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Primary leiomyosarcoma of the chest wall in a child : a case report

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Leiomyosarcoma is an uncommon soft tissue sarcoma of mesenchymal cell origin, which shows smooth muscle differentiation. Leiomyosarcoma is seldom found in the pediatric population, and accounts for fewer than 2% of all soft tissue sarcomas. Leiomyosarcoma of the chest wall is extremely rare in children. We report here a case of an 8-year-old boy with a primary leiomyosarcoma that was incidentally found as a rib mass. The patient underwent a complete resection for a suspected osteochondroma diagnosed by a three-dimensional chest computed tomography examination. Pathological findings of the mass revealed intersecting fascicles of spindle cells showing cigar-shaped nuclei, inconspicuous nuclear pleomorphism and occasional mitotic figures in the background of a suspected osteochondroma of the rib. This report documents the first description of a leiomyosarcoma possibly arising in an osteochondroma of the rib in a child. **(Korean J Pediatr 2008;51:98-101)**

Key Words : Leiomyosarcoma, Chest wall, Rib, Osteochondroma, Child

Introduction

Leiomyosarcoma is a malignant tumor of mesenchymal cell origin that shows smooth muscle differentiation. In adults, leiomyosarcoma is rare, making up only 7% of all soft tissue sarcomas[1]. The most common locations of leiomyosarcoma in adults are the retroperitonium and the extremities[2]. This tumor is seldom found in children, and accounts for fewer than 2% of all soft tissue sarcomas[3]. The most common primary site of leiomyosarcoma in children is the gastrointestinal tract, especially the stomach[3, 4]. The next most frequent sites are the extremities and the trunk[5]. Rarely, leiomyosarcoma has been reported in bones, mostly in the long bones. A primary chest wall leiomyosarcoma in a child is extremely rare. In this report, we describe a case with a rib mass as a leiomyosarcoma arising in a suspected osteochondroma of the rib.

Case Report

A previously healthy 8-year-old boy presented with fever, cough and sputum. Pneumonia was suspected and a simple chest X-ray was taken. A chest posterior-anterior radiograph showed an approximately 3 cm sized ossifying mass along the right anterior aspect of the sixth rib and pneumonic infiltration in the left lower lobe (Fig. 1A). On palpation, a fixed hard mass was revealed. On three-dimensional chest computed tomography (CT) images, the mass was highly attenuated and was in focal continuity with the underlying rib (Fig. 1B, C). The mass showed inward growth on the lung side, and the presence of an osteochondroma was suspected when partially heterogeneous attenuation was noted on bone setting images. The mass appeared as a hot lesion on bone scan images (Fig. 1D). The patient was discharged after a complete segmental resection of the rib was performed without incident.

Macroscopically, the resected rib showed a protruding lobulated mass. The cut surface of the mass showed an irregular mixture of pale gray fish-flesh like areas and yellow hard areas (Fig. 2A). Microscopically, the tumor was composed of cigar-shaped spindle cells in the background of a suspected osteochondroma. A largely calcified and bony projection containing a marrow cavity that was continuous with that of the underlying rib was noted (Fig. 2B). The spindle cells exhibited inconspicuous nuclear pleomorphism with occasional mitotic figures (4/10 high power fields) and an in-

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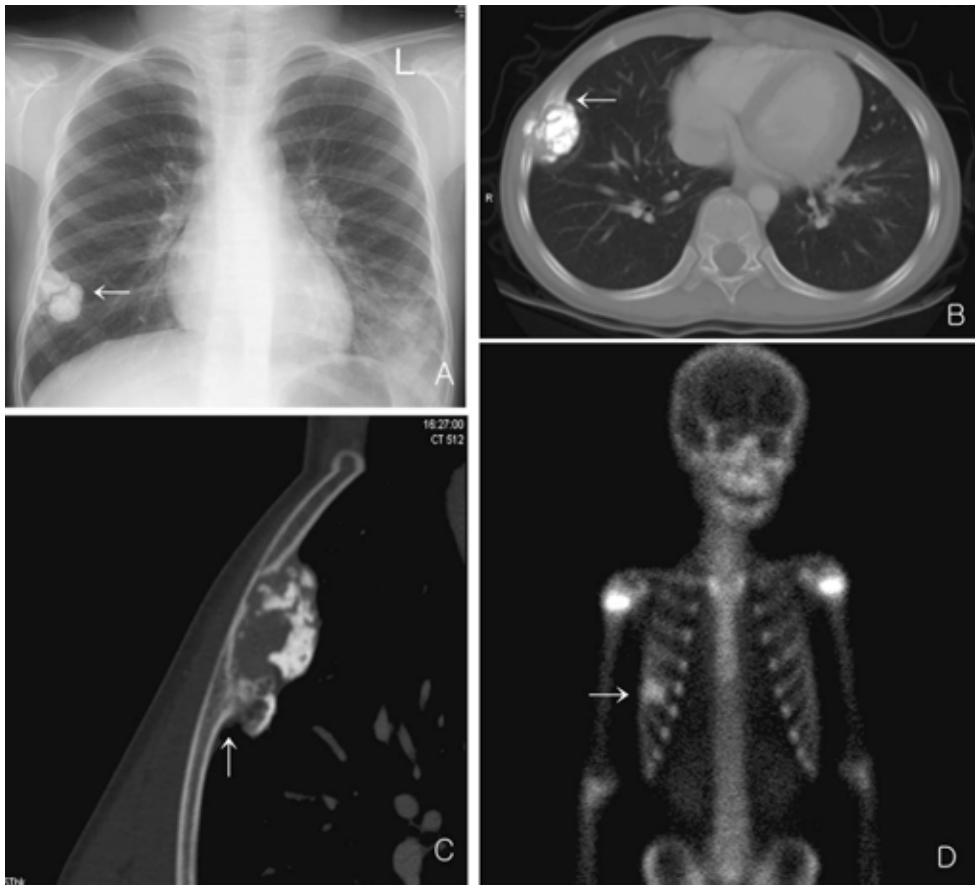

