the type of tissue components present (solid versus cystic). Magnetic resonance imaging provides excellent soft tissue definition, and is therefore preferable for surgical planning and following neoadjuvant therapy [3, 4, 5]. Staging is best done with CT chest/abdomen/pelvis. PET may be also used, however, it is an optional pretreatment assessment investigation only [6]. Imaging usually shows a soft tissue enhancing mass which may be partly cystic or solid. Histology of the abdominal mass in this patient shows cores of tissue composed of loose interfacing fascicles of plump spindle cells with vesicular nuclei and small nucleoli, surrounded by abundant loose fibrillar and myxoid stroma. No cytological atypia or necrosis is seen. Immunohistochemistry shows strong positivity for vimentin and beta-catenin, as well as patchy positivity for SMA and desmin. S100 and CD56 are negative. Differential diagnosis includes haemangioendothelioma, a liver tumour which may be mistaken for an abdominal mass. Lymphoma may also present as a large abdominal mass encasing the arteries.

Treatment options are based on whether the cancer has spread, the amount of tumour left after surgery, and whether the child has reached full growth. Surgery, radiation, and chemotherapy are all treatment options, either alone or in combination with one another [2, 3, 4]. This patient was put on weekly Methotrexate plus vinblastine combination. No surgery was considered at the moment.

**Differential Diagnosis List:** Retroperitoneal non-rhabdomyosarcoma soft tissue sarcoma (NRSTS), Desmoid tumour, Liposarcoma, Neuroblastoma, Haemangioendothelioma

**Final Diagnosis:** Retroperitoneal non-rhabdomyosarcoma soft tissue sarcoma (NRSTS)

**References:**

**Figure 1**

**a**

*Description:* An ill-defined large solid and cystic lesion is noted in the midline of the abdomen on ultrasound. *Origin:* BHRUT NHS Trust, Rom Valley Way, UK

**b**

*Description:* An ill-defined large solid and cystic lesion is noted in the midline of the abdomen on ultrasound. *Origin:* BHRUT NHS Trust, Rom Valley Way, UK
**Description:** Ultrasound colour Doppler study shows vascularity in the solid component of the lesion.

**Origin:** BHRUT NHS Trust, Rom Valley Way, UK
Figure 2

Description: Coronal STIR image shows fluid signal intensity lesion. Origin: BHRUT NHS Trust, Rom Valley Way, UK.
Description: Coronal T2 image shows the large predominantly cystic and solid mass with adjacent mass effect. Origin: BHRUT NHS Trust, Rom Valley Way, UK.
Description: Axial image shows the large retroperitoneal cystic/solid mass. Severe left hydronephrosis is noted. **Origin:** BHRUT NHS Trust, Rom Valley Way, UK.

Description: Axial image shows the large contrast enhancing retroperitoneal mass. **Origin:** BHRUT NHS Trust, Rom Valley Way, UK.
Description: Axial image shows the large cystic solid mass. Origin: BHRUT NHS Trust, Rom Valley Way, UK.
Description: Coronal image shows severe left hydronephrosis. Origin: BHRUT NHS Trust, Rom Valley Way, UK.