Positron Emission Tomography–Computed Tomography Evaluation of Maxillary Rhabdomyosarcoma in an Adult

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Abstract

Rhabdomyosarcoma is a rare primary head/neck tumor. This article illustrates the utility of multimodality imaging techniques in a 34-year-old male with metastatic alveolar rhabdomyosarcoma, including an apparently favorable metabolic response to chemotherapy between baseline and early post-treatment positron emission tomography–computed tomography. Clinical and histopathologic features of the disease are also presented.

Keywords

Alveolar rhabdomyosarcoma, magnetic resonance imaging, fluorodeoxyglucose positron emission tomography–computed tomography (F18-FDG PET-CT), myo-D1, Children's Oncology Group ARST0431 trial

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Rhabdomyosarcoma is the most common soft-tissue sarcoma in pediatric patients, with less frequent occurrence in young adults. The head/neck is a common site for this tumor with frequent involvement of the orbit, nasal cavity, nasopharynx, and paranasal sinuses. Although imaging features of head/neck rhabdomyosarcoma are non-specific and may resemble an aggressive fungal infection or other malignant neoplasm, histopathologic features are often definitive with characteristic myoid differentiation and immunohistochemical profile. This article presents a rare case of maxillary sinus rhabdomyosarcoma in an adult patient, with demonstration of a favorable metabolic response to multiagent chemotherapy between baseline and early post-treatment fluorodeoxyglucose positron emission tomography–computed tomography (F18-FDG PET-CT).

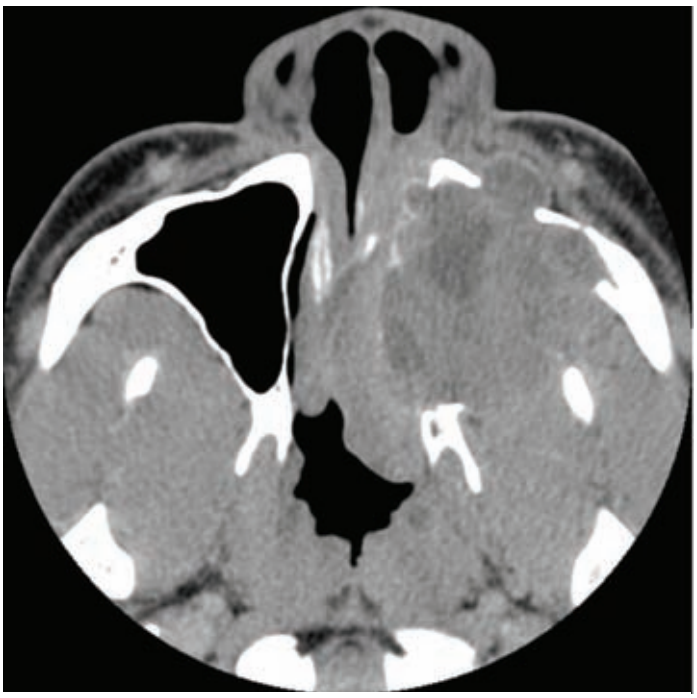
Case Report

A 34-year-old man presented to the emergency department (ED) with left facial pain, pressure, and nasal congestion of several weeks' duration. Within a three-month period, the patient had been seen in the ED twice and was treated for presumed sinusitis, demonstrating symptoms refractory to antibiotic treatment. A review of the patient's medical history was otherwise unremarkable.

Physical examination revealed an obvious left proptosis without abnormality of visual acuity. Extraocular movements were symmetric and intact. His otoscopic examination was unremarkable. Rhinoscopic examination revealed rightward deviation of the nasal septum with obliteration of the left nasal passage by an apparent soft-tissue mass beyond the vestibule. Oropharyngeal examination revealed no significant abnormality. No palpable lymph nodes of the neck were identified. Cranial nerve examination revealed hypesthesia in the distribution of the left maxillary nerve. Laboratory examination demonstrated a mild hypercarbia and lymphocytosis.

The patient underwent non-contrast CT of the sinuses, which revealed a large soft-tissue mass centered in the left maxillary sinus with significant osseous erosion into the hard palate, left orbit, and left nasal cavity, as well as displacement of left maxillary soft tissues (see *Figure 1*). Inflammatory changes were also seen in the ipsilateral frontal and sphenoid sinuses. Contrast-enhanced magnetic resonance imaging (MRI) of the sinuses demonstrated a heterogeneously enhancing cystic mass that was primarily T1 isointense to gray matter (see *Figure 2*). Additional MRI findings included anterior extension through the left infraorbital foramen and some erosion of the cribriform plate, without definite invasion into the anterior cranial fossa. Given the aggressive appearance of the lesion, differential considerations included neoplastic etiologies such as inverting papilloma or squamous cell carcinoma, or possibly an invasive fungal sinusitis. Pathology revealed a malignant small round blue cell tumor with an immunohistochemical profile consistent with rhabdomyosarcoma (alveolar type). Histopathologically, the tumor demonstrated a growth pattern of the solid variant, with periodic acid Schiff positivity of the neoplastic cells. The tumor cells showed positive immunoreactivity for desmin and myo-D1, with a nuclear staining pattern (see *Figure 3*)

