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늑골에서 발생한 거대세포종: 1예 보고 및 문헌 고찰

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거대세포종은 늑골에서 드물게 발생할 수 있으며, 후중격에서 발생한 종괴로 나타난 늑골의 거대세포종은 지금까지 4 예가 보고되었다. 38세 남자의 늑골에서 발생하여 후중격 종괴로 보인 거대세포종 1 예를 문헌 고찰과 함께 보고한다. 흉부 전산화 단층 촬영에서 우측 후상부 중격의 대부분을 차지하는, 경계가 명확한 다분엽성의 종괴가 우측 3번 늑골과 흉추를 침범하고 있었다. 임상적으로는 후중격에서 발생한 신경절신경종 혹은 그와 동반된 악성 변화를 의심하였다. 그러나 육안적으로 종괴는 우측 3번 늑골에서 발생하여 늑골 바깥쪽으로 성장하는 모습을 보였고, 현미경적으로 균일하게 산재된 다핵 거대 세포와 단핵 기질 세포로 구성되어 있어 늑골에서 발생한 거대세포종에 합당하였다. 거대세포종의 치료를 위해서는 재발과 전이의 가능성을 고려하여 광범위한 수술적 절제와 술후 방사선 치료를 고려해야 한다. 후중격 신경절신경종은 수술적 절제만으로 치료가 가능한 종양이므로, 거대세포종과 반드시 감별해야 한다.

색인 단어: 늑골, 거대세포종, 신경절신경종

INTRODUCTION

Giant cell tumor (GCT) represents 5% of all primary bone tumors and approximately 25% of benign bone tumors[1]. GCT usually involves the long bone epiphysis in skeletally mature patients and the metaphysis in skeletally immature patients[2]. GCT rarely occurs in the rib. Plain chest roentgenography shows a well-defined, expansile, osteolytic lesion, which is usually covered with a thin shell of reactive new bone. Computed tomography (CT) also shows a well-defined, expansile lesion of soft tissue density. Internal calcified septae and punctuate calcification are frequently seen[3]. When GCT occurs in the posterior arc of the rib, it may form a large destructive mass that presents

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as a posterior mediastinal mass[3]. We describe a case of GCT showing substantial growth out of the rib and mimicking a posterior mediastinal ganglioneuroma. We also review the relevant literature (GN).

CASE REPORT

A 38-year-old man presented with a 2-year history of pain in his right upper back and chest wall. He had recurrent episodes of this pain associated with decreased sensation in the right axilla. He had no remarkable past or family histories. On admission, the physical examination revealed no abnormalities. Routine laboratory tests, including serum calcium, phosphorus, alkaline phosphatase and tumor markers, were within normal range.

A plain chest roentgenography showed an ovoid, expansile upper paratracheal mass (Fig. 1). A chest CT showed a 5.5×4.2×4.3-cm well-defined, heterogeneous, multi-lobulated mass of soft tissue density with relative enhancement in the right posterior mediastinum. The mass appeared to invade the medial end of the right third rib and caused a bulging appearance in the posterior cortex (Fig. 2). Erosive change was also identified in the body of the third thoracic vertebra, but extensions into the spinal canal, esophagus or trachea were not identified. Although most of the mass was of soft tissue density, it also showed subtle, irregular calcified densities. The lung parenchyma was unremarkable.

A technetium-99m methylene diphosphonate bone scan showed focally increased uptake of the radionuclide around the right third costovertebral joint, consistent with rib invasion. Based on the radiologic finding, it was thought to be a posterior mediastinal GN. Because tumor invasion into the rib and vertebra was strongly suspected, ganglioneuroblastoma was included in the differential diagnosis. The mass, which occupied the mediastinum, the right third and fourth ribs and the right second to fourth intercostal muscles, was excised.

Grossly, there was a 2.2×0.7×0.7-cm red to brown firm tumor mass in the marrow cavity of the third rib, which was expanding and destroying the cortical bone. The third costovertebral joint was involved by

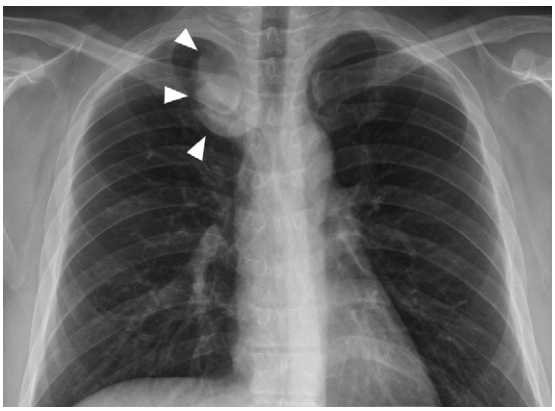

Fig. 1. Plain chest roentgenography shows an expansile upper paratracheal mass (arrowheads).

