J Korean Soc Radiol 2011 ; 64 : 117-121

The CT and MR Imaging Findings of Adulthood Sinonasal Alveolar Rhabdomyosarcoma with Disseminated Metastases on ^{18}F-FDG PET/CT: Report of Two Cases[1]

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We report herein two cases of adulthood sinonasal alveolar rhabdomyosarcoma that showed intense hypermetabolism and disseminated metastasis on ^{18}F-FDG PET/CT. The contrast-enhanced CT (CECT) and gadolinium-enhanced T1-weighted images (Gd-T1WI) showed multiple rings of intense enhancement (the "botryoid sign") of the mass. Adulthood rhabdomyosarcoma should be considered when a sinonasal mass shows the "botryoid sign" on CECT or Gd-T1WI, and intense hypermetabolism with disseminated metastases on 18F-FDG PET/CT.

Index words : Rhabdomyosarcoma, Alveolar
Fluorodeoxyglucose F18
X-Ray Computed Tomography
Magnetic Resonance Imaging

Rhabdomyosarcomas chiefly occur in infants and children and they are far less frequent in adults. Histologically, they are classified into three subtypes: embryonal, alveolar and pleomorphic. While the head and neck are the principal locations for childhood rhabdomyosarcoma, head and neck rhabdomyosarcoma is rare in adults (1). The overall survival is worse for adults than for children (2). The major sites of metastases are lung, bone, bone marrow, liver and kidney.

Several investigators have evaluated the utility of ^{18}F-FDG PET or PET/CT for the assessment of childhood rhabdomyosarcoma at various body sites (3-7). However, there are no reports in the medical literature regarding the ^{18}F-FDG PET/CT findings of adulthood sinonasal rhabdomyosarcoma.

We report herein two cases of adulthood sinonasal alveolar rhabdomyosarcoma that showed intense hypermetaboloism and disseminated metastases on ^{18}F-FDG PET/CT in a 48-year-old woman and a 68-year-old woman, along with the CT and MRI findings.

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Received August 11, 2010 ; Accepted November 24, 2010
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Case Reports

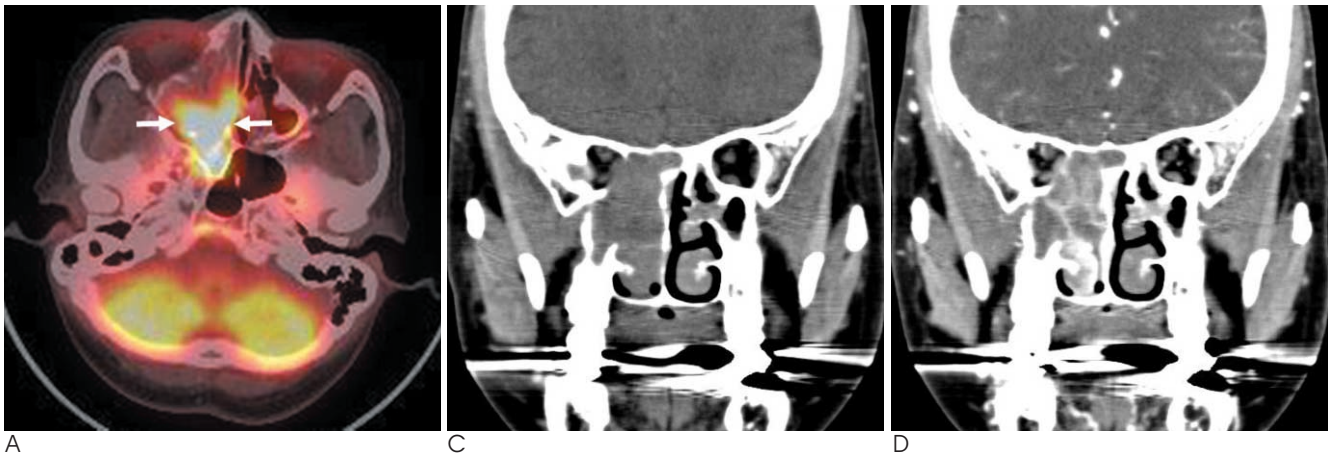
Case 1

A 68-year-old woman presented with the chief complaints of epistaxis and back pain. Physical examination revealed a polypoid mass at the right middle meatus. The microscopic and immunohistochemical findings of the specimen obtained by endoscopic biopsy of the mass were consistent with rhabdomyosarcoma of the alveolar subtype. The ^{18}F-FDG PET/CT obtained four days after biopsy showed a lobular mass with intense hypermetabolism (SUV_{max} = 14.6 g/mL) in the right sinonasal area (Fig. 1A). Also noted were diffuse areas of hypermetabolism involving the axial and appendicular skeleton and the lymph nodes of the right retropharyngeal chain and the left level VA, suggestive of disseminated involvement of the bone marrow and lymph

