

An Infrequent Case of Adult Alveolar Rhabdomyosarcoma with Pancreatic Metastases Detected in F-18 FDG PET/CT

Abstract

Rhabdomyosarcoma (RMS) is a rare cancer in adults, comprising about 1% of all forms of adult cancer. Common sites of metastases are lungs, skeletal system, lymph nodes, and brain. Metastases of RMS to pancreas are infrequent. We report a case of adult orbital alveolar RMS with pancreatic metastases detected in F-18-fluorodeoxyglucose-positron emission tomography/computed tomography.

Keywords: Fluorodeoxyglucose, metastases, pancreas, positron emission tomography, rhabdomyosarcoma

Introduction

Rhabdomyosarcoma (RMS) is a rare cancerous tumor in adults. RMS is an aggressive tumor and have a tendency to metastasize frequently. Common sites of metastases are lungs, skeletal system, lymph nodes and brain. Positron-emission tomography (PET) using F-18-fluorodeoxyglucose (FDG) is useful for detection, staging, and monitoring a variety of tumors in adults. We report a case of adult orbital alveolar RMS with pancreatic metastases detected in F-18 FDG PET/CT and review the relevant literature available.

Case Report

A 35-year-old male presented with right orbital swelling and diminution of vision for the past 1 month. Biopsy of the orbital mass [Figure 1] revealed small round malignant cell arranged in nests, surrounded by fibrovascular stroma. On immunohistochemical evaluation, the tumor cells were positive for desmin and myogenin. The histopathological features were consistent with alveolar variant of rhabdomyosarcoma (alveolar rhabdomyosarcoma [ARMS]).^[1] Subsequently, whole-body F-18-fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scan was performed 60 min after intravenous injection of 300 MBq of ¹⁸F-FDG, to assess the extent of disease. Maximum intensity projection image of the F-18-FDG-PET/CT scan [Figure 2] shows abnormal increased tracer uptake in the region

of the right orbit, bilateral cervical region, mid-abdomen (arrow), and mid-lumbar region (block arrow). Fusion images reveal that the uptakes correspond to recurrent primary tumor [Figure 2], cervical nodes, pancreatic deposit [Figure 3], and marrow of L4 vertebrae and right pelvic bone. Considering the aggressive presentation of rhabdomyosarcoma (RMS), these features were suggestive of extensive metastases. It is noteworthy that the pancreatic lesion is inconspicuous in contrast-enhanced CT.

Discussion

Soft tissue sarcomas (STSs) are rare cancers, comprising about 1% of all forms of cancer in the adult population.^[2] RMS is an STS thought to arise from myogenic precursor cells of various anatomical sites. Although RMS is infrequent in adult, it is the most common STS in childhood and adolescence.^[2] The histological subgroups of RMS are ARMS, embryonal RMS, pleomorphic RMS, RMS not otherwise specified with latter, and pleomorphic being frequent in adult patients.^[3] While head and neck is the common site of primary in children, extremity tumors are common in adults.^[4] Adult patients have generally poor prognosis compared to children.^[5] Other important prognostic factors include size of tumor, presence of metastases, histological subtypes, and molecular characteristics. With multimodality treatment, the overall survival of nonmetastatic disease has improved to

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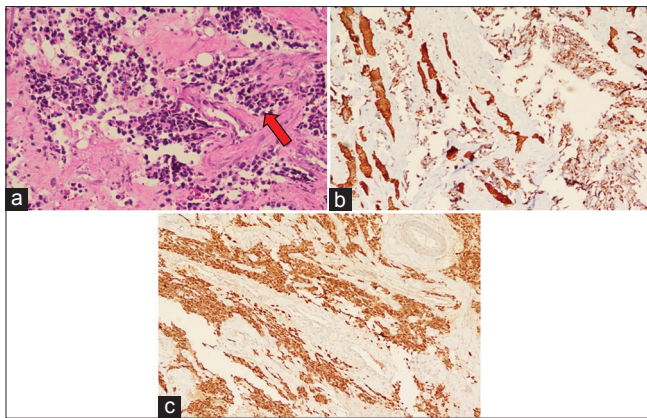


Figure 1: Photomicrograph (Hematoxylin and Eosin stain) (a) of the orbital mass reveals small round malignant cells arranged in nests (arrow), surrounded by fibrovascular stroma. On immunohistochemical evaluation, the tumor cells were positive for desmin (b) and myogenin (c) which are characteristic for rhabdomyosarcoma

