Alveolar Rhabdomyosarcoma

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History
Teenager with enlarging perianal mass.

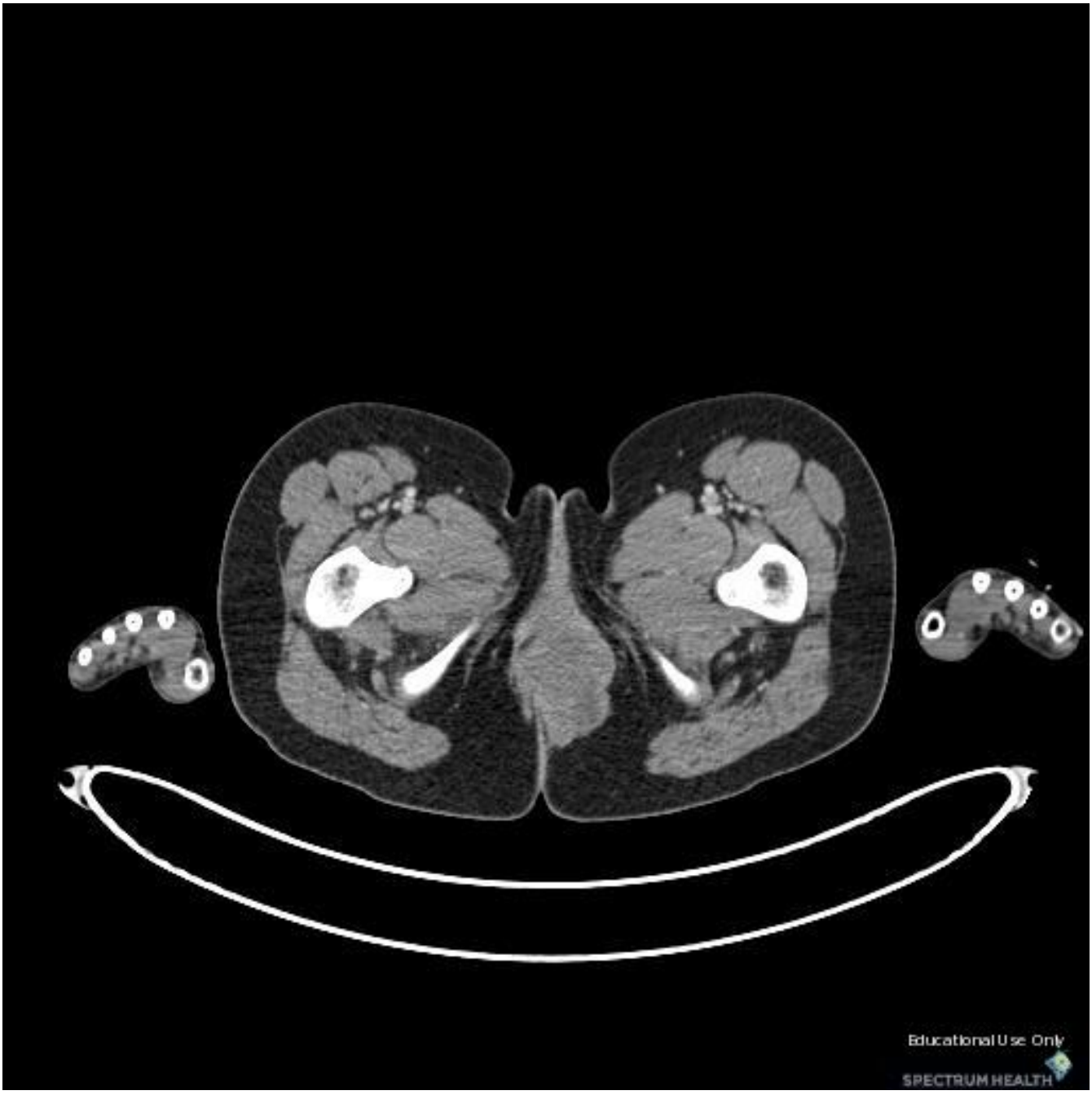
Diagnosis
Alveolar Rhabdomyosarcoma

Discussion
Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood. Its clinical, biological and pathological characteristics vary considerably. Rhabdomyosarcoma is almost exclusively a pediatric malignancy; the adult counterpart is malignant fibrous histiocytoma. Occurrence is bimodal, with are two age peaks: 2 – 6 years (usually head and neck and genitourinary tract sites) and 15 – 19 years (usually extremity, trunk, and paratesticular sites).
Rhabdomyosarcoma derives from primitive mesenchymal cells which demonstrate muscle differentiation on histological, immunochemical or electron microscopic evaluation. There are four histological types in decreasing order of frequency: embryonal, alveolar, undifferentiated and botryoidal. Each of these types may predict the location and prognosis. The embryonal type predominantly involves the head and neck and the genitourinary tract and has an intermediate prognosis. The undifferentiated and alveolar types tend to affect the extremities and have a poor prognosis. The botryoid type usually involves the urinary bladder, vagina, middle ear or nasopharynx and has a good prognosis.
The head and neck region account for about 28% rhabdomyosarcomas, whereas the extremities account for 24%, the genitourinary tract accounts for 18%, the trunk accounts for 11% and the orbit accounts for 7%. These tumors grow rapidly and invade adjacent structures and disseminate by lymphatic or hematogenous pathways. Rhabdomyosarcomas, especially of the extremity, trunk and paratesticular locations, present most often as a painless mass.
Rhabdomyosarcoma most commonly spreads to regional lymph nodes. Paratesticular rhabdomyosarcomas often metastasize to regional lymph nodes, while orbital lesions rarely have nodal metastases given few lymphatic channels in this region. Less commonly, there is distant spread to the lung, bone and bone marrow. Treatment failures can either manifest as local or regional recurrence or metastatic disease.