nodes (Fig. 1B). This was confirmed by biopsy of the bone marrow and a lymph node of the left level VA. Non-enhanced (NECT) and contrast-enhanced CT (CECT) (Figs. 1C, D) and the gadolinium-enhanced T1-weighted images (Gd-T1WI) showed a large mass with multiple rings of intense enhancement (the "botryoid sign") and bony destruction involving the right nasal cavity and the right ethmoid and sphenoid sinuses. She received 4 cycles of CYVADIC chemotherapy and 5 cycles of chemotherapy consisting of ifosfamide, etoposide and vincristine. She experienced subdural hematoma without a history of trauma 12 months after the initial diagnosis of sinonasal alveolar rhabdomyosarcoma. Sadly, in spite of performing craniotomy with removal of the subdural hematoma, she expired 13 months after the initial diagnosis.

Case 2

A 48-year-old woman presented with the chief complaints of left nasal obstruction and bloody discharge for several months. Physical examination disclosed a polypoid mass in the left middle meatus. Pre-biopsy NECT demonstrated a large hyperattenuating mass involving the left nasal cavity, the left ethmoid sinus and the bilateral sphenoid sinuses with bony destruction (Fig. 2A). The microscopic and immunohistochemical findings of the specimen obtained by incisional biopsy of the mass were consistent with rhabdomyosarcoma of the alveolar subtype. The Gd-T1WI obtained seven days after biopsy demonstrated a mass with the "botryoid



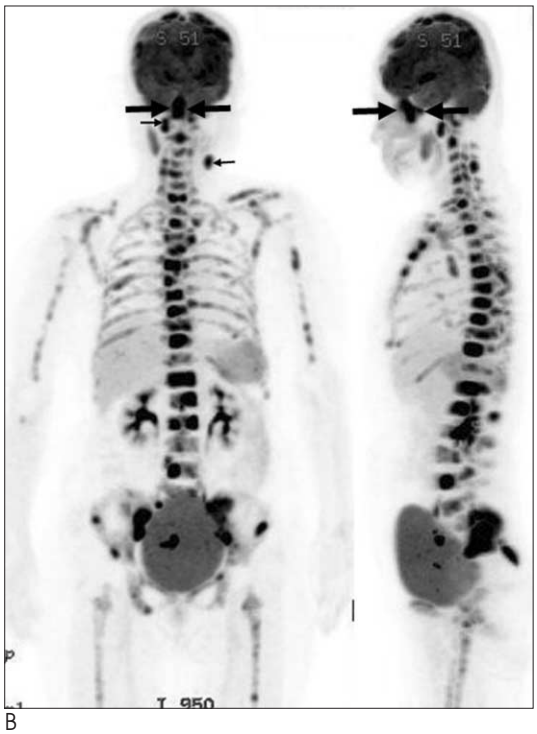


Fig. 1. Sinonasal rhabdomyosarcoma of the alveolar subtype with disseminated metastases to the bone marrow and lymph nodes in a 68-year-old woman.
A. An axial ^{18}F-FDG PET/CT image shows a lobular, intensely hypermetabolic mass (SUV_{max} = 14.6 g/mL) in the right sinonasal area (arrows).
B. The coronal and sagittal maximum intensity projection images of ^{18}F-FDG PET demonstrate intense hypermetabolism involving the axial and appendicular skeleton and the lymph nodes of the right retropharyngeal chain and the left level VA (small arrows), as well as the primary site (arrows).
C. A coronal reformatted non-enhanced CT image shows a right-sided sinonasal mass with bony destruction.
D. A coronal reformatted contrast-enhanced CT image reveals a mass with multiple rings of intense enhancement (the "botryoid sign").

sign" (Figs. 2B, C). ^{18}F-FDG PET/CT performed nine days after the biopsy demonstrated an intensely hypermetabolic mass (SUV_{max} = 11.9 g/mL) in the sinonasal area (Fig. 2D). Also noted were multiple bone marrow metastases in C3-6, T12 and L3, the sacrum, the pelvic bones, the scapula and the femur (Fig. 2E). She received four cycles of chemotherapy consisting of ifosfamide, etoposide and vincristine. A mass developed in her right breast six months after the initial diagnosis of sinonasal alveolar rhabdomyosarcoma, which was proven to be metastatic rhabdomyosarcoma by ultrasound-guided core-needle biopsy. Unfortunately, she experienced progressively aggravating sepsis and then expired 14 months after the initial diagnosis.

