

SPR 2018 Oncology Session
Saturday Workshop - May 19, 2018
SAM References

GENERAL ONCOLOGY SECTION

Bone Tumor Update

Kelley W. Marshall, MD

1. Which of the following is the strongest independent predictor of overall survival in osteosarcoma?
- A. Age at diagnosis
 - B. Tumor size
 - C. Presence of metastatic disease at diagnosis
 - D. Response to neoadjuvant chemotherapy

Correct Answer: D

Rationale: Poor histologic response to preoperative chemotherapy increases mortality by a factor of 2.4! Answer A is incorrect. Explanation: While younger presentation <14 yo can portend a worse prognosis, it is not the strongest prognosticator. Answer B is incorrect. Explanation: Large tumors and tumors with incomplete resection are associated with poorer outcome, but again this is not the strongest prognosticator. Answer C is incorrect. Explanation: While metastatic disease at diagnosis is associated with poorer outcomes, these patients have the potential for cure if they are good responders and any residual metastases are amenable to complete surgical resection.

Reference:

- Bielack SS, Kempf-Bielack B, Delling G, et al. Prognostic factors in high-grade osteosarcoma of the extremities or trunk: an analysis of 1702 patients treated on neoadjuvant cooperative osteosarcoma study group protocols. *J Clin Oncol* 2002;20(3):776–90.

2. All of the following are true of the Ewing Sarcoma Family of Tumors (ESFT) EXCEPT:
- A. 85% of tumors carry a specific genetic aberration
 - B. Local excision alone is curative in the vast majority of patients
 - C. Askin tumor is a member of the ESFT
 - D. Presence of metastatic disease at diagnosis is the strongest adverse clinical prognostic factor

Correct Answer: B

Rationale: Unlike osteosarcoma, the ESFT should be considered micrometastatic at presentation! Historically 95% of patients treated with local control alone died of later metastases. Answer A is true. Explanation: 85% of tumors have a pathognomonic balanced chromosomal translocation t(11;22) (q24;q12). Answer C is true. Explanation: Ewing sarcoma of bone, PNET, and extraskeletal Ewing sarcoma complete the family. Answer D is true. Explanation: Without metastatic disease 5 yr survival= 85%, with metastatic disease=27%.

Reference:

- Pediatric Malignant Bone Tumors: A Review and Update on Current Challenges, and Emerging Drug Targets Twana M. Jackson, MD, Mark Bittman, MD, and Linda Granowetter, MD *Curr Probl Pediatr Adolesc Health Care* 2016;46:213-228

Whole Body Imaging in Cancer Predisposition Syndromes

Stephan D. Voss, MD, PhD

3. Which is not an advantage of whole body MRI for tumor predisposition syndrome screening?

- A. Sensitive for soft tissue, osseous, and bone marrow disease
- B. No ionizing radiation
- C. Easy to bill for using accepted CMS codes
- D. Includes both neuro and non-neuro imaging in single exam

Correct Answer: C

Rationale: One of the challenges in standardizing whole body MRI exam protocols and implementing universal screening recommendations has been the inability to bill for the exam due to the lack of a wbMRI CPT code. A number of strategies can be used for reimbursement, but these are institution dependent.

The other choices are examples of advantages for use of wbMRI in CPS screening.

Reference:

- Greer MC, Voss SD, States LJ. Pediatric Cancer Predisposition Imaging: Focus on Whole-Body MRI. Clin Cancer Res. 2017. 23(11):e6-e13.
- Klenk C, Gawande R, Uslu L, Khurana A, Qiu D, Quon A, Donig J, Rosenberg J, Luna-Fineman S, Moseley M, Daldrup-Link HE. Ionising radiation-free whole-body MRI versus (18)F-fluorodeoxyglucose PET/CT scans for children and young adults with cancer: a prospective, non-randomised, single-centre study. Lancet Oncol. 2014. 15(3):275-85

4. All of the following are true regarding whole body surveillance imaging for cancer predisposition screening, EXCEPT:

- A. Can be performed without use of Gd-based contrast media
- B. Very few incidental findings
- C. Early detection of malignancy in LFS correlates with improved outcome (survival)
- D. Combining wbMRI with PET, DWI or use of iron-oxide agents can increase sensitivity for disease detection

Correct Answer: B

Rationale: In the study by Anupindi et al. a large number of incidental findings were reported (average 2.5/patient) during wbMRI screening. The other choices are all true statements.

