

288/20 740913

~~288/20 740913~~
42 YEAR OLD FEMALE. CORE BIOPSY FROM RIGHT RETROPERITONEAL MASS, ? SARCOMA. CT: POORLY ENHANCING 10 X 7.3 X 3.3CM SOFT TISSUE MASS IN THE RETROPERITONEUM, BEHIND AND TO THE RIGHT OF THE IVC WITH A TAIL OF THE ABNORMALITY EXTENDING OVER THE MIDLINE TO THE LEFT. FOCAL CALCIFICATIONS ARE SEEN. THE MASS IS IN CONTACT WITH IVC, AND RIGHT RENAL VEIN AND RIGHT RENAL ARTERY. IT REMAINS SEPARATE FROM THE AORTA, INFERIOR MARGIN OF THE SMA, RIGHT ADRENAL GLAND, KIDNEY, LIVER AND DIAPHRAGM. THERE IS NO RETROPERITONEAL OR PELVIC LYMPHADENOPATHY. NO MESENTERIC ABNORMALITY, AND NO MEASURABLE PERITONEAL DISEASE OR ASCITES. PROMINENT CYSTS IN THE RIGHT ADNEXA HAVE DEVELOPED BETWEEN THE CT FROM 8/11/19 AND 26/11/19, AND LIKELY REFLECT PHYSIOLOGIC FOLLICLES. REST OF THE ABDOMINAL VISCERA ARE UNREMARKABLE. NO INTRATHORACIC ABNORMALITY. NO SUSPICIOUS OSSEOUS LESION. RADIOLOGIC OPINION: POORLY ENHANCING ABNORMALITY IN THE RETROPERITONEUM OF UNCERTAIN SIGNIFICANCE, BUT CALCIFICATION AND POOR ENHANCEMENT SUGGEST A BENIGN ENTITY. FOLLOW UP IMAGING OR BIOPSY WOULD BE APPROPRIATE. NO PREVIOUS RMH HISTOLOGY

MACROSCOPY

Retroperitoneal mass: 6 cores ranging from 19-22mm. 1-6) AE.

HISTOLOGY

Cores of moderately cellular tumour, composed of patternless arrays or streams of spindle cells with elongated, hyperchromatic tapered nuclei and fibrillary cytoplasm in delicately collagenous stroma. The cells are plump, but no definite atypia is seen. Moderate numbers of essentially mature ganglion-cells, some containing pigment, are interspersed. Focally, (eg slide 3) the tumour is seen to infiltrative mature adipose tissue and in areas there are prominent psammoma bodies or small calcifications (eg slide 2 and 6), with an occasional osteoclast-like giant cells around the calcification (eg slide 4). No tumour necrosis is seen. No definite mitotic figures are seen in 10 hpf. There are small packets of haemorrhage/ degenerate blood, but no definite necrosis is noted.

The tumour is diffusely and strongly positive for S100 protein and CD56, and strongly positive for neurofilament in most cells, and SOX10 shows multifocal nuclear expression. CDK4 shows diffuse moderate expression. It is only focally positive for CD34. There is only very scattered p16, of uncertain significance. There is scanty weak h-caldesmon, although this marker is often aberrantly overexpressed in this laboratory. The tumour is negative for desmin, myogenin, CD117, DOG1, STAT6 and AE1/AE3. SMA is essentially negative with only very scanty weak expression seen. The proliferation fraction by MIB1 is very low (essentially negligible).

The features are in keeping with ganglioneuroma. The presence of psammoma bodies appears unusual, but the features do not support melanotic schwannoma (malignant melanotic nerve sheath tumour).

Dr Magnus Hallin/Dr Khin Thway

T: soft tissue t retroperitoneum m ganglioneuroma