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Pathology Report 001144078 CHP COOK AUSTIN REED 07/12/06 14:30

Name COOK AUSTIN REED
 MRN 001144078 CHP
 Physician KANE TIMOTHY
 Report Type Pathology Report
 Date of Event 07/12/06 14:30
 Date of Birth 02/23/2001
 Sex M
 Patient Status ... CHQ
 Document # CHS06-5156
 Authored by TEOT LISA
 Account # 0001027827557
 Hosp/Group CHP

PATIENT HISTORY:

Procedure: Left nephrectomy with removal of mass.

Pre-Op/Post-Op: Left renal mass./Same.

The patient is a 5 year-old boy with a ~~left kidney mass and a high grade tumor of the scalp and skull, most consistent with metastatic clear cell sarcoma of kidney.~~

FINAL DIAGNOSIS:

Kidney and adrenal gland, left, radical nephrectomy:

CLEAR CELL SARCOMA OF KIDNEY, 628 GRAMS, 13.2 CM, WITH RENAL SINUS INVOLVEMENT. (SEE NOTE)

ALL SURGICAL RESECTION MARGINS NEGATIVE.

THREE LYMPH NODES NEGATIVE FOR METASTATIC CLEAR CELL SARCOMA.

ACUTE AND CHRONIC TUBULOINTERSTITIAL NEPHRITIS OF UNINVOLVED KIDNEY.

UNREMARKABLE ADRENAL GLAND.

NOTE:

~~This tumor is stage IV based on metastasis to the scalp and skull.~~
 (CHS06-4862, 6/30/2006).

My signature is attestation that I have personally reviewed the submitted material(s) and the final diagnosis reflects that evaluation.

KLD/KLD

Pathologist: Lisa A. Teot, M.D.

Fellow/Chief Resident: Kudakwashe R. Chikwava

** Report Electronically Signed Out **

By Pathologist: Lisa A. Teot, M.D.

7/17/2006 18:12

My signature is attestation that I have personally reviewed the submitted material(s) and the final diagnosis reflects that evaluation.

GROSS DESCRIPTION:

The specimen is received fresh, labeled with the patient's name and designated "left kidney and mass." It consists of a left radical nephrectomy specimen that includes left adrenal. The specimen weighs 628 grams and measures 17.5 x 8.5 x 7.0 cm. It consists of a left kidney measuring 17.0 x 8.5 x 7.0 cm and a left adrenal gland measuring 7.2 x 1.8 x 0.5 cm. The kidney has attached renal artery and vein, each measuring 0.5 cm in length by 0.5 and 0.9 cm in diameter, respectively. The ureter measures 5.2 cm in length and 0.3 cm in diameter. The kidney is covered by a thin, Gerota's

fascia that appears pink, translucent, and bosselated. The specimen is bisected to reveal a 13.2 x 8.5 x 7.8 cm multilobated mass. The mass is located primarily in the upper and mid portions of the kidney, including the renal sinus and pelvis. The cut surface is firm and tan, with focal red-pink areas. ~~There are areas of hemorrhage and necrosis measuring up to 3.5 cm in greatest dimension in the lower portion of the renal mass. The specimen extends grossly into the distorted pelvis.~~ No extension into the renal arteries, renal vein, or ureter is identified. The tumor appears to be confined within the renal capsule/Gerota's fascia. The uninvolved kidney shows well demarcated corticomedullary junction. The cortex is dark red. The medulla is tan-brown. The hilum has moderate amounts of fat. Three lymph nodes, 1.5 cm in greatest dimension, are harvested from the hilar fat. Their cut surface is tan and uniform. The attached adrenal gland is grossly unremarkable. Digital images are taken and saved in CoPath. Fresh tissue is sent for cytogenetic studies and snap-frozen for potential molecular studies. Sections are submitted as follows: 1A renal artery, renal vein, and ureteric margins; 1B representative section of tumor, pilot section; 1C-1I tumor adjacent to renal capsule/Gerota's fascia; 1J-1N tumor and renal sinus; 1O-1Q tumor with adjacent kidney tissue; 1S uninvolved kidney; 1T representative sections of adrenal; 1R hilar lymph nodes.

DAG//KLD/KLD

MICROSCOPIC:

The tumor is comprised of a proliferation of small, round to oval to spindle cells arrayed in variable morphologic patterns. In some areas, the cells form nests and small sheets divided by hyalinized fibrous bands, with or without a chicken-wire vascular pattern. In other areas, cells appear more discohesive with abundant myxoid stroma. In other areas, spindle cells form fascicles with hyalinized stroma. The cells have variable cytoplasm ranging from scant to abundant and clear, and sometimes eccentric and eosinophilic. Nuclei are pale or clear with absent or inconspicuous nucleoli. Mitoses and apoptotic bodies are easily identified. Areas of necrosis, cystic degeneration and hyalinization are present. The renal capsule is intact and all surgical margins including Gerota's fascia, renal artery, renal vein, and ureter are free of tumor. Tumor extends into and focally replaces the renal sinus and vascular invasion is present. A fibrous pseudocapsule circumscribes the sarcoma in areas adjacent to uninvolved kidney. Entrapped tubular elements are observed at the periphery of the tumor. Three hilar lymph nodes show reactive features and no evidence of metastatic tumor. No nephrogenic rests are identified. ~~The uninvolved kidney shows acute and chronic tubulointerstitial inflammation with Tamm-Horsfall protein in tubules and the interstitium focally.~~ Dilated tubules contain erythrocytes, granular eosinophilic material, fibrin and/or inflammatory cells. No significant glomerular changes are observed. The adrenal gland is unremarkable.

