ROYAL MARSDEN NHS FOUNDATION TRUST - HISTOPATHOLOGY REPORT 749217: P. 1879 133 0202

Lab No	6625/20	Reported	9 Jul 2020	Pathologist DR HALLIN/DR THWAY
Source	Internal Operation	Sample Received	2 Jul 2020	CRITICAL CARE UNIT Ward (CHELSEA)
Sex	MALE	Age	73	Branch FULHAM ROAD
Clinical Diagnosis		Operation	1 Jul 2020	Consultant STRAUSS,MR D C

SITE **DIAGNOSIS**

SOFT TISSUE AND OTHER CONNECTIVE TISSUE A (T1X005)

B RETROPERITONEUM (TY4600)

SPINDLE CELL SARCOMA (Malignant) (M88013) SPINDLE CELL SARCOMA (Malignant)

(M88013)

73 YEAR OLD MALE. RECENT REVIEW OF OUTSIDE HISTOLOGY OF INCIDENTALLY DETECTED RETROPERITONEAL MASS SHOWED FEATURES IN KEEPING WITH PART OF SPINDLE CELL SARCOMA THE CELLS HAD A MARKEDLY STORIFORM PATTERN AND SHOWED ONLY MILD TO MODERATE ATYPIA AND WERE DIFFUSELY AND STRONGLY POSITIVE FOR CD34 (6024/20); THIS WAS DIFFICULT TO INTERPRET, AND WHILE DDL WAS CONSIDERED AT THIS INTRAABDOMINAL SITE, THE IMMUNOPROFILE WAS NOT SUPPORTIVE (FISH FOR MDM2 AMPLIFICATION STATUS AWAITED, ALONG WITH FISH FOR ALK GENE REARRANGEMENT). THE POSSIBILITY OF PERINEURIAL MPNST WAS ALSO CONSIDERED. SS18-SSX1/2 FUSION TRANSCRIPTS WERE UNDETECTABLE BY RT-PCR. MATERIAL WAS INSUFFICIENT FOR MOLECULAR INVESTIGATIONS TO ASSESS FOR COL1A/PDGFB FUSION TRANSCRIPTS/ GENE REARRANGEMENTS FOR DERMATOFIBROSARCOMA, ALTHOUGH THIS WOULD BE HIGHLY UNLIKELY INTRA-ABDOMINALLY. THIS SPECIMEN: EXCISION OF LEFT RETROPERITONEAL SARCOMA, WITH SPLEEN, KIDNEY AND COLON.

MACROSCOPY

A. Spleen: an unremarkable spleen weighing 82g and measuring 67x70x32mm. There is attached fatty tissue measuring 35x12x17mm. On slicing the spleen is largely unremarkable with a small focal area of ?scarring towards one aspect. This area measures 12x6x15mm. No other pathology is seen. Blocks 1) Representative section of normal spleen, 2&3) Representative sections of area of scarring, 4) Representative section of spleen with surrounding fatty tissue. Tissue remains. B. Left retroperitoneal sarcoma + kidney + colon: an unorientated retroperitoneal resection specimen measuring 230x224x132mm. There is an attached kidney measuring 121mm (pole to pole) x59x40. There is also an attached portion of colon measuring 310mm in length and 36mm in maximum diameter. Attached to the colon there is a multilobular retroperitoneal tumor mass measuring 173x114x152mm. One surface of this mass is covered in fragments of skeletal muscle, the opposite surface covered in smooth shiny connective tissue. On opening the colon the colonic mucosa appears macroscopically normal with no obvious infiltration of tumor. On opening the kidney the cut surface appears macroscopically normal with no obvious pathology identified. There is some distention of the ureter towards the hilum. The kidney lies 30mm away from the tumor mass. The adrenal gland is not present in the specimen. The tumor abuts the smooth serosal surface, the roughened surface and the colonic mucosa. On slicing the tumor is well circumscribed and heterogeneous with predominantly bland cream smooth areas with a central area of hemorrhage and necrosis. The necrosis equates approximately 25-30% of the tumor volume. Blocks 5) Tide ureteric resection margin. 6) Renal cortex to tumor. 7) Renal hilum adjacent to renal vessel resection margins. 8&9) Colonic resection margins. 10) Colonic mucosa to tumor. 11) Tumor to smooth serosal resection margin. 12) Tumor to roughened resection margin. 13&14) Representative sections of tumor. 15) Representative section of hemorrhagic/ necrotic area, central. Tumor and tissue remain.

HISTOLOGY A1-4. Spleen:

Sections from the areas of scarring described macroscopically show a prominently collagenized/ fibrotic area on the surface of the spleen, merging with the fibrous tissue/ capsule, and focally (slide 3) seen to extend from the surface into the parenchyma, with a well-defined border, without an infiltrative pattern present. This area is sparsely cellular to acellular and markedly fibrotic, with some possible focal areas of calcification. No cellular atypia, ghost cells, tumor necrosis or mitotic figures are seen.

This might represent organization/ scarring to previous pathology (e.g. splenic infarct), although the cause of this cannot be determined in this material. There is surrounding adipose tissue of mature type, without discernible atypia or septation. No other significant abnormality is noted within or around the spleen.

B5-15. Left retroperitoneal sarcoma, with kidney and colon: Sections show fibroadipose tissue containing cellular

tumor, frequently with similar features to those in the previous biopsy (6024/20), and comprising extensively storiform arrangements of moderately atypical spindle cells. In other areas, similar cells are disposed in streams or loose fascicles, and focally there is marked atypia, with enlarged vascular or hyperchromatic nuclei, including multinucleate forms. The mitotic index is up to 6/10hpf, with atypical forms, and there are areas of prominent necrosis (e.g. slide 15). A scattered mild chronic inflammatory infiltrate is intermingled. The tumor is seen to infiltrate the surrounding mature fat, but no definite atypia is noted within this. The tumor is focally close to the large bowel wall, but no infiltration is noted.

The features are consistent with spindle cell sarcoma with predominant prominent storiform architecture (grade 2). FISH for MDM2 amplification status is awaited on the previous biopsy. At this site, this more likely represents a storiform pattern of dedifferentiated liposarcoma, although the weak expression of CDK4 and absence of expression of p16 was noted in the core biopsy (no definite well- differentiated liposarcomatous component is identified in this specimen). Molecular investigations are awaited to conclusively exclude dermatofibrosarcoma, although this is highly unlikely morphologically and at this site. A possibility here is of perineurial malignant peripheral nerve sheath tumor/ malignant perineurioma, although this cannot be proven.

The tumor is present at the inked 'smooth' and 'roughened' resection margins. The section from the tied ureteric resection margin shows unremarkable ureter with surrounding fibroadipose tissue; no tumor, atypia or significant abnormality is noted. There is focally moderate inflammation in the ureteric wall, particularly around vessels, which likely represents changes secondary to the surgical process. The renal cortex shows surrounding mature adipose tissue, with occasional mild septation, but no conclusive tumor/ well-differentiated liposarcoma is seen. No significant pathology is noted in the renal parenchyma. The renal hilum shows unremarkable large vessels; the surrounding adipose tissue appears of mature type, without notable atypia. The sections from the colonic resection margins show focally slightly thinned (slide 9) large bowel mucosa and wall, with no significant pathology. There is focal mild serositis, and some surrounding mature adipose tissue; no tumor is seen.

Dr Magnus Hallin/Dr Khin Thway