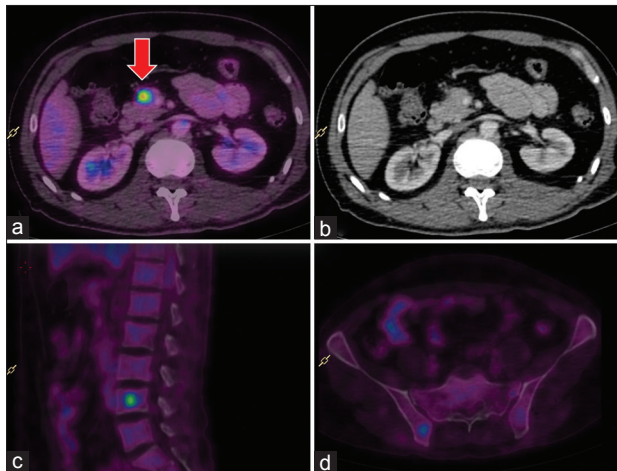


Figure 3: Transaxial view of fused image (a) shows fluorodeoxyglucose avid deposit in the pancreas (arrow), which is inconspicuous in corresponding contrast-enhanced computed tomography image (b). Sagittal view (c) and transaxial view (d) of fused images show focal hypermetabolism in L4 lumbar vertebrae and right iliac bone, respectively. These hypermetabolic foci in aggressive cancer such as rhabdomyosarcoma are suggestive of distant metastases

45%.^[6] Patients with metastatic disease have poorer prognoses and should be considered for novel treatment strategies, thus accurate staging is imperative.

Conventional investigations used for staging and restaging include cross-sectional imaging of the tumor, chest, abdomen, and pelvis; radiolabelled bone scan and bone marrow biopsy. A systematic review by Norman *et al.* concluded that FDG-PET/CT performed consistently better than conventional imaging in initial staging and restaging and has a potential role in assessing treatment response.^[7]

Common sites of metastases reported by conventional investigations are lungs, skeletal system, lymph nodes, and brain, with hematogenous being common route of metastases.^[8] However, anecdotal evidence of unusual metastases such as the breast, testes, subcutaneous

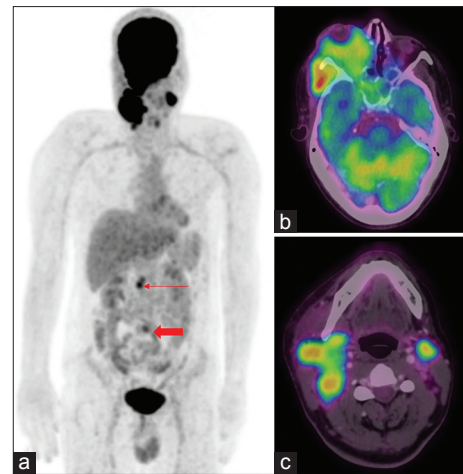


Figure 2: The maximum intensity projection image (a) of whole-body fluorodeoxyglucose positron emission tomography reveals increased tracer uptake in right orbital region, bilateral cervical region, mid-abdomen (arrow), and mid-lumbar region (block arrow). Transaxial-fused images reveal hypermetabolic right orbital mass (b) and metastatic bilateral cervical nodes (c)

tissue, and pancreas are not infrequent.^[9] An autopsy report of 57 children who died from ARMS had reported the prevalence of pancreatic metastases in 67% of patients.^[10] Jha *et al.* in a retrospective analysis reported the high prevalence of 11.2% for pancreatic metastases in ARMS.^[11]

While increased blood supply to the developing breast has been attributed to breast metastases in adolescents, cause of pancreatic metastases is poorly understood. Influence of chromosomal translocation (PAX3/7-FKHR fusion gene, characteristic for ARMS) over insulin-like growth receptors of the pancreas has been hypothesized for the cause of pancreatic metastases.^[11]

Conclusion

FDG-PET/CT has superior diagnostic accuracy over conventional imaging for the evaluation of distant metastases of RMS. Pancreatic metastases of RMS though infrequent are not as rare as initially thought to be especially in alveolar variant.

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Conflicts of interest

There are no conflicts of interest.

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