Reference:

- Greer MC, Voss SD, States LJ. Pediatric Cancer Predisposition Imaging: Focus on Whole-Body MRI. Clin Cancer Res. 2017. 23(11):e6-e13.
- Klenk C, Gawande R, Uslu L, Khurana A, Qiu D, Quon A, Donig J, Rosenberg J, Luna-Fineman S, Moseley M, Daldrup-Link HE. Ionising radiation-free whole-body MRI versus (18)F-fluorodeoxyglucose PET/CT scans for children and young adults with cancer: a prospective, non-randomised, single-centre study. Lancet Oncol. 2014. 15(3):275-85

CNS Manifestations in Cancer Predisposition Syndromes

Sarah S. Milla, MD, FAAP

5. What CNS neoplasm is seen in patients with von Hippel Lindau?

- A. Schwannoma
- B. Hemangioblastoma
- C. Optic Nerve Glioma
- D. Ganglioglioma

Correct Answer: B

Rationale: B is correct. Hemangioblastoma is seen in VHL patients. A is incorrect. This is not typical and concern for low grade neoplasm should be raised. C is incorrect. This is not typical and other causes should be considered. D is incorrect. This is not typical and other causes should be considered

Reference:

- Vijapura C, et al. Genetic Syndromes Associated with Central Nervous System Tumors. RadioGraphics 2017; 37: 258-280.
- Monsalve J, et al. Imaging of Cancer Predisposition Syndromes in Children. RadioGraphics 2011; 31(1): 263-280
- Dunbar EM, et al. Genetic counseling and tumor predisposition in neur-oncology practice. Neuro-Oncology Practice 2018; 3(1): 17-28.

6. What location is typical for T2 FLAIR signal abnormalities in NF?

- A. Globus pallidus
- B. Frontal white matter
- C. Caudate
- D. Corticospinal tract

Correct Answer: A

Rationale: A is correct and is a classic location for involvement in NF1. B is incorrect. This is not typical and concern for low grade neoplasm should be raised. C is incorrect. This is not typical and other causes should be considered. D is incorrect. This is not typical and other causes should be considered.

Reference:

- Vijapura C, et al. Genetic Syndromes Associated with Central Nervous System Tumors. RadioGraphics 2017; 37: 258-280.
- Monsalve J, et al. Imaging of Cancer Predisposition Syndromes in Children. RadioGraphics 2011; 31(1): 263-280
- Dunbar EM, et al. Genetic counseling and tumor predisposition in neur-oncology practice. Neuro-Oncology Practice 2018; 3(1): 17-28.

Pros and Cons of PET-CT and PET-MRI in Pediatric Oncology

Michael S. Gee, MD PhD

Lisa States, MD

7. PET/MR offers benefit over PET/CT in CPS because;

- A. It replaces whole body MRI.
- B. Diffusion weighted imaging adds value.
- C. Radiation dose is decreased.
- D. Sedation is eliminated.

Correct Answer: C

Rationale: Radiation dose is increased. Further investigation in this population includes using micro-dose techniques +/- increased acquisition times for whole body imaging or focused evaluation of a finding on whole body MRI. Answer A is incorrect. Explanation: Whole body MRI will remain the surveillance tool in the evaluation of CPS, such as Li Fraumeni Syndrome, due to lack of ionizing radiation. Answer B is incorrect. Explanation: The added value of diffusion weighted imaging in the diagnosis and response to therapy is an active area of research. Answer C is incorrect. Explanation: Sedation requirements for PET/MR are greater than that for PET/CT.

Reference:

- Detection of Small Pulmonary Nodules with Ultrashort Echo Time Sequences in Oncology Patients by Using a PET/MR System, Burriss NS, et. Al, Radiology. Vol 278, No 1, Jan 2016

8. PET/MR evaluation of the pulmonary nodules can be improved using;

- A. Ultrashort TE (UTE) technique
- B. Diffusion weighted imaging
- C. STIR
- D. Blanket coil

Correct Answer: A

Rationale: Free breathing UTE has a high sensitivity for the detection of small pulmonary nodules and requires further investigation. Answer B is incorrect. Explanation: Diffusion weighted imaging is susceptible to motion and susceptibility artifact. Answer C is incorrect. Explanation: STIR imaging is part of lung imaging protocols but is not superior to 3D-GRE sequences. Answer D is incorrect. Explanation: The blanket coil increases patient comfort but has not yet been studied for the evaluation of pulmonary nodules.