Discussion

Sinonasal rhabdomyosarcoma in adults presents a diagnostic challenge because most benign and malignant sinonasal tumors are seen as solidly enhancing mass on CT or MRI, and they cannot be differentiated from rhabdomyosarcoma. Hagiwara et al. (8) noticed that

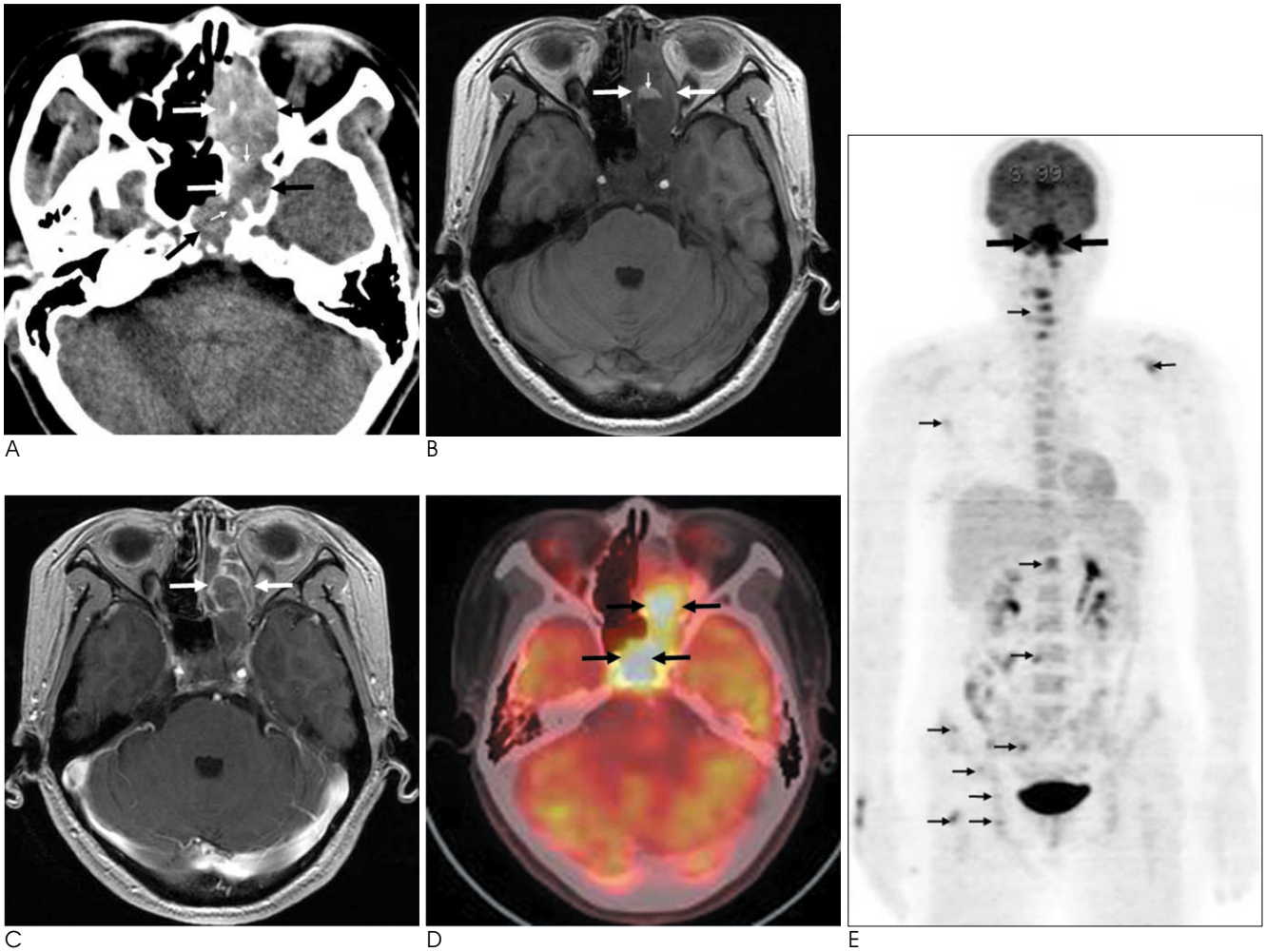


Fig. 2. Sinonasal rhabdomyosarcoma of the alveolar subtype with disseminated bone marrow and breast metastases in a 48-year-old woman.