The following statement applies to all immunohistochemistry, Insitu Hybridization Assays (ISH & FISH), Molecular Anatomic Pathology, and Immunofluorescent Testing:

The testing was developed and its performance characteristics determined by the University of Pittsburgh, Department of Pathology, as required by the CLIA '88 regulations. The testing has not been cleared or approved for the specific use by the U.S. Food and Drug Administration, but the FDA has determined such approval is not necessary for clinical use.

HISTO TISSUE SUMMARY/SLIDES REVIEWED:

Part 1: Kidney, Nephrectomy and mass

Taken: 7/12/2006 14:30 Received: 7/12/2006 14:54

Stain/cnt	Block
H&E x 1	A
Recut x 1	A
H&E x 1	B
Recut x 2	B

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Pathology Report 001144078 CHP COOK AUSTIN REED 06/30/06 11:40

Name COOK AUSTIN REED
 MRN 001144078 CHP
 Physician TYLER-KABARA ELIZABETH
 Report Type Pathology Report
 Date of Event 06/30/06 11:40
 Date of Birth 02/23/2001
 Sex M
 Patient Status ... CHQ
 Last Disch Date .. 07/02/06 10:18
 Document # CHS06-4862
 Authored by JAFFE RONALD
 Account # 0001027756269
 Hosp/Group CHP

PATIENT HISTORY:

Procedure: Excision of left skull lesion.

Pre-Op/Post-Op: Skull lesion.

FINAL DIAGNOSIS:

Scalp tumor biopsy:

HIGH GRADE TUMOR MOST CONSISTENT WITH METASTATIC CLEAR CELL SARCOMA OF KIDNEY (SEE NOTE).

NOTE: In view of the presence of tumors within the kidney, this is likely to represent a metastasis. The tumor morphology and a panel of immunostains that is almost entirely non-reactive has largely excluded the following from the differential diagnosis: Rhabdoid tumor, Neuroblastoma, PNET, Wilms', Rhabdomyosarcoma and Synovial Sarcoma. The diagnosis of Clear Cell Sarcoma is made, to some degree, by exclusion since it has no unique markers, but this is a metastatic site. FISH for Synovial and PNET-type translocations will be reported in an addendum.

The findings were discussed with Dr. P Shaw on 7/3 and 7/7/2005.

My signature is attestation that I have personally reviewed the submitted material(s) and the final diagnosis reflects that evaluation.

MJY/MJY

Pathologist: Ronald Jaffe, MB. BCh.

** Report Electronically Signed Out **

By Pathologist: Ronald Jaffe, MB. BCh.

7/7/2006 16:13

My signature is attestation that I have personally reviewed the submitted material(s) and the final diagnosis reflects that evaluation.

GROSS DESCRIPTION:

The specimen is received in two parts.

Part 1 is received in formalin, labeled with the patient's name and designated "skull lesion." It consists of a single fragment of round tissue soft in consistency and red-pink in appearance, measuring 1.2 x 1.0 x 0.5 cm. A portion of the specimen is snap frozen for potential molecular genetic tests. The remainder of the specimen is entirely submitted in cassette labeled 1A.

Part 2 is received in formalin, labeled with the patient's name and designated "skull lesion." It consists of two fragments of tissue. The

larger fragment is oval, soft in consistency, and red-pink in appearance measuring 2.8 x 1.1 x 0.5 cm. The smaller fragment is round, soft in consistency, and red-pink in appearance. It measures 1.2 x 1.0 x 0.5 cm. A portion of the larger fragment is snap frozen for potential molecular genetic tests and preserved in EM fixative for potential electron microscopy. The remainder of the specimen is submitted entirely in cassettes labeled 2A 2C (cassette 2B is fixed in B plus fixative).

KRC//MJY/MJY

INTRAOPERATIVE CONSULTATION:

Specimen #1 is received for intra-procedural consultation in the operating room. The specimen is identified as "skull lesion". Clinical question: Diagnosis? One imprint is prepared and stained with H&E. One imprint is prepared and stained with Pinacyanol Chloride. Imprint diagnosis: Metastatic neoplasm. RH

MICROSCOPIC:

1. and 2. Sections show the presence of tumor cells in nests and aggregates with varying amounts of fibrovascular stroma between the tumor nests, and minimal production of neuropil by the tumor cells. The tumor cell nuclei are generally large with stippled chromatin and nucleoli that are not prominent. There is a moderate amount of likely eosinophilic cytoplasm. Immunostains reveal that the cells stain strongly for vimentin with some paranuclear accentuation. Stains for cytokeratins CAM5.2 and AE1/AE3 are negative, as is EMA. Pgp and Synaptophysin are negative and CD99 staining is not above background. Desmin and WT-1 stains are non-reactive.

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HISTO TISSUE SUMMARY/SLIDES REVIEWED:

Part 1: Skull mass biopsy

Taken: 6/30/2006 11:40 Received: 6/30/2006 12:15

Stain/cnt	Block
Blank x 4	A
Blank x 4	A
CAM 5.2 x 1	A
AE1/3 x 1	A
Desmin x 1	A
EM Hold x 1	A
EMA x 1	A
Ewing12E7 x 1	A
H&E x 1	A
Recut x 1	A
PGP x 1	A
Synapto x 1	A
Vimentin x 1	A
WT-1 x 1	A
Imp H&E x 2	(none)
Imp Pina x 2	(none)

Part 2: Skull mass biopsy

Taken: 6/30/2006 11:40 Received: 6/30/2006 12:15

Stain/cnt	Block
Blank x 4	A
H&E x 1	A
Imp H&E x 2	A
Imp Pina x 2	A