Reference:

- Detection of Small Pulmonary Nodules with Ultrashort Echo Time Sequences in Oncology Patients by Using a PET/MR System, Burriss NS, et. Al, Radiology. Vol 278, No 1, Jan 2016

Novel Radiotracers for Oncologic Imaging

Scott E. Snyder, PhD

9. Which of the following PET radiotracers does not accumulate by a metabolic trapping mechanism?

- A. [¹⁸F]Fluorodeoxyglucose (FDG)
- B. ⁶⁸Ga-DOTA-tyr3-Octreotide (⁶⁸Ga-DOTATOC)
- C. [¹¹C]Methionine (MET)
- D. meta-[¹⁸F]Fluorobenzylguanidine (MFBG)

Correct Answer: B

Reference:

- Abongwa C, Mott S, Schafer B, McNeely P, Abusin G, O'Dorisio T, Zamba G, O'Dorisio MS, Menda Y.. Safety and accuracy of ⁶⁸Ga-DOTATOC PET/CT in children and young adults with solid tumors. *Am J Nucl Med Mol Imaging*, 2017, 7:228-235.

10. Which of the following IS NOT a limiting factor specific to the development of novel radiopharmaceuticals for pediatric use?

- A. Typically can't recruit healthy volunteer control subjects
- B. Necessity for sedation of many subjects
- C. RDRC limits on allowable radiation dose to minors
- D. Lack of funding for clinical trials

Correct Answer: D

Reference:

- Abongwa C, Mott S, Schafer B, McNeely P, Abusin G, O'Dorisio T, Zamba G, O'Dorisio MS, Menda Y.. Safety and accuracy of ⁶⁸Ga-DOTATOC PET/CT in children and young adults with solid tumors. *Am J Nucl Med Mol Imaging*, 2017, 7:228-235.

RARE TUMORS SECTION

Neuroendocrine Tumors: What's New in Imaging?

Andrew T. Trout, MD

11. Which of the following is correct regarding preparing a patient for a Ga-68 DOTATATE PET scan?

- A. Patients must fast for 6 hours to reduce soft tissue uptake of the radiotracer
- B. Patients should be warmed to reduce brown fat uptake of the radiotracer
- C. Patients should consume a high fat, low carb diet for 2 meals prior to their scan to reduce cardiac uptake of the radiotracer
- D. Short acting somatostatin analogues should be stopped for 24 hours prior to the scan

