Multiple Non Ossifying Fibromas in 20 year old patient with Speckled Lentiginous Nevus syndrome; Jaffe-Campanacci syndrome?

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ABSTRACT

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validation of the software, statistical analysis as a biomarker for outcome was restricted to 12 patients displaying initial BSI between 3 and 13. Three additional patients with higher tumor loads were analyzed separately. Results: Manual corrections were necessary in one patient due to obstruction induced urine uptake in the upper urinary tract and in two patients due to incontinence induced urine contamination (3/30 scans). In one patient pelvic metastasis could not be discriminated from bladder activity. Comparison of BSI Version 1.8 and 2.1 for 15 patients revealed higher values in particular at higher tumor burden both for baseline (mean 6.3 vs 8.7) and for follow up (mean 7.1 vs 9.4). However, difference between baseline and interim-staging was not significant between both versions (p=0.83). 5/15 patients showed a decrease of bone scan index after the first two cycles of radium-223. In contrast, only two patients with initial low levels of serum PSA (<100 ng/ml) presented a PSA decline at this time point. Cox regression analysis showed a trend towards validity for bone scan index to serve a predictive marker similar to serum PSA (p=0.035 and p= 0.18 for BSI version 1.8 and 2.1; p=0.034 for PSA) Conclusion: The bone scan index is a valuable and easily applicable tool to quantify tumor load and treatment response on bone scans in mCRPC patients undergoing Radium-223 treatment.

EP010

Multiple Non Ossifying Fibromas in 20 year old patient with Speckled Lentiginous Nevus syndrome; Jaffe-Campanacci syndrome?

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INTRODUCTION: Speckled lentiginous nevus syndrome is a recently described syndrome with male to female ratio of 4:3; usually present at birth but may appear during first years of life. Cafe au lait macules can be topped by either browns or black macules or papules. Patients usually present with musculosceletal and neurological anomaly at ipsilateral site. Nonossifying fibromas (NOFs) are the most common benign bone tumor in children. The tumors affected long bones as the humerus, the tibia, the femur, the fibula. Approximately 8 % of people with NOF will have more than one tumor. It is uncommon to have more than two or three tumors except in certain very rare conditions like Jaffe - Campanacci 's syndrome. This syndrome can present only with skin patches and NOFs, but also as a part of more severe systemic presentation like cardiovascular, renal, ocular abnormalities and mental retardation. CASE PRESENTATION: a 20 year old female with a known diagnosis of Speckled lentiginous nevus Syndrome presented to our Department in 2012 for evaluation of prolonged discomfort in the right leg. Performed MRI showed atrophy in right leg and hyperintensive ovale zone (24x10 mm) in distal part of left femur. Three phase bone scan revealed decreased uptake in all 3 phases in right lower extremity, without focal lesions in the left and increased uptake in right humerus (initally evaluted as a bone infarct or eosinophilic granuloma). Bone scan performed in 2013 showed increased left - right asymmetry uptake between lower extremities. Since than she developed paresthesia in right arm. X-ray showed a 41x10 mm zone in proximal diaphysis of right humerus suspected for NOF. Bone scan performed in 2016 showed same changes in right humerus as 2012, but with new focal increased uptake in distal part of diaphysis of left femur with same scintigraphy pattern as in the right arm. Diagnosis of multiple non-ossifying fibromas was presumed. CONCLUSION: Regarding the clinical presentation and bone scan, diagnosis of Jaffe-Campanacci syndrome was presumed.

EP011

Volumetric FDG-PET indices for the assessment of histological response to neoadjuvant chemotherapy and outcome in paediatric patients with Ewing sarcoma and osteosarcoma

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Aim: Response to chemotherapy is a prognostic factor in patients with Ewing sarcoma (EWS) and Osteosarcoma (OST). Several studies showed high diagnostic accuracy of positron emission tomography using 18F-fluoro-2-deoxy-glucose (FDG-PET) for response to initial chemotherapy (CHT) in these patients and a correlation with outcome. Despite a potential impact of age on the prognosis of bone sarcoma patients, most of these studies mixed adult and paediatric populations. The objective of this retrospective work was to evaluate the prognostic value of quantitative indices derived from FDG-PET in an homogeneous paediatric bone sarcoma population including EWS and OST. Methods: 31 paediatric patients with EWS (median age 14.7 years) and 31 with OST (median age 12.8 years) were included. All patients were treated with CHT, and underwent surgery for local control. All patients had FDG PET at diagnosis and after induction CHT, prior to surgery. Several parameters for assessment of response of the primary tumour to therapy by FDG PET were evaluated: SUV_{max}, SUV_{peak}, SUV_{mean}, metabolic tumor volume (MTV), total lesion glycolysis (TLG), Tumor to contralateral Background Ratio (TBR), 7 textural features (TF) and 3 shape features (SF). For each metric requiring a segmentation step, 3 delineation methods were compared including an adaptative approach, 40% of SUVmax and SUV>2.5. Each pair of FDG-PET scan were also classified as responding (R) and non-responding (NR) according to PERCIST criteria. Results were then compared to histopathological regression of the resected tumour as defined by Salzer-Kuntschik and to the clinical follow-up for survival evaluation (median of follow-up: 5 years). Results: After CHT, the absolute SUVs, MTV, TLG, TF, SF or TBR, the reductions of SUVs, TF or SF, TBR and PERCIST criteria were neither significantly associated with histologic response nor overall survival or progression free survival for either EWS or OST. The MTV and TLG reduction (segmentation SUV > 2.5) were the only quantitative indices that significantly discriminated histopathological responders from non-responders using the entire patient population ($\Delta MTV_{2.5}$, p=0.0109; $\Delta TLG_{2.5}$, p=0.0076) as well as in the subgroup of OST patients ($\Delta MTV_{2.5}$, p=0.0032; $\Delta TLG_{2.5}$, p=0.0040), but not in the EWS subgroup. Conclusion: Only MTV and TLG reduction seem to enable accurate non-invasive assessment of histopathological reduction after CHT in the OST paediatric population but reveal limited additional prognostic value in paediatric patients with EWS.

EP012

Prognosis value of SUV-based metrics, textural and shape features derived from initial FDG-PET in paediatric patients with Ewing sarcoma and osteosarcoma

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Purpose: Histologic response, measured by the percentage of tumor cells remaining after neoadjuvant chemotherapy (CHT) is the main prognostic factor in Osteosarcoma (OST) and Ewing sarcoma (EWS). Previous positron emission tomography using 18F-fluoro-2-deoxy-glucose (FDG-PET) studies tried to determine the value of FDG uptake as an accurate and non-invasive preoperative marker of response. Yet most of them explored heterogeneous populations mixing different histological