A. An axial non-enhanced CT image shows a large hyperattenuating mass (arrows) in the left nasal cavity, the ethmoid sinus and the bilateral sphenoid sinuses with bony destruction (small arrows).

B. An axial spoiled gradient-echo T1-weighted image (T1WI) (TR/TE = 7.3/3.0, FA = 20) reveals a sinonasal mass (arrows), which is hypointense compared to that of the adjacent muscles, and the mass contains a focus of hyperintense hemorrhage (small arrow).

C. An axial gadolinium-enhanced spoiled gradient-echo T1WI (TR/TE = 7.3/3.0, FA = 20) demonstrates a mass with multiple rings of intense enhancement (the "botryoid sign") (arrows).

D. An axial ^{18}F-FDG PET/CT image shows a lobular intensely hypermetabolic mass (SUV_{max} = 11.9 g/mL) in the sinonasal area (arrows).

E. A coronal maximum intensity projection image of ^{18}F-FDG PET demonstrates multiple bone marrow metastases involving the axial and appendicular skeleton (small arrows), as well as the primary site (arrows).

four of eight cases of rhabdomyosarcoma of the head and neck showed multiple rings of enhancement that resembled a bunch of grapes (the "botryoid sign", on Gd-T1WI. The "botryoid sign" was identified in our two cases on both CECT and Gd-T1WI. The "botryoid sign" is known to be caused by abundant mucoid stroma surrounded by a thin layer of tumor cells (9). Several investigators have reported the SUV_{max} of rhabdomyosarcoma at various sites of the body (3–5). Our two cases of adult alveolar rhabdomyosarcoma showed SUV_{max} of 11.9 g/mL and 14.6 g/mL, respectively, which were far higher than those reported by other studies in which the majority of cases were in the pediatric age group (3–5).

The initial presentation of alveolar rhabdomyosarcoma with concurrent metastases to the bone marrow and lymph nodes mimicking acute leukemia/lymphoma, has rarely been reported in children (6, 7) or adults (10). Our cases represented sinonasal alveolar rhabdomyosarcoma manifesting intense hypermetabolism of the primary site and disseminated bone marrow metastases at the initial presentation, as demonstrated by ^{18}F-FDG PET/CT. Case 1 presented a diagnostic dilemma because the disseminated metastases involved the bone marrow and cervical lymph nodes at the initial presentation and this mimicked malignant lymphoma/leukemia. Case 2 represented adulthood alveolar rhabdomyosarcoma with breast metastasis, which might represent a sign of disseminated disease and so a poor prognosis. Our review of the literature has yielded two cases of alvelolar rhabdomyosarcoma, each in the orbit and foot, respectively, of two 16-year-old girls, and the tumor showed intense hypermetabolism of the primary sites and disseminated bone marrow metastases (6, 7). We hypothesized that the intense hypermetabolism of the primary site might be associated with disseminated metastases. Our cases also suggest the potential usefulness of ^{18}F-FDG PET/CT not only for the detection of the primary site, but also for the staging of adulthood alveolar rhabdomyosarcoma.

In summary, the diagnosis of adulthood rhabdomyosarcoma can be considered when a sinonasal mass demonstrates the "botryoid sign" on CECT or Gd-T1WI, and intense hypermetabolism and disseminated metastases on ^{18}F-FDG PET/CT. Intense hypermetabolism of the primary site may herald disseminated metastases and thus a poor prognosis. Care must be taken to avoid misdiagnosing sinonasal alveolar rhabdomyosarcoma as lymphoma or leukemia.

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J Korean Soc Radiol 2011;64:117-121

대한영상의학회지 2011;64:117-121

미만성 전이를 보인 성인 비부비동 폐포형 횡문근육종의 ^{18}F-FDG PET/CT, CT 및 MR 영상소견: 2예 보고[1]

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반 영 은 · 이 상 권

저자들은 ^{18}F-FDG PET/CT에서 강한 대사활성도와 미만성 전이를 보였던 성인에서 발병한 비부비동 폐포형 횡문근육종 2예를 보고하고자 한다. 이들 종양들은 조영증강 후 CT 및 T1-강조영상에서 포도송이 모양의 테두리형 조영증강을 보였다("botryoid sign"). 비부비동 종괴가 조영증강 후 CT 및 T1-강조영상에서 "botryoid sign"을 보이며, ^{18}F-FDG PET/CT에서 강한 대사활성도 및 미만성 전이를 동반할 경우 성인에서 발병한 횡문근육종을 고려하여야 한다.