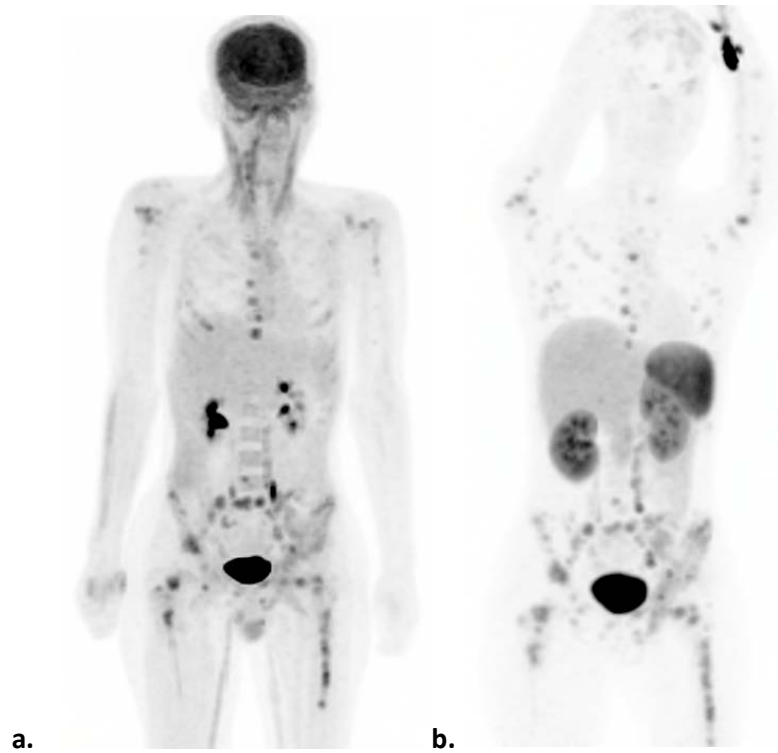
Correct Answer: D

Rationale: Somatostatin analogues competitively inhibit uptake of Ga-68 DOTATATE. Scans should be performed immediately prior to a dose of long acting analogues and short acting analogues should be held for 24 hours prior to scanning. Answer a is incorrect. Explanation: Fasting is more relevant to F-18 FDG scanning to optimize biodistribution. Answer b is incorrect. Explanation: Brown fat uptake is not an issue with Ga-68 DOTATATE. Answer c is incorrect. Explanation: This preparation is useful for myocardial F-18 FDG PET imaging but is not relevant to Ga-68 DOTATATE imaging.

Reference:

- Hofman MS, Lau WF, Hicks RJ. Somatostatin receptor imaging with 68Ga DOTATATE PET/CT: clinical utility, normal patterns, pearls, and pitfalls in interpretation. Radiographics. 2015 Mar-Apr;35(2):500-16.

12. Of these PET scans, which is the Ga-68 DOTATATE scan?



Correct Answer: B

Reference:

- Hofman MS, Lau WF, Hicks RJ. Somatostatin receptor imaging with 68Ga DOTATATE PET/CT: clinical utility, normal patterns, pearls, and pitfalls in interpretation. *Radiographics*. 2015 Mar-Apr;35(2):500-16.

Zebras in Pediatric Oncology

Sue C. Kaste, DO

13. Growing teratoma syndrome;

- Represents transformation of benign teratoma into malignant immature germ cell tumor
- Represents benign metastatic disease
- Is considered treatment failure of non-seminomatous germ cell tumors
- Is diagnosed by elevated alpha fetoprotein

Correct Answer: B

Rationale: Answer A is incorrect. Explanation: This syndrome is associated with treated malignant germ cell tumor but represents benign disease, not the transformation of benign into malignant disease. Answer C is incorrect. Explanation: This syndrome is often confused with treatment failure and development of progressive disease. One hypothesis suggests that effective treatment of malignant components may actually select out benign components that continue to slowly grow. Answer D is incorrect. Explanation: Serum markers are normal.

Reference:

- Kataria SP et al. *Indian J Surg Oncol* March 2017 8(1):46-50 DOI 10.1007/s13193-016-0568-3
- Logothetis CJ et al. *Cancer* 1982 Oct 15; 50(8):1629-35

14. Malignant ectomesenchymoma;

- Used to be called triton tumor
- Most commonly arises in the thalamus
- Is cytogenetically related to osteosarcoma
- Contains intermixed foci of rhabdomyosarcoma and neuroblastic

Correct Answer: D

Rationale: Answer A is incorrect. Explanation: A Triton tumor has benign histology and does not contain ganglion cells. Answer B is incorrect. Explanation: This tumor arises in the soft tissues. Answer C is incorrect. Explanation: This tumor is cytogenetically related to rhabdomyosarcoma, not osteosarcoma.

References:

- Dantonello TM et al. *Pediatr Blood and Cancer* 2013; 60:224-229
- Huang SC et al. *Am J Surg Pathol* 2016; JUL 40(7):876-885
- Griffin BB et al. *Int J Surg Pathol* 2017; DOI:10.1177/1066896917734915
- Freita AS et al. *Pediatr Neurosurg* 1999 Jun;30(6):320-30

Update on RECIST

M. Beth McCarville, MD

15. The revised RECIST 1.1 Criteria do not address:

- A. Lymph node measurement
- B. Maximum number of target lesions
- C. The use of PET-CT in assessing response
- D. Distinguishing pseudo-progression from true progression

Correct Answer: D

Reference:

- Eisenhauer EA et al., Eur Jo Cancer 45(2009):228-247

16. The PERCIST criteria do not require:

- A. Measurement of background FDG uptake in the liver or aorta
- B. The use of the Standardized Uptake Value corrected for lean body mass (SUL)
- C. Measurement of the same target lesion at each response imaging time point
- D. Use of a spherical volume of interest (VOI)

Correct Answer: C

Reference:

- Hyun J et al., Radiol 280(2):Aug 2016:576-584