Figure 1: Axial Unenhanced Computed Tomography Image



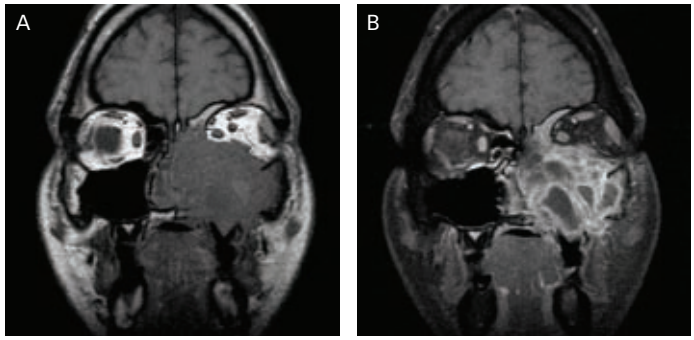
This image demonstrates a large mass of heterogeneous attenuation centered in the left maxillary sinus, with erosion of the maxillary sinus walls. Adjacent soft-tissue infiltration and left nasal cavity obstruction are also evident.

Oncologic staging using F18-FDG PET-CT revealed significant metabolic activity associated with the primary neoplasm in the maxillary sinus (see *Figure 4*). An enlarged ipsilateral level IIA lymph node in the neck with moderate metabolic activity (maximum standardized uptake value [SUV] 4.9) was demonstrated. Histopathologic study of this node revealed findings consistent with metastatic rhabdomyosarcoma. The PET-CT also demonstrated an osteolytic lesion in the ipsilateral left iliac bone with moderate hypermetabolism (maximum SUV 6.5) (see *Figure 5*); cytologic examination of this lesion also revealed malignant small round blue cells consistent with rhabdomyosarcoma. Cytologic examination of the cerebrospinal fluid was performed given the questionable intradural extent of the primary tumor, with no malignant cells identified. The patient was determined to have stage 4 disease and was enrolled in the Children's Oncology Group ARST0431 trial, to include intensive multiagent chemotherapy with vincristine/doxorubicin/cyclophosphamide (VDC) and planned adjunctive regional external-beam radiation therapy at 20 weeks. Follow-up PET-CT four weeks after initiation of chemotherapy showed a favorable early metabolic response at the site of the primary lesion (see *Figure 6*) as well as within the nodal and osseous metastatic foci (see *Figure 7*). Repeat bone marrow cytology of the left iliac bone metastasis was repeated, with no malignant cells identified

Discussion

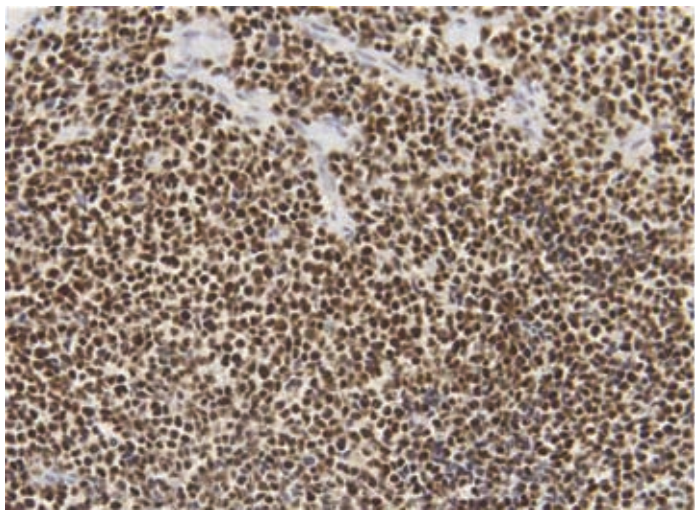
Although rhabdomyosarcoma is considered the most common pediatric solid tumor of the head and neck, it is a rare primary head/neck tumor in adult patients.[1,2] Based on the modified World Health Organization classification, two main morphologic subtypes are described: embryonal rhabdomyosarcoma (ERMS), which encompasses the botryoid subtype, and alveolar rhabdomyosarcoma (ARMS). The

Figure 2: Coronal Pre-/Post-contrast T1-weighted Magnetic Resonance Images



The pre-contrast image (A) demonstrates a left maxillary sinus mass which is isointense to gray matter. The post-contrast (B) T1-weighted image demonstrates intense heterogeneous enhancement with non-enhancing cystic components as well as erosion of the cribriform plate. No definite invasion into the anterior cranial fossa is seen.