Rhabdomyosarcoma is a curable disease with more than 70% survival at 5 years after the diagnosis. Relapses are uncommon after 5 years although more often with stage III, stage IV disease, or large primary tumors; relapse survival is best with initial low stage and low risk disease. Operative management is the initial treatment of choice although surgery may be limited by the location and extent of tumor and the desire to preserve function. Hyperfractionated radiation or conventional fractionated radiation is employed for local control when surgery cannot ablate the mass or with infiltrative lesions. Chemotherapy (usually vincristine, dactinomycin and cyclophosphamide or ifosfamide and etoposide) regimens are continually being refined. Recurrent disease is treated with combination chemotherapy; intensive chemotherapy followed by autologous bone marrow transplantation is also being investigated.
The Intergroup Rhabdomyosarcoma Study (IRS-IV) pretreatment clinical staging is based on TNM

classification scheme. Imaging studies and surgical findings determine the extent of disease. Further, treatment is prescribed by a postsurgical grouping classification.
Rhabdomyosarcoma and other soft tissue sarcomas have been shown to be FDG avid and consequently PET-CT would be expected to be useful for initial diagnosis and staging as well as in the detection of recurrent disease. On initial staging PET-CT examination, evaluation of size and extension of the primary tumor and the presence of local lymph node spread and metastatic sites is important. On re-staging PET-CT examinations, locoregional recurrence is most common, however pulmonary, osseous and bone marrow metastatic disease needs to be excluded. PET-CT has been successful in assessing the efficacy of neoadjuvant chemotherapy with reduction in avidity of greater that 50% in soft tissue sarcomas associated with increased time to recurrence and improved overall survival.

Findings
PET/CT- CT shows a soft tissue mass which is markedly fluorodeoxyglucose avid with no evidence of metastatic disease.

