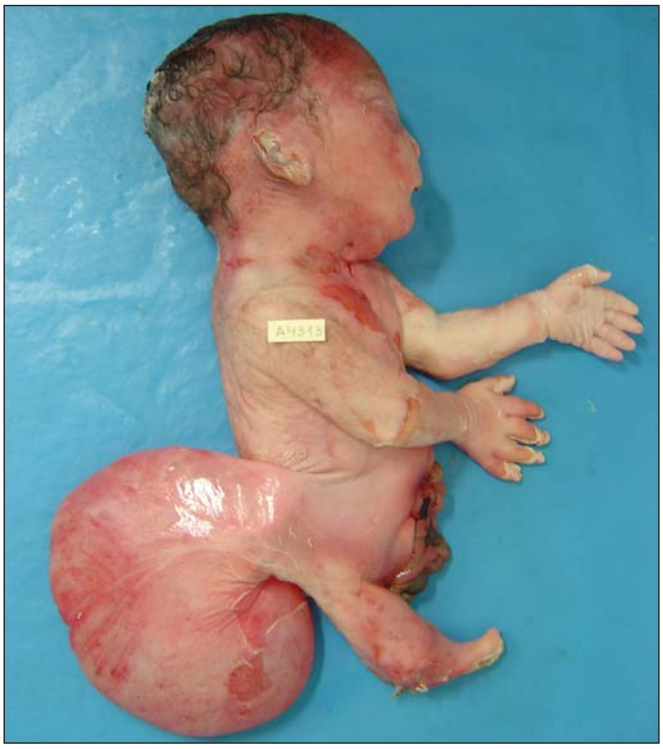

Figure 2. Postmortem evaluation of a 29-week stillborn fetus with sirenomelia.

congenital anomalies, such as congenital heart disease and gastrointestinal anomalies[1]. The prenatal diagnosis is based on identification of this pattern of malformation in imaging studies.

Sirenomelia is considered a primary developmental field defect affecting multiple midline primordia[3]. In the case reported here, MRI allowed us to make the diagnosis of myelomeningocele, which was identified as cystic hygroma on prenatal ultrasound, and bilateral renal agenesis, thereby confirming severe fetal impairment, which allowed the termination of pregnancy to be authorized. However, not all of the associated malformations were identified prior to the stillbirth; the interventricular communication and gastroschisis were identified only during the autopsy. Congenital heart disease has been associated with sirenomelia[1,4], and the fetus evaluated here was also exposed to angiotensin-converting enzyme inhibitors, which could also explain the occurrence of the cardiac defect[5].

The combination of interventricular communication and gastroschisis is not very common; in fact, only two cases, both identified by prenatal ultrasound, have been reported[6]. In a recent review, Feldkamp et al.[7] suggested that gastroschisis is a primary malformation. Our case showed the importance of using a combination of different imaging methods for the diagnosis of a rare congenital anomaly. Although ultrasound continues to be the main diagnostic tool for use during pregnancy, MRI has many advantages, mainly in identifying the fetal morphology[8]. In the case presented here, despite the high quality of the images, the associated malformations were identified only through pathological studies. The unusual anomalies identified in this case were defects of blastogenesis. The combination of prenatal imaging and postnatal autopsy is important to defining the spectrum of associated malformations even when the congenital anomaly is part of a primary developmental field defect.

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Complementary findings on ^{18}F-FDG PET/CT and ^{18}F-NaF PET/CT in a patient with Erdheim-Chester disease

Dear Editor,

A 27-year-old male presented with polydipsia, polyuria, xerostomia, and mild bone pain, being diagnosed with and treated for diabetes insipidus. Thereafter, he presented with diffuse and severe bone pain, xanthomas, xanthelasmas, exophthalmia, and cholelithiasis. After a complete medical investigation, Erdheim-Chester disease (non-Langerhans cell histiocytosis) was considered the most probable clinical diagnosis. Among the imaging exams performed, he was referred for ^{18}F-FDG PET/CT and ^{18}F-NaF PET/CT.

The initial ^{18}F-NaF PET/CT showed that ^{18}F-NaF uptake was more intense in the distal femora and throughout the tibiae, as well as in the fibulae (proximal and distal), tarsi, and maxillas, than in the other bones (Figure 1A). The ^{18}F-FDG PET/CT study revealed increased glycolytic metabolism in the pituitary stalk, proximal left femur, proximal fibulae, ankle, and feet, less intense uptake being observed in other areas (Figures 1B and 1C). It is of note that the ^{18}F-FDG PET/CT was performed 9 months after the ^{18}F-NaF PET/CT, showing a heterogeneous response of the lesions to the various treatment modalities the patient underwent, and that, over the course of the follow-up, he alternated between periods of clinical stability and disease progression.

