malignancy. Isolated EMPD, without a coexisting internal primary lesion, is usually an indolent, slow-growing cancer that rarely metastasizes. Rare invasive EMPD has a propensity to metastasize to inguinal nodal basins[5]. EMPD involving the external genitalia has a strong association with gastrointestinal and genitourinary adenocarcinomas[4,6]. A small subset of invasive EMPD cases show signet-ring cell morphology with extracellular mucin. Immunohistochemical analysis establishes the distinction between signet-ring cells intrinsic to EMPD and those originating from coexisting visceral neoplasms[7,8]. Poor prognostic factors include dermal invasion, nodular skin lesions, lymph node involvement, and distant metastasis[3]. Given the multiple presentations of EMPD and their varying prognoses, there is a need to identify distant metastases and the primary visceral tumor: that effort is facilitated by functional ^{18}F-FDG PET/CT imaging[1–6].

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Congenital lobar emphysema

Dear Editor,

A 34-year-old asymptomatic woman underwent a chest radiography examination as an admission requirement for a new job. The X-ray showed focal hyperlucency in the left upper lobe of the lung (Figure 1A). High-resolution computed tomography (HRCT) was performed to confirm the findings (Figures 1B and 1C). The HRCT findings were characteristic of congenital lobar emphysema (CLE).

The evaluation by imaging methods in pediatrics has been the subject of a series of recent publications in the radiology literature of Brazil[1–6]. CLE is characterized by hyperinflation of one or more lung lobes in the absence of extrinsic bronchial obstruction[7]. It is a rare disease and its incidence is 20–30 cases/1000 births, most commonly affecting a single lobe of the lung (typically the left upper lobe), although multiple lobes or specific lobar segments may be involved[7,8]. The disease has a variety of causes, including bronchial cartilage deficiency (bronchomalacia) and endobronchial lesions, resulting in narrowing of the airway lumen and obstruction with air trapping, as well as progressive lobar overexpansion, usually with compression of the remaining areas of the ipsilateral lung[9].

CLE is generally diagnosed during early infancy, presenting with persistent progressive respiratory distress. It is known that CLE can occur in association with other malformations, especially cardiac malformations, which are present in 20% of cases[7]. In rare cases, it is diagnosed in adulthood and must be differentiated from other causes of localized pulmonary hyperlucency, because the treatments differ[9]. In such cases, the patients are usually asymptomatic and the disease can go unnoticed, resulting in underestimation of the true incidence of this condition.

Conventional chest X-rays are typically used in order to establish the diagnosis of CLE, showing a unilateral hyperlucent hemithorax. This finding is also present in a variety of other conditions, which include tension pneumothorax—the main differential diagnosis on routine chest radiography[7]—as well as
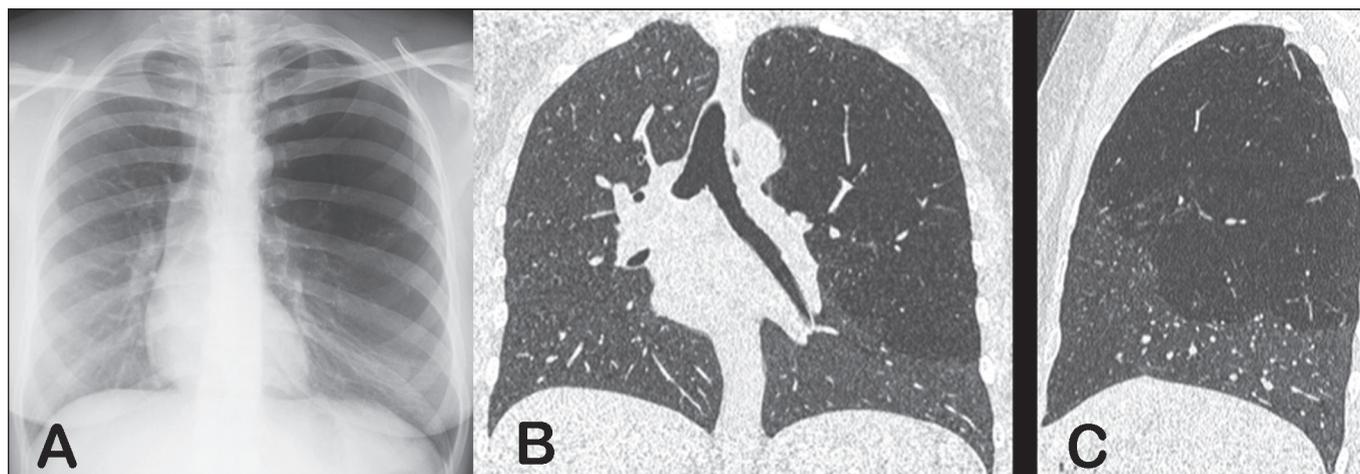

Figure 1. Anteroposterior chest X-ray (**A**) showing radiolucency and hyperinflation of the upper two thirds of the left lung. HRCT with coronal and sagittal reconstructions (**B** and **C**, respectively) showing hyperinflation of the left upper lobe of the lung, as well as vessel attenuation.

bullous disease, pneumatocele, Swyer-James syndrome, endobronchial mass, unilateral pulmonary agenesis, proximal interruption of the pulmonary artery, scimitar syndrome, diaphragmatic hernia, and Poland syndrome[8]. It can also exclude an intrathoracic mass or vascular ring. HRCT is useful for confirming radiographic findings, delineating the affected lobe and showing relative narrowing of the bronchus associated with hyperinflation and attenuated vessels in the hyperlucent lobe, which facilitate the differential diagnosis.

