# Extra-renal malignant rhabdoid tumor of head and neck region: Characteristics of tracer uptake on FDG PET/CT in tumor with rare histology

Sir,

A 40-year-old female presented with complaints of diminished vision and deviation of eyes since 15 days, with a single episode of convulsion. CT scan of head and neck showed a large mass arising from the sphenoid sinus, eroding the skull base. Endoscopy guided biopsy was done elsewhere which was suggestive of myoepithelial tumor. Since the extent of lesion made it inoperable, patient was referred to our Institution for whole body F-18 Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography (FDG PET/CT) study, for staging, prior to definitive

radiotherapy (RT). Maximum Intensity Projection (MIP) image showed no appreciable tracer uptake in head and neck region (1a), or elsewhere in the body. Axial CT and fused PET/CT images showed an enhancing extensive soft tissue mass involving sphenoid sinus, invading the cavernous sinus and encasing bilateral carotid arteries, spreading into the clivus and skull base [Figure 1b and c - arrow]. It had a maximum Standardised Uptake value of 3.6. No discrete cervical adenopathy was noted. There was no evidence of distant metastases. Since the pattern of uptake was unlike myoepithelial tumors, which usually show intense FDG avidity, biopsy was repeated from the area with maximum SUV. Histopathology [H and E, ×40 - Figure 2a] showed high grade malignant tumor with tumor cells arranged in sheets, and majority showing 'rhabdoid' morphology. On immunohistochemistry (2b), loss of staining was seen for IN1 antibody, which was confirmatory for diagnosis of extra-renal rhabdoid tumor. Malignant Rhabdoid Tumor (MRT) was first described as a rare sarcomatous variants of Wilms tumour,[1] primarily because of its renal origin, in children. Though originally described as primary renal neoplasm, tumors with similar histological characteristics have been identified at extra-renal sites such as thymus, liver, central nervous system, heart, chest wall and extremities; these are termed as extra-renal malignant rhadboid tumors (ERRT).[2] Given its aggressive nature, early diagnosis and treatment becomes important. Though the diagnosis is purely based on histology and immunochemistry, as seen in our case, at the same time, it is essential to know the uptake characteristics in these rare histological variants. Oda *et al.,* have shown that the characteristic 'rhabdoid' cells of ERRTs are also seen in certain soft tissue sarcomas such as synovial sarcomas, extraskeletal myxoid chondrosarcoma and leiomyosarcoma.[3] As per the existing literature, ERRTs demonstrate intense FDG uptake at the primary and metastatic sites.[4] However, our case is an exception wherein the tumor shows very low-grade tracer concentration. Also, the pattern of uptake is quite uniform, and cannot be appreciated on Maximum Intensity Projection (MIP) images. There are only two such reports in literature, especially in children, wherein low-grade FDG uptake is seen in ERRTs.[5,6] Thus, ERRTs, which are rare, aggressive histological variants, are an addition to the list of tumors, which may show low grade metabolic activity on PET/CT imaging.

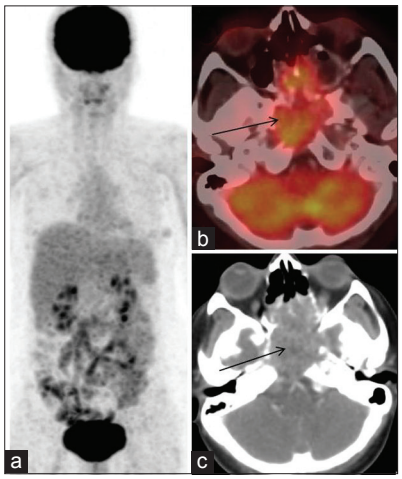


**Figure 1:** (a) MIP image shows no appreciable increased tracer uptake, whereas low-grade metabolic activity (b - arrow) is seen in enhancing ill-defined soft tissue mass in the sphenoid sinus (c - arrow), on axial PET/CT and CT images, with evidence of clival erosion suggestive of skull base extension

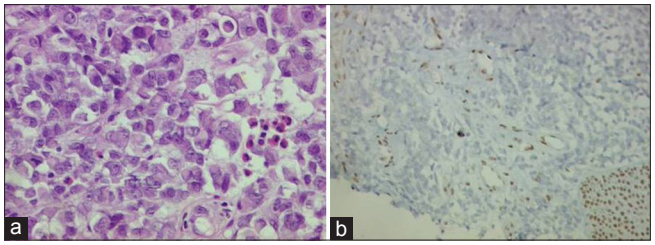


**Figure 2:** (a) (H and E stain, ×40 magnification) Extra-renal rhabdoid tumour containing sheets of malignant cells. Tumor cells contain large nuclei and prominent nucleoli. Eosinophilic cytoplasm is conspicuous imparting 'rhabdoid' phenotype to the tumor cells. (b) Immunohistochemistry for antibody to INI1; tumor cell nuclei do not show any staining while nuclei of overlying epithelium and endothelial cells lining blood vessels show positive staining (serving as internal positive control)

**Ameya D. Puranik, Nilendu Purandare, Mukta Ramadwar[1], Archi Agrawal, Sneha Shah, Venkatesh Rangarajan**

Departments of Nuclear Medicine and Molecular Imaging and [1]Pathology, Tata Memorial Hospital, Mumbai, Maharashtra, India

**Address for correspondence:**
Dr. Venkatesh Rangarajan,
Department of Nuclear Medicine and Molecular Imaging,
Tata Memorial Centre, Mumbai, Maharashtra, India.
E-mail: drvrangarajan@gmail.com

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