Figure 3: Low-power Micrograph of Solid-variant Alveolar Rhabdomyosarcoma with Cells

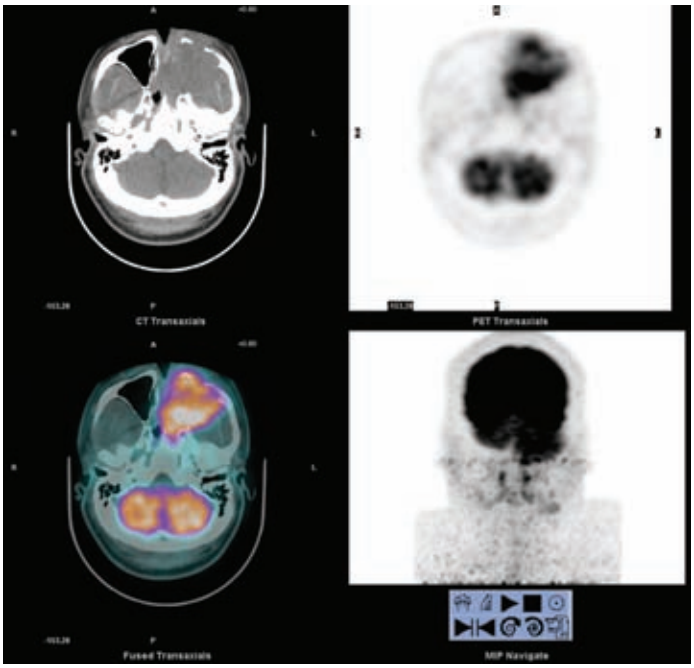


This image demonstrates positive immunoreactivity of the tumor to myo-D1, a nuclear regulatory protein in skeletal muscle differentiation. Note the underdevelopment of the alveolar pattern in this solid-variant specimen, which exhibits a paucity of intervening fibrous septae.

diagnostic distinction between ERMS and ARMS is important because the alveolar subtype portends a worse prognosis owing to more frequent metastatic dissemination, requiring a modified therapeutic regimen.[2,3] The case described above is a rare case of primary ARMS in an adult patient with extensive local bony erosion and soft-tissue infiltration, ipsilateral nodal extension, and distant metastatic involvement. The tumor demonstrates a solid variant pattern of growth, as previously described by Tsokos and Triche.[2,4] Positive staining for desmin, a muscle-specific actin, and myo-D1, a myogenic nuclear regulatory protein,[2,3] provided precise diagnosis and eliminated other more common head/neck neoplasms such as squamous cell carcinoma, lymphoma, and melanoma.

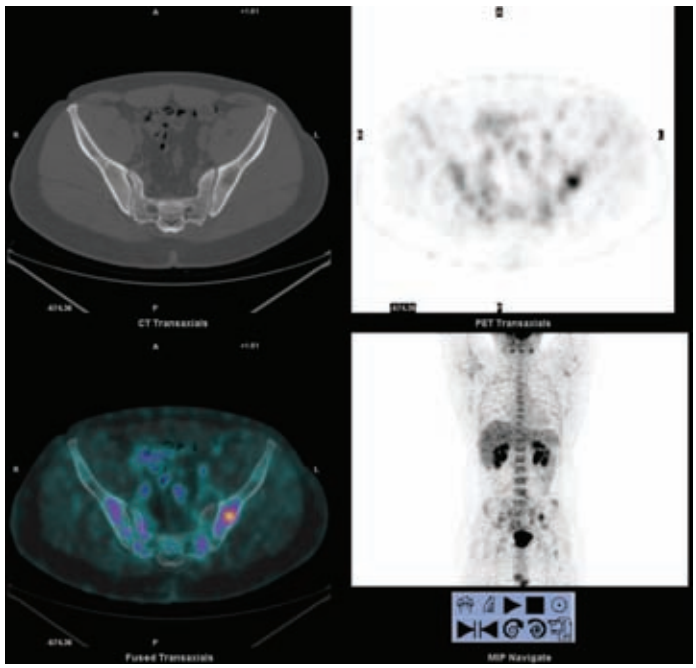
This case also provides an example of the potential utility of PET-CT in following pathologically confirmed treatment response in the adult patient with ARMS. Here, the initial pre-treatment PET-CT

Figure 4: Baseline Staging Axial Positron Emission Tomography–Computed Tomography



