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Two smooth muscle tumors in the airway of an HIV-infected child

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Abstract. Tumors of smooth muscle origin are rare in childhood. We report a case of multiple bronchial leiomyomata in a seven year old girl with clinical HIV infection who presented with new onset of wheezing. Clinical details of this case have been published elsewhere, but without imaging studies [1].

We know of eight previously reported cases of smooth muscle tumors in HIV-infected children, suggesting a non-random association. These cases have primarily involved the bronchi and GI tract. Smooth muscle tumors should be considered in an HIV-infected child with new or unexplained respiratory or GI symptoms.

Case report

A cachectic 7-year-old black female presented to the emergency room with severe respiratory distress, wheezing and a temperature of 103 degrees. She had been diagnosed as HIV positive at the age of two and her mother was a known intravenous drug abuser and HIV positive. Past medical history revealed many admissions for sinusitis, pneumonias, myocarditis and AIDS encephalopathy. Medications on admission included AZT, Bactrim, Digoxin, and Nystatin as well as IV gamma globulin. There was a severe depletion of CD4 helper cells as well as a markedly decreased CD4/CD8 ratio.

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Initial chest X-ray revealed a mild reticulonodular pattern throughout both lung fields unchanged from prior films of earlier admissions. A soft tissue mass (Fig. 1) bulged into the tracheal lumen from the upper left wall. CT (Fig. 2) confirmed an 8 × 5 mm mass in the upper left tracheal wall impinging upon the lumen. The patient had a history of intubation and this was thought to possibly represent a granuloma. Despite broad spectrum antibiotics, the patient continued to spike fevers with several episodes of severe respiratory distress. Bronchoscopy was performed with excisional biopsy of the tracheal lesion. Pathology revealed a 1 cm papillary mass with immunochemistry studies positive for desmin, specific for smooth muscle origin, representing a leiomyoma. The patient did well for a week post surgery when she began to have new episodes of cyanosis and respiratory distress. Chest X-ray (Fig. 3) now revealed a mass in the distal left mainstem bronchus with air trapping in the left lung. The left upper lobe was hyperinflated (Fig. 4) with narrowing of the left main stem bronchus and non-visualization of the left upper lobe bronchi and lingular bronchi. Multiple pulmonary nodules were noted as well as a left lower lobe infiltrate. At repeat bronchoscopy a mass arising from the left upper lobe bronchus was identified and biopsied. The pathology revealed a second smooth muscle tumor, with possible sarcomatous features. Despite attempted laser resection of the masses the patient continued to have respiratory difficulty and left lower lobe atelectasis and to require supplemental oxygen. She died 5 months later of cardiorepiratory arrest.

Discussion

The association of neoplasms with congenital and acquired immunodeficient states is well known. In particular, HIV-infected adults have a high incidence of aggressive B cell lymphoma and Kaposi sarcoma as well as malignant tumors of the head and neck, colon, and anorectal areas [2]. The occurrence of neoplasms in HIV-infected children, while well known, is considerably less common than in adults, being seen in less than 2% of such children [3].

Benign and malignant smooth muscle tumors are rare in childhood; leiomyosarcomas constitute less than 2% of childhood soft tissue sarcomas [3]. Scattered previous reports of benign and malignant smooth muscle tumors in children have included a few non-HIV immunocompromised patients (two with renal transplants, one with ALL in remission) as well as eight cases in HIV infected children (four mentioned as a personal communication) [3]. Most of these patients have had multiple tumors, involving the tracheobronchial tree and/or the GI tract.

A number of possible mechanisms for the association HIV and leiomyomatous tumors have been proposed. These include defective immuno-surveillance, chronic antigenic stimulation, the presence of multiple coinfecting organisms and elaboration by HIV-infected T cells of growth factors which stimulate tumors [2]. In addition, the HIV virus itself may have a direct role in tumorigenesis, as has been suggested by the work of Vogel and colleagues on the induction of Kaposi-like spindle cell tumors in transgenic mice containing an HIV gene in their genome [2]. The relative contributions of the immunodeficient

