ROYAL MARSDEN NHS FOUNDATION TRUST - HISTOPATHOLOGY REPORT NHS Number: 630 255 0882

Lab No

5855/20

Reported

23 Jun 2020

Pathologist DR HALLIN/DR THWAY

Source Sex

Internal Operation Sample Received 10 Jun 2020

MALE

Age

56

Ward Branch

FULHAM ROAD

Clinical Diagnosis

Operation

9 Jun 2020

Consultant STRAUSS, MR D C

SITE

DIAGNOSIS

SOFT TISSUE AND OTHER

NEOPLASM MALIGNANT / SPINDLE CELL SARCOMA (Malignant) / A CONNECTIVE TISSUE (T1X005) RHABDOMYOSARCOMA (Malignant) (M80003 / M88013 / M89003)

NEOPLASM MALIGNANT / SPINDLE CELL SARCOMA (Malignant) /

B FLANK (TY1310)

RHABDOMYOSARCOMA (Malignant) (M80003 / M88013 / M89003)

56 YEAR-OLD MALE, WHO PRESENTS WITH 12-MONTH HISTORY OF ENLARGING LEFT FLANK LESION, WHICH HAS STEADILY GROWN SINCE SEPT 2019, AND RECENTLY DEVELOPED BLEEDING BLISTERS. NO SIGNIFICANT PMH. OUTSIDE CT. SUBCUTANEOUS LESION, ?DFSP, WITH SOME AREAS OF NODULARITY, POTENTIALLY SUSPICIOUS FOR SARCOMATOUS CHANGE. THIS SPECIMEN: WLE OF LEFT FLANK FUNGATING TUMOR, ?DFSP WITH SARCOMATOUS CHANGE. NO PREVIOUS RMH HISTOLOGY

MACROSCOPY

Wide excision left flank fungating tumor; stitch single 12 o'clock, double = 3 o'clock: an unorientated elliptical excision specimen measuring 76mm (12 to 6 o'clock), x180mm (3 to 9 o'clock), x27mm (superficial to deep). The anterior surfaces entirely covered by skin which bears a central raised multinodular area measuring 52mm (3 to 9 o'clock), 33mm (12 to 6 o'clock) to a height of 12mm. There is an area of fungation centrally which measures 9mm in maximum diameter. The specimen has been inked 12 o'clock = blue, 3 o'clock = red, 6 o'clock = green, 9 o'clock = orange and deep = black. Slicing reveals a multifocal cream homogeneous tumor extending 57mm (3 to 9 o'clock), x37mm (12 to 6 o'clock), x18mm (superficial to deep). The tumor lies 14mm (12 o'clock margin), 9mm (6 o'clock margin), 37mm (3 o'clock margin) and 48 (9 o'clock margin). Tumor lies 13mm form closest to deep margin. No obvious macroscopic necrosis is identified. Blocks 1) Cruciate of 3 o'clock margin. 2) Cruciate of 9 o'clock margin. 3) Tumor to closest 6 o'clock margin. 4) Tumor to closest 12 o'clock margin. 5) Tumor to closest deep margin. 6) Representative section of tumor at area of fungation. 7&8) Further representative sections of tumor. Tissue and tumor remain.

HISTOLOGY

Sections show centrally ulcerated skin and subcutis, with skeletal muscle deeply. The dermis and subcutis contain focally polypoid/ exophytic, ill-defined cellular tumor, composed of loose fascicles or ill-defined nests of cells with moderately to focally markedly atypical ovoid or spindled vesicular or hyperchromatic nuclei with fibrillary or moderate amounts of palely eosinophilic cytoplasm. In many areas, particularly the nested foci, the cells have a slightly more ovoid and plasmacytoid morphology. Tumor giant cells are also present. The mitotic index exceeds 20/10hpf, with atypical forms. Focal tumor necrosis is present (eg slide 8). No morphologic epithelial differentiation is noted. No skeletal muscle invasion is identified. The surrounding fibroadipose tissue shows moderate numbers of lymphoid aggregates, which are a mixture of CD20- and CD3- positive small lymphocytes. The overlying squamous epithelium is variably hyperkeratotic with some focal mild atrophy, but no significant pathology is seen within it, and no connection with/ transition from the overlying epidermis is noted.

The tumor is essentially diffusely and strongly positive for desmin, including in the more rounded/ plasmacytoid cells in (block 5), with scanty but focal nuclear myogenin expression (block 6); myogenin is essentially negative in block 5, with some possible cytoplasmic expression in mast cells, without conclusive expression in tumor nuclei (positivity in one or two possible tumor nuclei only). There is some focal SMA expression. In one block (slide 5), AE1/AE3 shows very strong, essentially diffuse expression in the rounded cell/ plasmacytoid component, with multifocal strong expression in the spindle cells. In the other block (block 6), there is focal, predominantly moderate granular positivity for AE1/AE3 at the superficial aspect of the tumor; this does not identify any epithelial elements, and this likely represents aberrant staining. CD138 shows weak to moderate granular expression, interpreted as aberrant staining, in one of the blocks (block 6), and is negative in the other block (block 5). The tumor is negative for CD34. STAT6, S100 protein, SOX10, MUC4, CD45, CD20 and CD3. INI1 and BRG1 are retained in nuclei. The proliferation fraction by MIB1 is high to very high.

This is a high-grade malignant neoplasm with myoid/

rhabdomyosarcomatous differentiation (rhabdomyosarcoma NOS, grade-3) and keratin expression; the latter may be aberrant, but sarcomatoid carcinoma cannot be excluded. The morphology does not support alveolar rhabdomyosarcoma, but molecular investigations are awaited to exclude this. FISH is also awaited to assess for EWSR1 and FUS gene rearrangements, seen in a very small subset of

rhabdomyosarcomas. The tumor is at least 20mm from the 12 o'clock margin, at least 21mm from the 3 o'clock margin, at least 26mm from the 9 o'clock margin, at least 28mm from the 3 o'clock margin, and focally 7.5mm from the deep margin (case also kindly seen by Dr Terlizzo (consultant skin pathologist), who agrees with the findings).

Dr Magnus Hallin/Dr Khin Thway

SUPPLEMENTARY REPORT 23/6/20

The tumor is diffusely and strongly positive for MyoD1.

The interpretation remains as above; this is a high-grade malignant neoplasm with myoid/ rhabdomyosarcomatous differentiation (rhabdomyosarcoma NOS, grade 3) and keratin expression; the latter may be aberrant, and sarcomatoid carcinoma cannot be entirely excluded, but appears less likely overall.

Dr Magnus Hallin/Dr Khin Thway