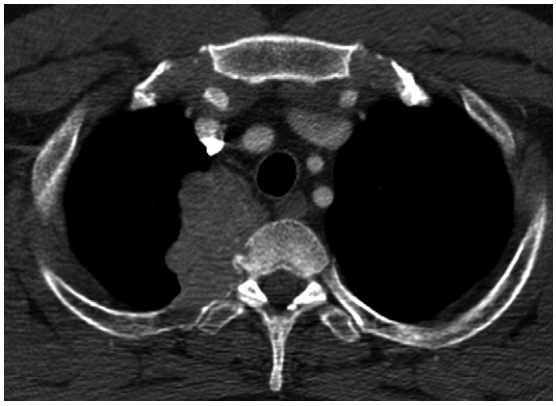

Fig. 2. Contrast-enhanced computed tomography shows a well-defined, lobulated, heterogeneous mass of soft tissue density associated with destruction of the right third rib and thoracic vertebra in the right posterior mediastinum.

the tumor. Fragmented mediastinal masses were relatively well-circumscribed. The largest mass measured 5×4.5×3 cm and was partly enclosed by a yellow to pink calcified capsule. The cut surfaces of the mass showed beefy-red viable-appearing areas accompanied by multiple foci of hemorrhage (Fig. 3). Histologically, the tumor consisted of diffusely interspersed, multi-nucleated, osteoclast-type giant cells and oval to spindle-shaped stromal mononuclear cells without significant cytologic atypia (Fig. 4). The appearance of the giant cell nuclei was similar to that of the stromal cells. Mitotic activity was apparent in the stromal cells but not in the giant cells; no atypical mitosis was identified. Some areas of the tumor contained small, multi-focal, aneurismal bone cyst-like spaces. Reactive osteoid and woven bone were also found, especially at the periphery.

The pathologic diagnosis was a GCT of the rib involving the posterior mediastinum. Twelve months after the surgery, the postoperative course has been uneventful, except for mild chest discomfort and anhidrosis of the right side of the face and the right hand due to the sympathectomy.

DISCUSSION

Although GCT of the rib usually arises in the posterior arc, no reports have mentioned the differential diagnosis between GCT and GN, which is the most common benign tumor arising in the posterior mediastinum. We found several radiologic findings in our case that may contribute to a misdiagnosis of GN. CT showed that the lesion formed a large paravertebral mass and that the medial end of the rib adjacent to the mass showed cortical thinning and expansion.

Based on these findings, the radiologist's interpretation was that the paravertebral mass had invaded the rib, not that the primary rib tumor extended into the mediastinum, which was the case. In addition, internal calcification was punctuated and more subtle than that of typical GCT, and neither a sclerotic rim nor peripheral enhancement was identified. Based on these findings, we considered GN, which is the

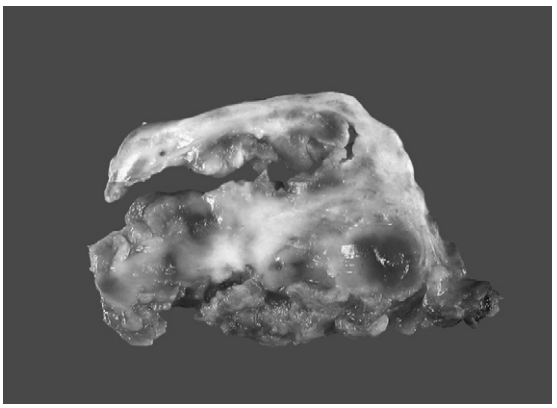

Fig. 3. Grossly, the mediastinal mass is partly enclosed by a calcified capsule and consists of beefy red to yellow, viable-appearing areas and multiple foci of hemorrhage. There are cleft-like empty spaces beneath the capsule.