Lobectomy is the treatment for nearly all cases of CLE with respiratory distress. According to Karnak et al.[10], lobectomy is the recommended treatment for CLE in all infants under two months of age and in older infants who present with severe respiratory symptoms. Apparently, the earlier the presentation is, the greater is the need for surgery. Conservative management, with close outpatient follow-up, can be used in older children who present with mild to moderate symptoms. Because our patient had remained asymptomatic throughout her life, her case was managed with clinical and radiographic follow-up.

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Erdheim-Chester disease with isolated neurological involvement

Dear Editor,

A 25-year-old female patient presented with a seven-month history of progressive dysphagia, dysphonia, diplopia, ptosis of the right eyelid, weight loss, and sporadic pulsatile headache on the right side of the face. She had a history of hypertension, diabetes, unspecified thyroid disease, and smoking. The physical examination revealed satisfactory general health, although the patient was found to be malnourished, as well as to have deficits in the right third, fifth, and sixth cranial nerves. Magnetic resonance imaging (Figure 1) showed an expansile lesion located in the right sellar and juxtasellar region. A transsphenoidal biopsy was performed. The pathology and immunohistochemical study showed xanthomatous macrophages, together with CD 68 positive and CD1A negative histiocytes, consistent with a diagnosis of Erdheim-Chester disease. Computed tomography of the chest and abdomen showed no abnormalities.

Erdheim Chester disease is currently considered a clonal disorder, the pathogenesis of which is mediated primarily by a chronic, uncontrolled inflammatory process[1]. The Th1-type immune response involves activation of the following cytokines: IFN- , IL-1/IL-1Ra, IL-6, IL-12, and MCP-1/CCL2. In studies of Erdheim-Chester disease, the most commonly reported gene mutation is that occurring in the BRAF V600E gene, which is seen in 57–75% of patients diagnosed with the disease. Mutations have also been reported in the MAPK (NRAS and MAP2K1) and PIK3 (PIK3CA) pathways[2].

Histopathologically, Erdheim-Chester disease is a non-Langerhans cell histiocytosis, characterized by numerous macro-
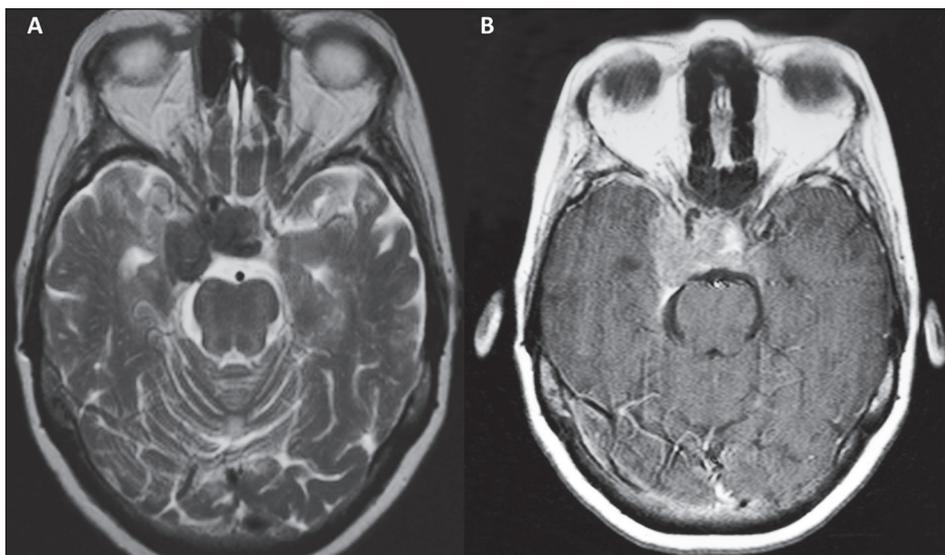

Figure 1. Magnetic resonance imaging scan showing a well-defined, compact, lobulated expansile lesion, measuring 3.0 × 1.5 × 3.0 cm, located in the right sellar and juxtasellar region, invading and occupying the sella turcica and the right juxtasellar region, with a hypointense signal on a T2-weighted image (**A**) and intense enhancement on a T1-weighted image acquired after gadolinium contrast administration (**B**). Note the reduction in the caliber of the intracavernous carotid arteries. The lesion is compressing and dislocating the optic chiasm anteriorly, cavitating the right medial temporal region, surrounding the pituitary gland, and extending to the suprasellar cistern. Anteriorly, it reaches the right optic canal.