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LETTER TO THE EDITOR
Interferon Alpha for Alveolar Soft Part Sarcoma

To the Editor: Alveolar soft-part sarcoma (ASPS) is a rare malignant tumor for whom chemotherapy has little or no efficacy. Although we reported on some patients responding to conventional chemotherapy, results were ultimately disappointing if the tumor was not completely resected [1]. Thus, new approaches are needed for patients with unresectable ASPS. The possible activity of interferon alpha (IFNα) against ASPS has been previously documented [2]. More recently Roozendaal et al. [3] described a partial remission induced by IFNα 2b in a patient with ASPS. We therefore decided to use IFNα in a patient with a chemoresistant ASPS.

A 7-year old girl presented in September 1996 with a rapidly-growing mass in her right buttock. A biopsy was performed confirming the diagnosis of ASPS. Staging revealed multiple lung metastases. Chemotherapy was administered according to the European MMT4 protocol without benefit. Further chemotherapy with topotecan and cyclophosphamide achieved no improvement and a very slow increase in the number and size of the pulmonary lesions was demonstrated on CT scan. Because the patient was otherwise in very good general condition, in March 1997 we agreed with the family to stop any further treatment, apart from irradiating the buttock lesion to diminish the local discomfort.

Towards the end of 2003, the patient's general condition suddenly deteriorated, with rapid weight loss (from 45 to 37 kg in approximately 1 month) and increasing respiratory insufficiency (oxygen saturation in the range of 91%–93%). New imaging studies showed an increase in the metastatic lesions (Fig. 1a) and revealed an intracerebral mass in the frontal lobe. With the family's and patient's consent, treatment with IFNα 2b (3×10^6 IU s.c. day) was initiated. During the treatment, she suffered further weight loss and fever. Oral steroids (prednisone 25 mg daily) were given to ameliorate the symptoms. The fever disappeared and food intake improved. IFNα treatment was never interrupted and a new radiological evaluation after 3 months showed a further increase in the lung lesions (Fig. 1b). We decided to stop the IFNα therapy and the patient died 2 months later.

We were consequently unable to confirm the encouraging report by Roozendaal et al. [3] and Kuriyama et al. [2]. Although it was demonstrated that most solid tumors of childhood express IFNα receptors, and that IFNα treatment may inhibit their growth in vitro [4], other mechanisms—including immuno-mediated antitumor effects—may be relevant to its in vivo efficacy in ASPS. IFNα may not be effective when the tumor burden is exceedingly high, as seemed the case in our patient by comparison with the imaging studies on the two reported cases in which this treatment was effective, although this is

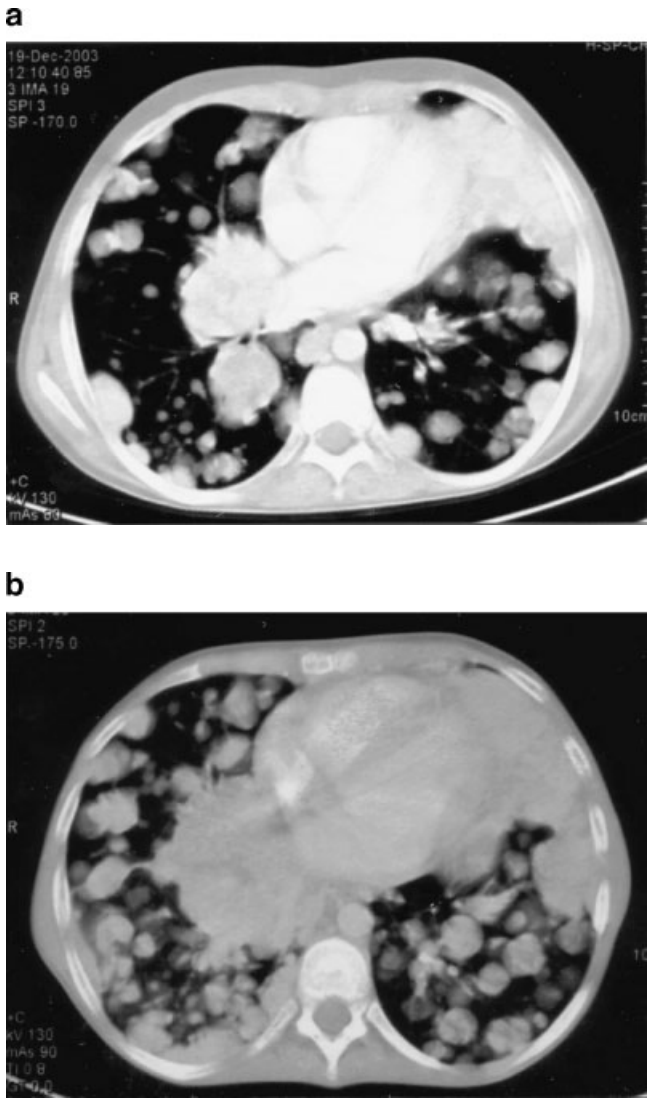

Fig. 1. Progression of lung metastases is evident comparing the axial CT section at the beginning (**a**) and after 3 months (**b**) of IFN alpha 2b therapy.

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highly speculative. The role of IFNα in ASPS deserves further study in order to better define its role and mechanisms of action.

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