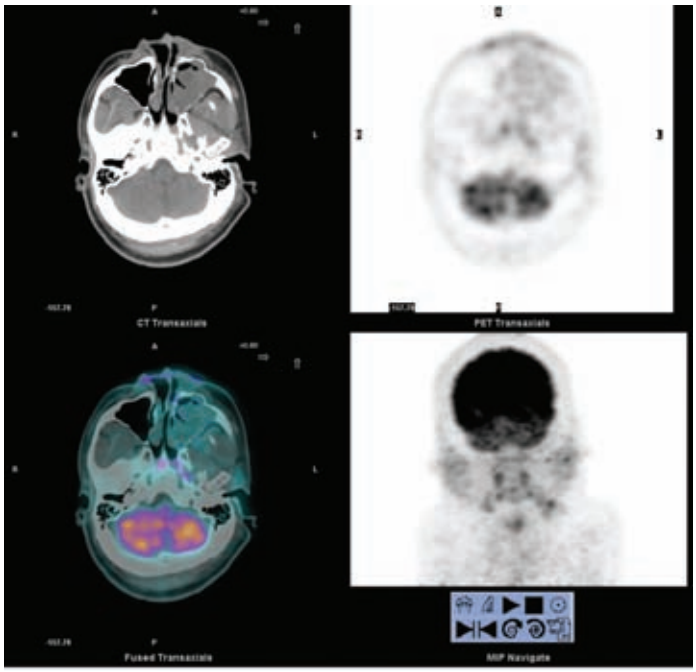
These images show that the primary neoplasm within the left maxillary sinus is F18-FDG avid.

Figure 5: Baseline Staging Axial Positron Emission Tomography–Computed Tomography



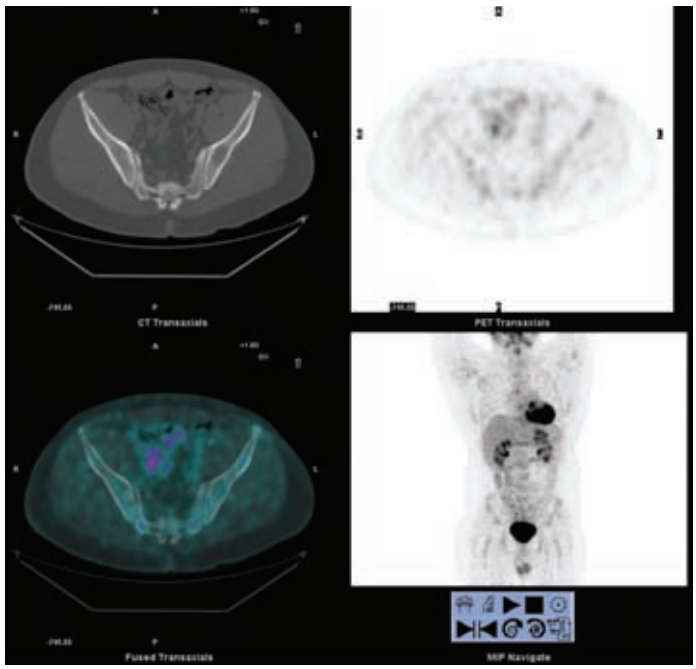
These images demonstrate a left iliac bone osteolytic lesion with F18-FDG avidity. Subsequent biopsy showed metastatic rhabdomyosarcoma.

Figure 6: Positron Emission Tomography–Computed Tomography Four Weeks After the Initiation of Chemotherapy



These images show a significant decrease in metabolic activity of the primary neoplasm.

Figure 7: Positron Emission Tomography–Computed Tomography Images Four Weeks After the Initiation of Chemotherapy



These images show a significant decrease in metabolic activity of the left iliac bone osteolytic lesion, suggesting a favorable early response. Biopsy showed no residual neoplastic tissue.

demonstrated multiple foci of moderate to intense hypermetabolic activity, with imaging four weeks post-chemotherapy revealing a significant decrease in uptake of the primary lesion and bony metastasis. The early post-treatment PET-CT therefore supports the notion of F18-FDG PET-CT documenting early favorable metabolic response to chemotherapy in the adult patient with ARMS. Favorable

imaging response to treatment of rhabdomyosarcoma has been documented in the literature in retrospective studies of pediatric patients.[5,6] Although the imaging response to treatment has not been studied extensively in adult patients, the biologic behavior of rhabdomyosarcoma in the young adult has demonstrated similarity to pediatric cases in one retrospective series.[2]

Conclusion

In conclusion, this case report illustrates the potential of F18-FDG PET-CT in monitoring early response to chemotherapy. Findings of this case should prompt further investigation of the utility of PET-CT in predicting outcome and prognosis in adult patients with head/neck rhabdomyosarcoma following treatment. ■

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