Erdheim-Chester disease is systemic, although variable in extent, and bone involvement is quite typical. Classical radiological findings include sclerotic and osteolytic lesions in the cortical layer of long bones, occurring bilaterally and symmetrically in their metaphysis and diaphysis, sparing the epiphysis and the axial skeleton. Approximately 50% of patients with Erdheim-Chester disease present extraosseous impairment, including changes in the

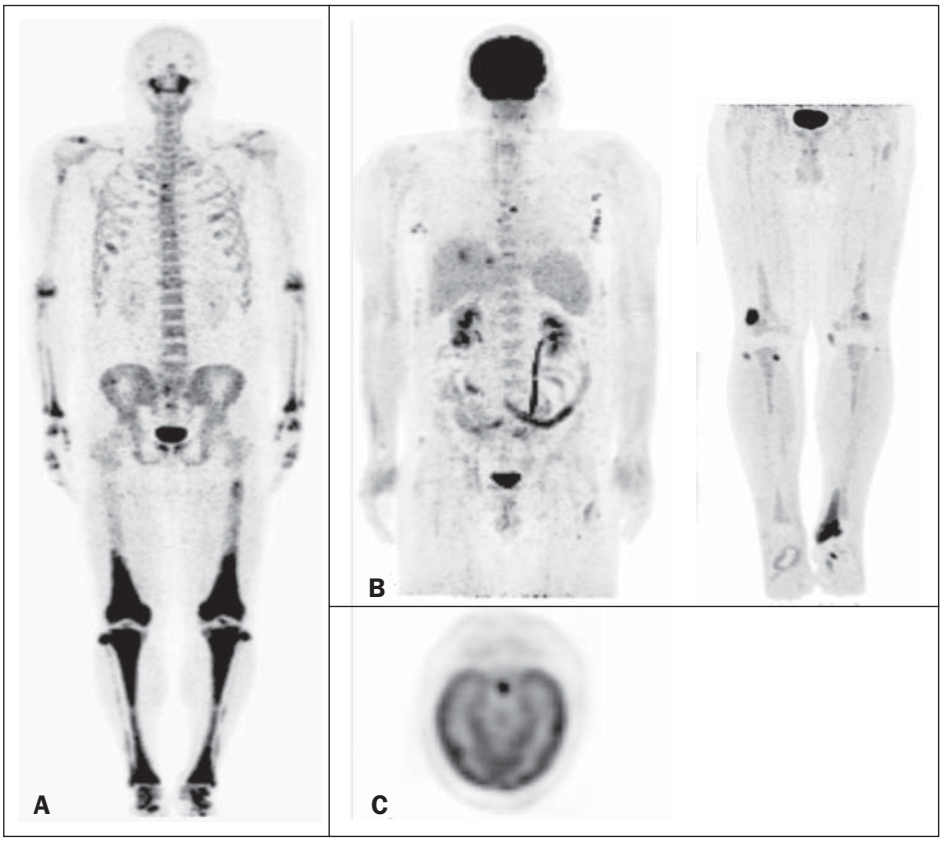

Figure 1. A: ^{18}F-NaF PET/CT maximum-intensity projection image showing intense NaF uptake in the distal third of the femora; throughout the tibiae; in the proximal and distal extremities of the fibulae; in the tarsi; and in the maxillas. Note also the uptake in the proximal third of the right humerus, proximal diaphysis of the left femur, acromioclavicular joints, pubis, elbows, joints of the hands, and thoracic girdle. **B:** ^{18}F-FDG PET/CT maximum-intensity projection images depicting diffuse nodular lesions in the thoracic and abdominal walls; distal metaphyseal region of the femora and tibiae; left femoral diaphysis; left Achilles tendon; and feet—the imaging criteria indicating disease progression in comparison with the findings of previous exams (not shown). **C:** PET axial brain image demonstrating high FDG uptake in the hypophysis, corresponding with the nodular thickening of the pituitary stalk seen on an MRI scan (not shown).

hypothalamus, posterior hypophysis, eyes, retroperitoneum, skin, lungs, and heart[1].

^{18}F-NaF PET/CT has the advantage of being a whole-body study with high sensitivity, thereby detecting bone impairment in Erdheim-Chester disease. The use of imaging methods enables clinical suspicion for early diagnosis and patient follow-up, including therapy response assessment[2]. In comparison with ^{99m}Tc-MDP, ^{18}F-NaF shows better pharmacokinetic characteristics, including faster blood clearance and two-fold higher uptake in bone[3]. Data from a number of studies, all involving small patient samples, have shown that ^{18}F-NaF PET has higher sensitivity and specificity than do conventional ^{99m}Tc-based bone scans[4–7]. In the present study, ^{18}F-NaF PET/CT revealed some bone lesions in the ribs and arms that were not detected by ^{18}F-FDG PET/CT, indicating that the former has greater sensitivity for detecting bone lesions.

In Erdheim-Chester disease, extraosseous impairment can occur in almost every organ, which suggests that ^{18}F-FDG PET/CT has potential value as a diagnostic tool. However, its main advantage is probably therapy response assessment, although that has not been well established[8]. This imaging modality also allows guided percutaneous biopsies (by identifying areas of high metabolic activity). Therefore, the role of ^{18}F-FDG PET/CT in the initial diagnosis of Erdheim-Chester disease remains unclear, especially because the systemic presentation patterns of the disease are extremely variable, and it is likely to prove much more valuable for patient follow-up[8,9].

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