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Bronchioloalveolar Carcinoma in a Juvenile Rhadomyosarcoma Patient

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Primary tumors of the lung are uncommon in pediatric patients, particularly bronchioloalveolar carcinoma (BAC). An 11-year-old female suffering from back pain for 1 month was referred to Seoul St. Mary's Hospital for treatment of a pathologic fracture of the lumbar spine. Comprehensive evaluation disclosed numerous pulmonary metastases of rhabdomyosarcoma (stage IV). During chemotherapy, most of the lung lesions regressed, with the exception of two nodules. Wedge resections, intended for diagnosis and cure, yielded a histologic diagnosis of BAC.

Key words: 1. Lung neoplasms
2. Adenocarcinoma, bronchiolo-alveolar
3. Pediatric
4. Rhabdomyosarcoma

CASE REPORT

An 11-year-old female was referred to our care for the treatment of a pathologic fracture of the lumbar spine. One month prior, she had fallen during exercise, causing persistent back pain. When symptoms failed to abate, computed tomography (CT) of the spine was performed, revealing a fracture at the L-2 level that was most likely pathologic. Given the circumstances, she was admitted to Seoul St. Mary's Hospital as a pediatric hematology patient. A chest X-ray (Fig. 1) revealed a left lower lobe infiltrate with pleural effusion, and the chest CT revealed bilateral pleural effusions with multiple, diffuse nodules of both lungs. Positron emission tomography (PET)-CT further disclosed whole-body tumor involvement. Biopsies of the bone marrow and the sole of the right foot confirmed stage IV rhabdomyosarcoma. After 5 months of chemotherapy, a complete metabolic response was evident by PET-CT; and after 11 months of treatment, the lung lesions and pleural effusions had essentially resolved. However, two nodules persisted in the left lung—one in the upper lobe and the other in the superior segment of the lower lobe (Fig. 2B). For diagnostic and curative purposes, wedge resection of these two nodules was performed via mini-thoracotomy. The patient's recovery was uneventful, and she was discharged on postoperative day 6.

Grossly, the resected nodules were firm and ovoid, measuring 3 mm in diameter. In histologic sections, tumor proliferation was lepidic, marked by cuboidal to columnar cells with scanty cytoplasm following alveolar contours. Thus, bronchioloalveolar carcinoma (BAC), non-mucinous type (Fig. 3) was diagnosed. The patient was tumor-free after 6 months, but a new mass developed on her left wrist 10 months

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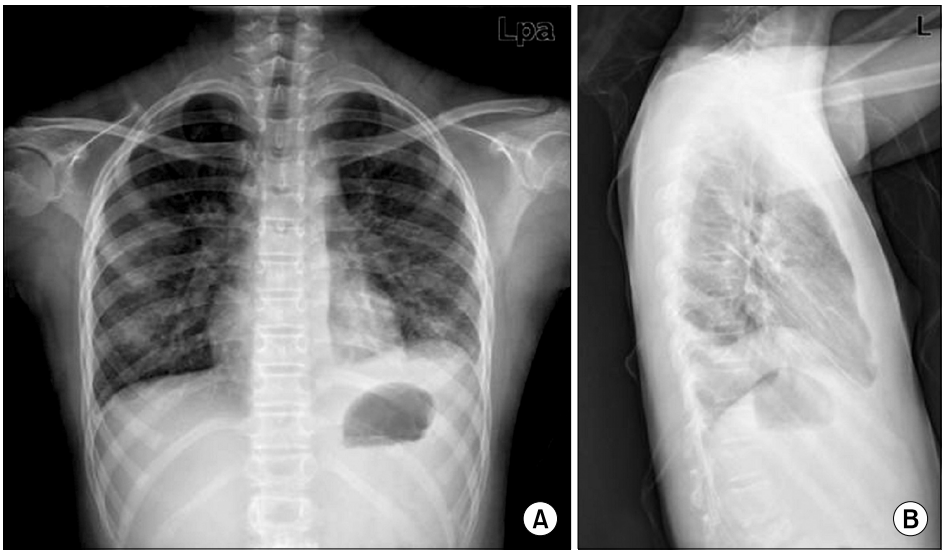

Fig. 1. Initial chest X-ray. (A) Posterioranterior (PA) view: pleural effusion and lower lobe infiltrate on left. (B) Left lateral view: left pleural effusion with thickened fissure, indicative of pleural lesion.

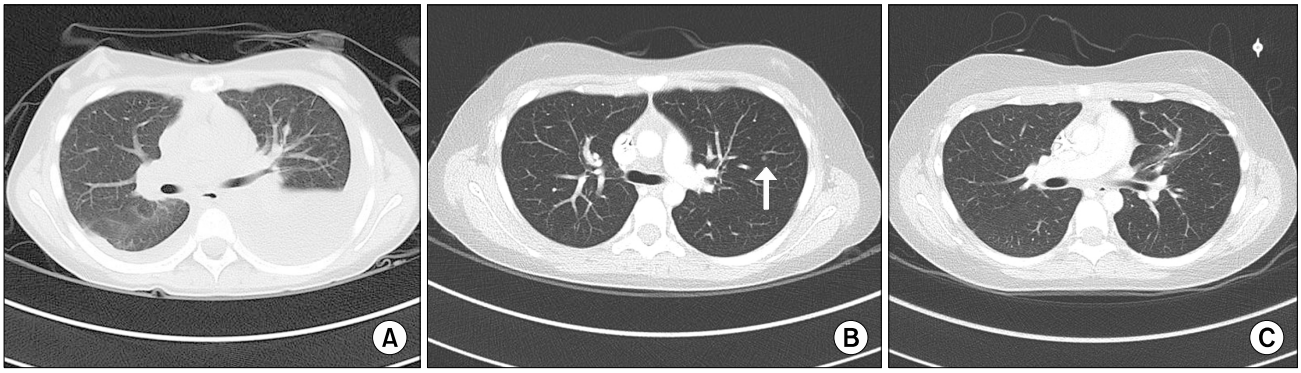

Fig. 2. Serial chest computed tomography. (A) Initial scan: bilateral pleural effusions and multiple small nodules throughout lungs. (B) After chemotherapy (11 months): nodules and pleural effusion absent, with persistent nodule <5 mm (arrow) of left lower lobe. (C) After surgery (13 months): multiple nodules throughout lungs and postoperative change of left lung.

postoperatively. Whole-body recurrence of rhabdomyosarcoma was subsequently verified by PET-CT.