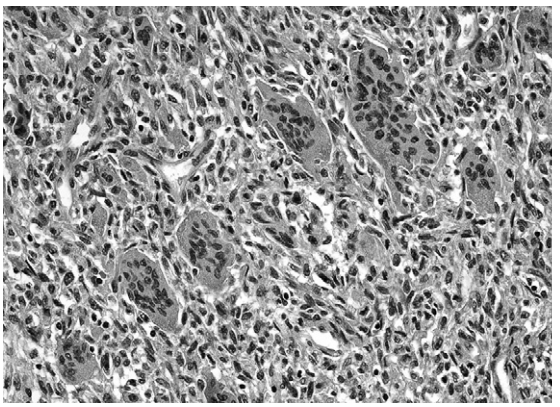

Fig. 4. Microscopically, the tumor consists of diffusely interspersed multi-nucleated, osteoclast-like giant cells on a background of oval to spindle-shaped mononuclear stromal cells (Hematoxylin & eosin stain. Original magnification, ×400).

most common tumor of the posterior mediastinum.

On a plain chest roentgenography, posterior mediastinal GN typically appears as a large, well-defined round or oval paravertebral mass[4]. By contrast-enhanced CT, it manifests as a well-defined, lobulated, homogenous or mildly heterogeneous paravertebral mass with mild to moderate enhancement. Fine, speckled or coarse calcifications may be also detected. We suggest that when there is a large, expansile soft tissue mass in the posterior mediastinum, GCT of the rib should be differentiated from posterior mediastinal GN. Because GCT is locally aggressive, with a high local recurrence and low metastatic potential, radical surgical resection with postoperative radiation therapy is required to control local recurrence.

Neoplastic lesions that can present as posterior mediastinal masses include primary tumors of the chest wall, posterior mediastinum or lung, as well as metastatic tumors. Among these, primary chest wall tumors are relatively uncommon, and primary tumors involving the bony skeleton of the chest wall are even more rare. In particular, primary rib tumors comprise only 5 to 7% of all primary bone neoplasms[5]. About 0.5% of GCTs occur in the rib[1]. From 1991 to 2008, 19 cases of GCT were reported in the rib, including 16 in the English and 3 in the Korean literature. The clinicopathologic features of these reports and for our case are summarized in Table 1.

Of 17 cases that reported tumor locations, 10 cases arose in the anterior arc of the rib and 7 in the posterior arc. In 4 of 7 cases where GCT arose in the posterior arc, the tumor subsequently invaded the thoracic vertebra or posterior chest wall. In cases with vertebral invasion, complete excision may be difficult due to anatomical and neurologic limitations. In 2 cases, the tumors could not be resected due to massive bleeding and extensive involvement for nearly half of the vertebral body[6]. These 2 patients received postoperative radiation therapy for residual tumors.

In fact, the use of postoperative radiation therapy for GCT has not been recommended due to the possibility of malignant transformation. Campanacci et al. reported malignant transformations in 8 of 27 GCT patients (29%) following irradiation[7]. In a series by McGrath et al., 5 of 21 GCT patients developed sarcomatous changes at the irradiation site[8]. In contrast, Nair et al. reported that details regarding energy and radiation schedules were lacking for most of the earlier studies[9]. Recent reports suggest that mega-voltage radiotherapy is an effective, well-tolerated alternative to surgery without risk of malignant transformation[9]. Nonetheless, optimal management of GCT of the bone is complete tumor resection with wide margins, if possible[10]. Postoperative radiation therapy may be necessary to treat patients with extensive, aggressive or incompletely resected GCT. In our case, although the tumor invaded the costovertebral joint, complete excision was possible. The patient received no postoperative radiation therapy.

In addition, 3 patients received preoperative radiation therapy for improving resection outcomes or chemoradiation therapy due to tumor aggressiveness and pain. Additional studies on the therapeutic effects of preoperative radiation and chemoradiation should be performed. Although tumors did not recur in any of these patients, follow-up periods longer than 5 years after surgery were available for 2 cases only. Longer observa-

Table 1. Clinicopathologic features of giant cell tumor of the rib reported in the English and Korean literature