Fig. 1. Imaging studies of the mass. A) A chest posterior-anterior radiograph shows an approximately 3.2×2.6 cm sized ossifying mass (arrow) along the anterior aspect of the right 6th rib. B), C) A computed tomography scan shows osteocartilaginous exostosis (an osteochondroma) originating from the right 6th rib and focal continuity (arrow) with the underlying rib. (D) A hot uptake at the right 6th rib was seen in a bone scan (arrow).

tersecting fascicular pattern of growth (Fig. 2C). The surgical resection margins were free of the tumor. As determined by immunohistochemical analysis, the tumor cells were positive for smooth muscle markers such as vimentin, smooth muscle actin and desmin (Fig. 2D), but the tumor cells were negative for epithelial membrane antigen, cytokeratin, S-100, CD34 and C-kit. These pathological features are consistent with a low grade leiomyosarcoma possibly arising in a pre-existing osteochondroma of the rib.

Postoperative adjuvant therapy was not administered to the patient. A whole body positron emission tomography-computed tomography (PET-CT) examination performed 6 months after the complete resection showed no abnormal findings. Bone scans were also negative. At 12 months after surgery, the patient showed no local recurrence or distant metastasis.

Discussion

A primary leiomyosarcoma of the chest wall in children is extremely rare[6]. Until now, to the best of our knowledge, there were only two previous reports in the clinical literature describing a primary chest wall leiomyosarcoma in children[7, 8]. This report is therefore the third reported case of a primary chest wall leiomyosarcoma occurring in a child. In this case, based on CT findings, the mass was initially diagnosed as an osteochondroma as the bony portion of the mass was contiguous with the underlying rib. However, microscopically, the mass was diagnosed as a low grade leiomyosarcoma in the background of a suspected osteochondroma. Thus, this is the first report of a leiomyosarcoma in the background of a suspected osteochondroma.