Reference
Arush B, et. al. Assessing the use of FDG PET in the detection of regional and metastatic nodes in alveolar rhabdomyosarcoma of the extremities. J Ped Hemat/Oncol 2006; 28:440-445.
Klem ML, et. al. PET for staging in rhabdomyosarcoma: An evaluation of PET as a adjunct to current staging tools. J Ped Hemat/Oncol 2007; 29:9-14.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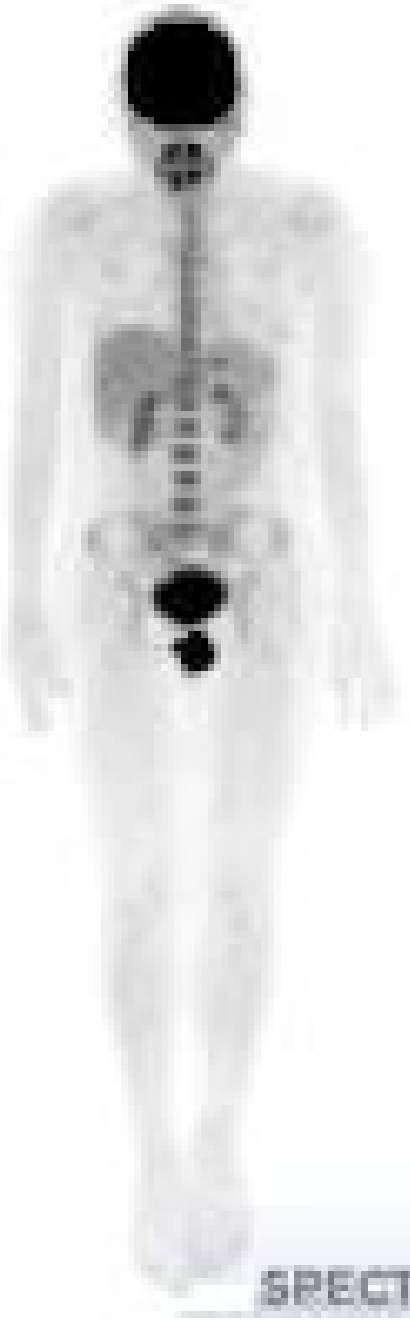

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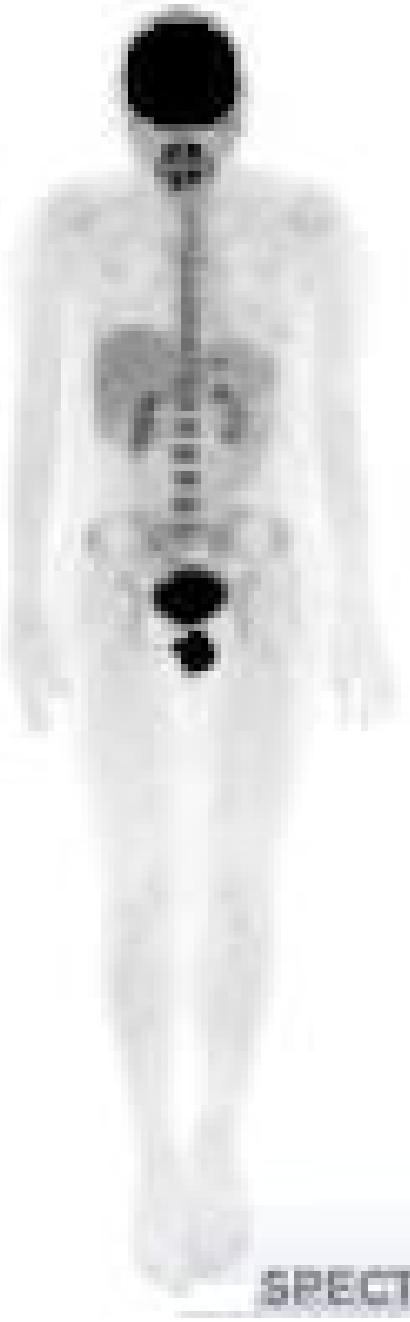

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