No	Age/sex	Location		Size (cm)	Treatment	Presentation	Involvement	Recurrence	Follow-up	Reference
1	38/M	Posterior	3rd	5.5×4.2	CR	Chest wall pain	CV joint	No	12 months	Our case
2	28/M	Entire hemithorax		25.0×17.0	Preop. RT + CR	Atelectasis	No	No	24 months	Cordeiro et al. (2008)
3	28/F	Anterior	4th	15.0×11.0	CR	Breast lump	No	N/A	N/A	Rashid et al. (2007)
4	34/M	Posterior	10th	N/A	CR	Back pain	N/A	N/A	N/A	Maki et al. (2007)
5	30/F	Anterior	5th	N/A	CR	Breast swelling	No	No	4 years	Kumar et al. (2007)
6	46/F	Anterior	9th	8.0×5.0	CR	Painless mass	No	No	N/A	Al-Otaibi et al (2006)
7	36/M	Posterior	3rd	11.5×10.0	CR + Postop. RT	Mediastinal mass	Chest wall	No	14 months	Volmar et al. (2004)
8	57/M	Posterior	5th	7.0×6.5	CR + Postop. RT	Back pain	Vertebra	No	6 years	Sakao et al. (2003)
9	25/M	Anterior	8th	8.0×5.0	CR	Painless mass	No	N/A	N/A	Reddy et al. (2003)
10	12/F	Posterior	9th	8.0×8.0	CR	Painless mass	No	No	12 months	Athanassiadou et al. (2003)
11	27/F	Anterior	2nd	12.0×8.0	CR	Painful mass	No	No	5 years	Briccoli et al. (2003)
12	40/M	Anterior	2nd	8.0×6.5	CR	Painful mass	No	No	11 months	Shin et al. (2002)
13	25/F	Posterior	4th	6.0×3.5	CR	Painless mass	No	No	6 months	Ninomiya et al. (2002)
14	47/M	Anterior	6th	5.0×5.0	CR	Painful mass	No	No	9 months	Chang et al. (2002)
15	20/M	Anterior	3rd	6.0×6.0	CR	Painful swelling	No	No	N/A	Gupta et al. (2000)
16	51/M	Posterior	7th	12.5×6.5	Preop. CRT + CR	Chest wall pain	Vertebra	No	N/A	Tanaka et al. (1996)
17	31/M	N/A	1st	9.0×8.0	CR	Chest wall mass	N/A	N/A	N/A	Song et al. (1993)
18	29/M	Anterior	9th	8.0×6.0	Preop. CRT + CR	Painful mass	No	No	19 months	Hanna et al. (1992)
19	50/M	Anterior	3rd	7.0×6.0	CR	Painful mass	No	No	6 months	Hendra et al. (1991)
20	24/M	Posterior	2nd	N/A	IR + Postop. RT	Painful mass	Vertebra	No	N/A	Ju et al. (1991)

CR indicates complete resection; CV, costovertebral; preop., preoperative; postop., postoperative; RT, radiation therapy; CRT, chemoradiation therapy; IR, incomplete resection; N/A, not available.

tion periods and more cases are needed to clarify prognoses.

In summary, we described a GCT arising in the posterior arc of the rib. Because GCT has a high potential for local recurrence and metastasis, it must be differentiated from posterior mediastinal GN that can be treated with surgical excision alone. For GCT, a wide surgical excision with elective postoperative radiation therapy remains the mainstay for management. Appropriate clinical, radiologic and pathologic assessments may facilitate diagnosis and avoid delays in treatment.

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Abstract

Giant Cell Tumor of the Rib: A Case Report and Review of the Literature

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Giant cell tumor (GCT) of the rib may present as a posterior mediastinal mass when it involves the posterior arc. Only 4 cases of GCT of the rib presenting as a posterior mediastinal mass have been reported. We report a case of a 38-year-old man with GCT of the rib. Computed tomography revealed a well-defined, multi-lobulated, heterogeneous mass in the right supero-posterior mediastinum, which appeared to invade the right third rib and thoracic vertebra. It was thought to be a posterior mediastinal ganglioneuroma or its malignant transformation. Grossly, the tumor mass arose in the posterior arc and showed substantial growth out of the rib. Microscopically, the tumor consisted of interspersed multi-nucleated giant cells and stromal mononuclear cells, compatible with GCT. For GCT, a wide excision with elective radiotherapy should be considered. GCT must be differentiated from posterior mediastinal ganglioneuroma that can be treated by surgical excision alone.

Key Words: Rib, Giant cell tumor, Ganglioneuroma

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