We were unable to define whether the origin of the leio-

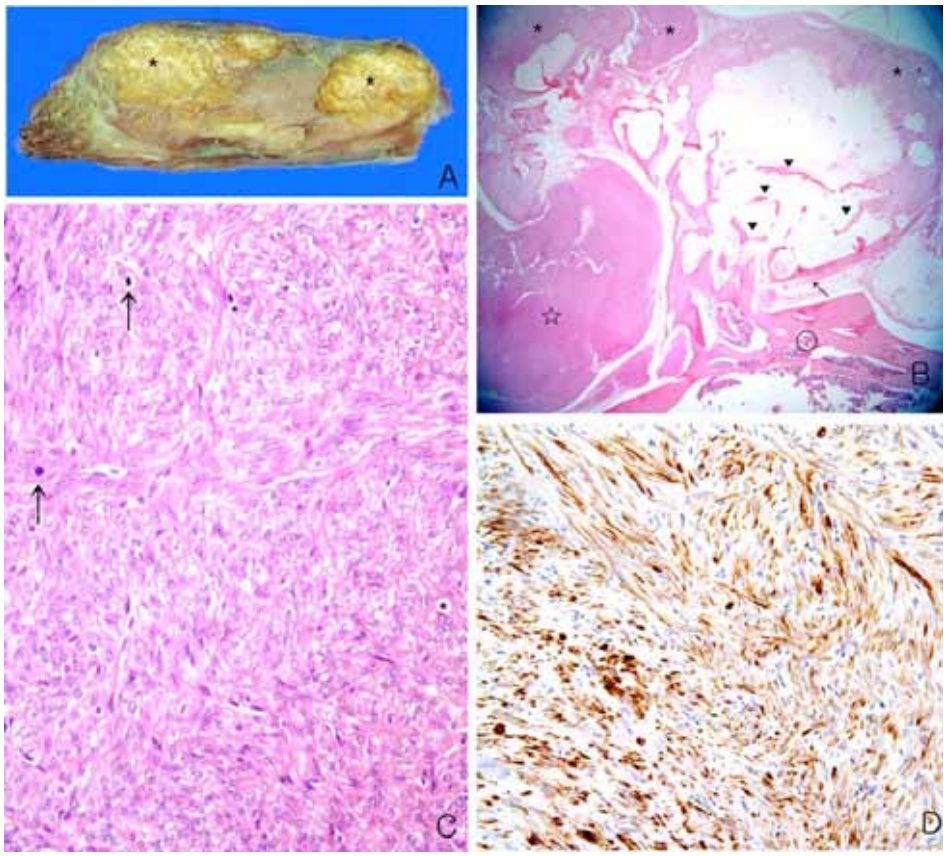

Fig. 2. Macroscopic and microscopic features of the mass. A) The cut surface of the mass is pale gray and firm with extensive yellowish calcified areas (asterisks). B) A low power photomicrograph shows a compactly cellular tumor (☆) associated with extensive calcification (asterisks) and mature bone tissue which suggests the presence of a pre-existing osteochondroma containing a marrow cavity (▼) that was continuous (arrow) with that of the underlying rib (◯) (hematoxylin-eosin staining, × 12). C) The tumor is composed of intersecting fascicles of spindle cells showing cigar-shaped nuclei, inconspicuous nuclear pleomorphism and occasional mitotic figures (arrows) (hematoxylin-eosin staining, × 400). D) The tumor cells are immunoreactive to desmin (immunohistochemical staining, × 400).

myosarcoma was a rib (bone) or the soft tissue around the rib. It was difficult to infer whether the osteochondroma was pre-existing or whether the leiomyosarcoma of the soft tissue was associated with extensive calcification and ossification. Findings favoring the former are that the lesion was seen as a bony projection containing a marrow cavity that was continuous with that of the underlying rib. Radiologically, the mass demonstrated focal continuity with the cortex of the underlying rib, and the mass was diagnosed as an osteochondroma of the sixth rib. Microscopically, the leiomyosarcoma was present in the background of a suspected osteochondroma that presented as a protruding bony mass containing a marrow cavity continuous with that of the underlying rib. Therefore, a leiomyosarcoma arising in an osteochondroma was suggested for this case. In contrast, a leiomyosarcoma of the bone shows no specific clinical or radiographic features when compared with other bone sarcomas that do not produce an extracellular matrix[9]. For the diagnosis of leiomyosarcoma of the bone, most of the sarcomatous tissue (≥70%) has to be intramedullary located with only limited extraosseous extensions[10]. The mass was protruding from the sixth rib and most of the mass was not intramedullary in location. Therefore, we could not confirm that this mass was a leiomyosarcoma of the bone. Calcification within the leiomyosarcoma was neither uncommon nor exclusive[11].

It was very interesting that a pre-existing osteochondroma was strongly suspected based on the radiological and pathological findings in this case. Therefore, this leiomyosarcoma may have formed within an osteochondroma of the

underlying rib. As a leiomyosarcoma arising in an osteochondroma has not previously been reported, it is recommended that confirmative diagnosis by biopsy should be performed to diagnose additional cases like this in the future.

Treatments for leiomyosarcoma are still under debate. A high grade leiomyosarcoma is very aggressive and preoperative chemotherapy, surgery with wide resection or amputation and postoperative adjuvant therapy is often performed[9, 10]. The most commonly used chemotherapeutic agents are doxorubicin, ifosfamide and mesna, and radiotherapy is often given for postoperative adjuvant therapy[10]. However, there is no significant statistical difference between the use of surgery alone and surgery with chemotherapy and/or radiotherapy[10]. Low grade leiomyosarcoma is often treated with surgery alone[10].

In children, wide local excision is the most important treatment. Although the use of chemotherapy and radiotherapy is still under debate, it is often used in postoperative adjuvant therapy[4].

In this case, the mass was diagnosed as a low grade leiomyosarcoma. It was completely resected and the margin of the cutting surface was free of malignant cells. Whole body PET-CT and bone scans showed no evidence of residual tumor or recurrence. During the 1 year follow-up after surgical removal, the patient remained healthy without adjuvant therapy.

In summary, we have presented a case of a primary chest wall leiomyosarcoma in the background of a suspected osteochondroma that was located on the rib of an 8-year-old boy.

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한 글 요 약

소아의 흉벽에서 진단된 평활근육종 1례

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평활근육종은 중간엽세포 기원의 평활근 분화를 보이는 연부조직육종으로 소아에서는 전체 연부조직육종의 2% 이하의 발생률을 보이는 매우 드문 질환이다. 특히 흉벽의 평활근육종은 더욱 드물다. 저자들은 우연히 발견된 흉벽 종양을 방사선학적 검사로 골연골종으로 추측하여 완전절제술을 실시하였고, 조직검사 결과 저등급 평활근육종으로 진단된 1례가 있어 보고하는 바이다.

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