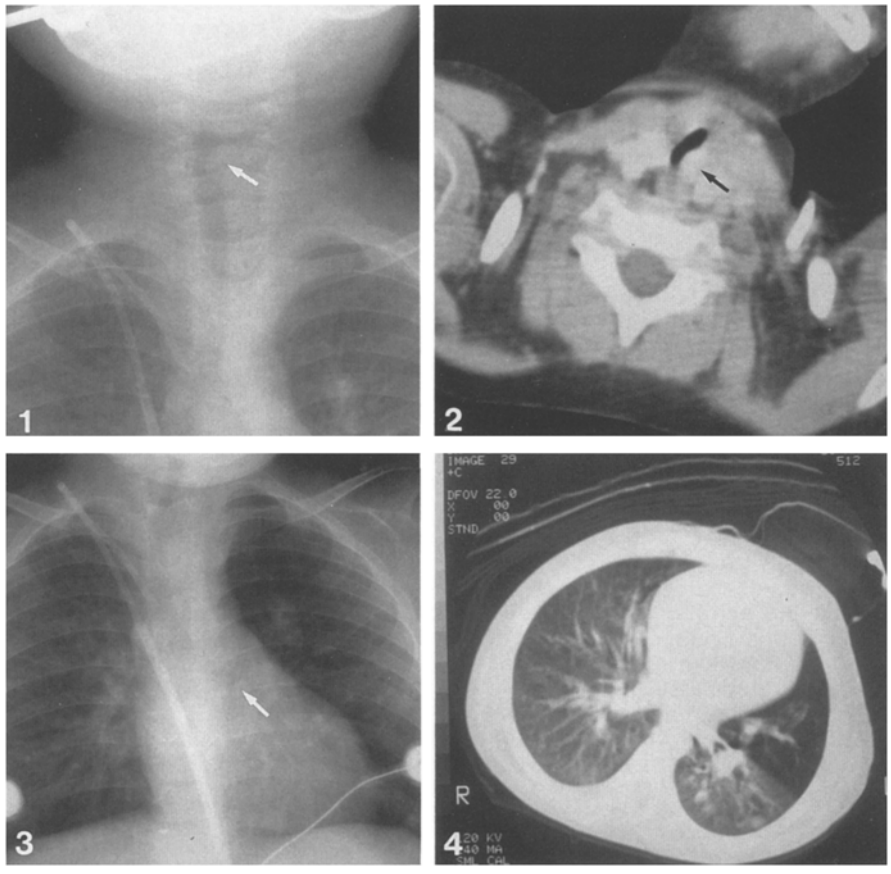

Fig. 1. AP view of upper trachea showing nodular mass *(arrow)* encroaching upon the airway

Fig. 2. CT cut of neck with contrast at same level as Fig. 1 shows mass *(arrow)* deforming and narrowing the trachea

Fig. 3. AP chest shows hyperlucency of the left upper lung indicating air trapping, as well as a nodular mass *(arrow)* in the distal left mainstem bronchus

Fig. 4. CT cut in midthoracic area showing hyperlucency and oligemia in the left upper lobe, representing obstructive emphysema

state, of humoral factors, and of direct viral oncogenesis in HIV patients remain under study [4]. In addition, desmin positive pseudo-spindle cell tumors resembling leiomyomas have been seen in patients with HIV and underlying mycobacterium avium intracellulare infection, actually representing a hyperplastic process. However, no MAI organisms were seen in this patient and the lesions were pathologically diagnosed as true tumors [5].

It is easy to overlook the onset of tumors in these chronically ill children because non-neoplastic forms of pathology such as opportunistic infections and liver and biliary dysfunction are much more common than neoplasms in the respiratory and GI tracts of HIV-infected children. Indeed, many children with neoplasms have other concurrent pathology. However, an awareness of the association, albeit uncommon, between HIV infection and smooth muscle tumors will facilitate the early recognition of these tumors in children with HIV who present with new or unexplained symptoms in the tracheobronchial tree or in the GI tract. In the tracheobronchial tree, the symptoms may be obstructive in nature [6]. With the growing pediatric AIDS population and the increase in the length of the illness, the number of such cases is likely to increase.

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Editorial commentary

In the past several years, scattered accounts of leiomyoma and/or leiomyosarcoma have been published in HIV infected pediatric patients who had prior LIP. The tumors have been noted in the liver, gastrointestinal tract but mostly in the lung, both in the trachea, bronchi and lung parenchyma.

The tumor cells are Desmin positive and, on light and electron microscopy, are felt to be of smooth muscle origin. They must be distinguished from the collections of spindle cells that seem to be a pseudotumor in HIV patients with mycobacterium avium intracellulare; the cases of this latter pseudotumor have been in the adult HIV literature.

W. E. Berdon (Managing Editor)