DISCUSSION

Primary lung tumors are rare in the pediatric age group. Solitary pulmonary nodules are largely due to inflammation, but the neoplasms that do occur are generally malignant, with the incidence of metastatic disease outnumbering that of primary lung cancer [1]. Most primary cancers of the lung are bronchogenic in origin, typically arising during the sixth or the seventh decade of life. In the second decade, the reported incidence is only 0.9%, and prior to the age of 40 years, this figure changes little (1.2%) [1]. BAC is a variant of adenocarcinoma. Although its etiology and risk factors are not fully appreciated, smoking and environmental exposures are unlikely contributors [2]. Instead, an association between congenital pulmonary airway malformation (CPAM), type I and BAC has been reported [3], and BAC has been encountered as a second malignancy among survivors of other pediatric malignancies, including Ewing's sarcoma, Hodgkin's lymphoma, hepatoblastoma, and testicular teratocarcinoma [4].

In this instance, there were no signs of CPAM in the preoperative chest X-ray or chest CT, and the lung lesions were not cystic. Shiraishi et al. [5] similarly reported three young patients (14, 17, and 23 years of age) with osteosarcoma and

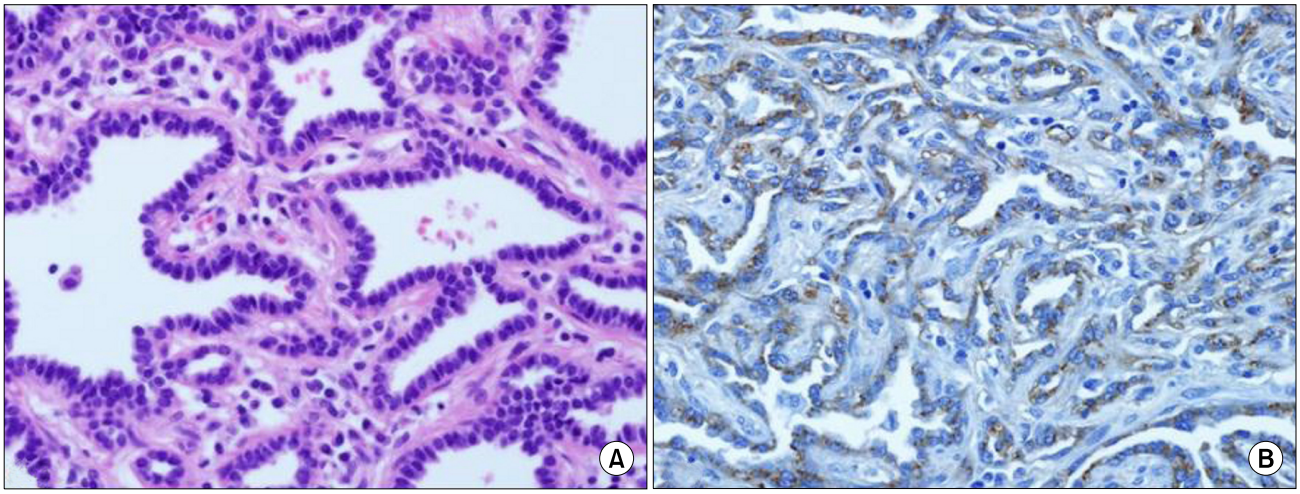

Fig. 3. (A) Cuboidal to columnar cells nondestructively follow alveolar contours (H&E, ×400). (B) Immunohistochemistry (EGFR), histopathology of bronchioloalveolar carcinoma; positive EGFR immunostaining (×400). EGFR, epidermal growth factor receptor.

concurrent malignancy (adenocarcinoma) or premalignancy (atypical adenomatous hyperplasia) of the lung. All lung pathology was proven before the chemotherapy for osteosarcoma, but the significance of this association remains to be seen. Our patient presented with bilateral pleural effusions and boggy lungs, precluding the identification of BAC. Without a definitive timeline, BAC could not legitimately be linked to chemotherapy. Kowalczyk et al. [6] reported that the probability of the second malignancy was greater in pediatric acute lymphoblastic leukemia patients than in the normal group. We believe that soft tissue malignancy could affect the development of the second malignancy. Further studies including genetic predisposition need to be followed.

Pediatric patients with BAC are usually asymptomatic, which hampers discovery and diagnosis. Therefore, our patient was under CT surveillance for disseminated sarcoma. Hence, the detection of concurrent BAC was possible. Ultimately, it is important to recognize that solitary lung nodules in young patients with sarcomas can be primary malignancies, particularly BAC.

As in adults, the prognosis for BAC in juveniles is better than that for other lung malignancies, provided that the surgical resection is adequate [2]. Unfortunately, rhabdomyosarcoma recurred in this patient 10 months after surgery. Multiple lung lesions reappeared at this time, consistent with metastases. At present, the patient is still living and will continue to be follwed.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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