Primary pediatric endobronchial Ewing sarcoma family of tumors

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Summary

Background: Ewing sarcoma family of tumors is the second most common primary bone tumor of childhood. Extraosseous Ewing sarcoma family of tumors is rare. We present a pediatric case of primary endobronchial Ewing sarcoma family of tumors.

Case Report: A 12-year-old boy presented with dyspnea and chest radiography showed right pulmonary atelectasis. Chest computed tomography demonstrated tumor in the right main bronchus. Histopathological examination of the resected tumor demonstrated Ewing sarcoma family of tumors. No other lesions were detected throughout the body and the right main bronchus was thought to be the primary site. As of 1 year and 6 months after further resection of residual tumor followed by chemotherapy and radiotherapy, the patient remains disease-free.

Conclusions: Extraosseous Ewing sarcoma family of tumors arises in soft tissues of the trunk or extremities, but primary endobronchial Ewing sarcoma family of tumors has rarely been reported. Although quite rare, Ewing sarcoma family of tumors should be considered among the differential diagnoses for pediatric bronchial tumor.

key words: pediatric endobronchial tumor • atelectasis • Ewing sarcoma family of tumors

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BACKGROUND

Ewing sarcoma family of tumors (ESFT) is highly malignant and represents the second most common primary bone tumor of childhood and adolescence [1]. These tumors usually arise in bone, but can also develop in soft tissues. However, primary endobronchial ESFT is extremely rare. We present a pediatric case of primary endobronchial ESFT.

CASE REPORT

A previously healthy 12-year-old boy presented with dyspnea and was referred to our hospital. On admission, he was afebrile and dyspneic, with a respiratory rate of 24 breaths/min. Physical examination revealed extremely diminished right-sided breath sounds. Chest radiography showed right pulmonary atelectasis (Figure 1). Chest computed tomography (CT) demonstrated tumor in the right main bronchus (Figure 2). Bronchoscopic examination revealed a pedunculated mass (20×15×8 mm) in the right main bronchus, which was obstructed by the tumor (Figure 3A, 3B). The tumor was excised with a laser, with respiration improving immediately following removal of the mass. Histopathological examination of the excised specimen demonstrated small round cells. Immunostaining yielded positive results for CD99 and synaptophysin and negative results for desmin, compatible with a diagnosis of ESFT (Figure 4A, 4B). EWS-FLI1 chimeric transcripts were not identified in the tumor by reverse-transcriptase polymerase chain reaction. No other lesions were identified following whole-body CT scan and the right main bronchus was thought to represent the primary site. Because residual tumor was found on chest CT after the first excision, he first underwent further bronchoscopic excision of the tumor, followed by five courses of chemotherapy (vincristine, cyclophosphamide, pirarubicin, etoposide, and ifosfamide) and radiotherapy (45 Gy in 25 fractions). Surgery was not conducted, because the patient had no prognostic risk factors [2] and surgical intervention was considered too invasive. As of 1 year and 6 months after completion of the chemoradiotherapy, the patient remains disease-free.

DISCUSSION

Endobronchial tumor is extremely rare in children and its true incidence is unclear [3]. Histologically, the most frequent tumor in adults is squamous cell carcinoma, whereas
cartinoid tumor is the commonest in children, followed by mucoepidermoid carcinoma [4,5]. Extraosseous ESFT has been reported in skin, kidney, small intestine, and other soft tissues of the trunk and extremities [6–8]. However, primary endobronchial ESFT is extremely rare and relatively few cases have been reported to date [9].

Because no significant difference was found in patient demographics and tumor characteristics between extraosseous and skeletal ESFT [10], treatment of extraosseous ESFT usually involves intensive multi-agent chemotherapy and local control including surgery and irradiation, as for skeletal ESFT [11]. Outcomes for extraosseous ESFT resemble those of skeletal ESFT [10]. In the present case, we first considered multidisciplinary treatment, including surgery. However, as the patient had neither metastatic disease at diagnosis nor any other prognostic risk factors [2], we considered tracheobronchoplasty would be overly invasive and decided to conduct irradiation for local control.

Conclusions

Endotracheal tumor may not be immediately suspected in pediatric patients presenting with dyspnea or atelectasis, but should be considered among the differential diagnoses. Although quite rare, ESFT should be considered in cases of pediatric endobronchial tumor.

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