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An Uncommon Case of Pediatric Esthesioneuroblastoma Presenting as SIADH: $^{18}$F-FDG PET/CT in Staging and Post-Therapeutic Assessment

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Abstract: Esthesioneuroblastoma (ENB) is an uncommon neuroendocrine tumor originating from the olfactory neuroepithelium and accounts for 3–6% of all intranasal tumors [1]. ENBs can be locally aggressive and cause invasion and destruction of surrounding structures. Histological grading and clinical stage at presentation are highly predictive of survival and especially presence of lymph node and distant metastases are determining prognostic factors [2–5]. Thus, reliable imaging is essential in these patients. Conventional imaging modalities for staging ENB are magnetic resonance imaging (MRI) and computed tomography (CT). However, fluorine-18 fluoro-2-deoxy-D-glucose positron emission tomography/CT ($^{18}$F-FDG PET/CT) has been reported as a valuable adjunct and was found to upstage 36% of ENB patients compared to conventional imaging [6]. We present a case demonstrating the diagnostic work-up and follow-up with $^{18}$F-FDG PET/CT in a young patient with ENB with a highly atypical clinical presentation.

Keywords: esthesioneuroblastoma; $^{18}$F-FDG-PET/CT; pediatric oncology; paraneoplastic syndromes
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18F-FDG PET/CT in Staging and Post-Therapeutic Esthesioneuroblastoma Presenting as SIADH: An Uncommon Case of Pediatric 

Interesting Images 

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Figure 1. Initial scan—(left, middle): CT and fused 18F-FDG PET/CT, coronal view; (right): 18F-FDG PET, maximal intensity projection. A 17-year-old girl with no previous medical history was admitted for psychiatric evaluation due to sudden onset of symptoms consisting of hypomania and hallucinations. Prior to admission the patient had suffered from recurrent emesis for a couple of days. As a part of the standard diagnostic work-up, laboratory tests found severe hyponatremia with a serum sodium level of 110 mmol/L (normal range: 135–147 mmol/L), normal serum potassium of 3.7 mmol/L (normal range: 3.8–4.7 mmol/L), low serum ionized calcium 1.14 mmol/L (normal range: 1.18–1.25 mmol/L), high urine sodium level 93 mmol/L (normal range: 20–40 mmol/L) and normal urine osmolality 406 mmol/kg (normal range 300–900 mmol/kg). The patient was clinically normovolemic and had no abnormal clinical signs besides the psychiatric symptoms. Hypothyroidism and adrenal insufficiency were excluded and syndrome of inappropriate antidiuretic hormone (SIADH) was suspected as the cause of hyponatremia. Treatment with fluid restriction and intravenous isotonic sodium chloride was initialized. In suspicion of an underlying intracranial pathology and as part of standard diagnostic work-up a CT scan of the cerebrum was performed. This revealed no pathological intracranial lesions, but a polypoid mass with calcifications in the left maxillary sinus protruding into the left nasal cavity. Subsequently, the patient was referred to 18F-FDG PET/CT for characterization and staging, which demonstrated the mass in the left maxillary sinus to be highly FDG avid (SUV_max 7.4). Besides symmetrical FDG uptake in the pharyngeal lymphatic tissue, which is considered benign, there were no sites of increased FDG uptake in the cervical lymph nodes. Furthermore, no other pathological foci suspicious of metastatic disease were found on the whole-body scan. The tumor was resected en bloc endoscopically under the use of image guidance through a medial maxillectomy. Following surgery, the patient’s symptoms resolved and the electrolytes normalized quickly. Histological and immunohistochemical examination of tumor material were consistent with an ectopic esthesioneuroblastoma Hyams grade I positive for chromogranin, synaptophysin, neuron-specific enolase, calretinin, and vasopressin. Histologically the tumor cells reached the margins of the specimen, and the operation was considered non-radical. The patient therefore received post-operative radiation therapy to the surgical area, with no elective neck volumes. Clinical examination and renewed 18F-FDG PET/CT scan three months post-treatment showed complete response (Figure 2).
The authors declare no conflict of interest.

References


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