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Case Report

Giant Fibrous Dysplasia of the Rib: A Case Report

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ABSTRACT

Fibrous dysplasia is a benign disease in which normal bone tissue is resorbed by fibrous tissue and immature bone structure. Fibrous dysplasia has been reported to account for approximately 5%–7% of all benign bone tumors; the rib involvement is rare. Surgical resection is the preferred treatment method in symptomatic patients, and the reconstruction should be carried out according to the size of the defect. Reconstruction technique involves stabilizing the thorax and closed soft tissue defects with a graft. This study aimed to present a case of an 18-year-old patient diagnosed with fibrous dysplasia characterized by atypical symptoms such as shortness of breath and pain reconstructed with sandwich graft.

INTRODUCTION

Fibrous dysplasia is a slowly progressive, benign disease in which normal medullar bone is replaced by abnormal fibro-osseous tissue.[1] It was first described by Lichtenstein in 1938.[2] Although its etiology is not completely known, somatic gene mutations are considered to be responsible for this disease.[3] It constitutes 5%–7% of bone tumors.[4] Fibrous dysplasia is of three types: monostatic type (80%), polystatic type (3%–4%) type, and McCune–Albright syndrome (3%).[5] Fibrous dysplasia usually starts in childhood, and the frequency increases in puberty and adolescence period. However, its prevalence decreases in adulthood.[6] The recommended treatment approach is extensive resection. Different reconstruction techniques are used to maintain thoracic wall stability and protect intrathoracic organs.[7]

This study presented the case of an 18-year-old patient with fibrous dysplasia reconstructed with a sandwich graft. The patient had a mass on the anterior wall of the thorax and complained of dyspnea.

CASE REPORT

The physical examination of the patient revealed no palpable mass on visiting the thoracic surgery clinic. However, the breathing sounds were not perceived in the left bottom lung zone during auscultation. No abnormalities were detected in his biochemistry and blood count. The pulmonary function test (PFT) of the patient who did not have a smoking history revealed the following: FEV1, 75%; FVC, 78%; FEV1/FVC, 96%; FEF 25%–75%, 115%; PEF, 64%; FEF 50%, 109%. His posterioanterior chest x-ray showed a round and radiolucent mass image, 8 cm in diameter at the level of fifth rib which extended to the left diaphragm and covered most of the left bottom zone but did not block the costodiaphragmatic angle (Figure 1a). Contrast-enhanced thoracic computed tomography (CT) was used to de-

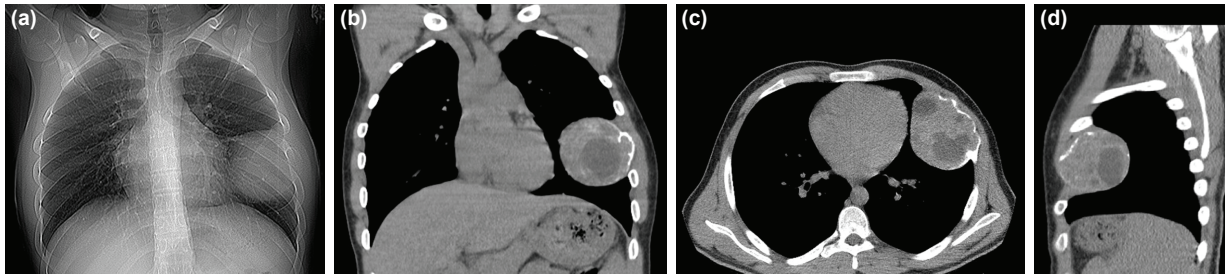

Figure 1. (a) Posteroanterior (PA) lung image. **(b)** Computed tomography image (frontal) **(c)** Computed tomography image (sagittal) **(d)** Computed tomography image (coronal).

termine the involvement of the main vascular structures, pleura, and mediastinum and also to direct the surgical technique. A solid lesion containing necrotic components, which caused destruction on the neighboring fifth costa with axial dimensions of approximately 10×7.5 cm^2 at the left lung lingular segment level, was detected in the tomography (Figures 1b–d). The surgical excision was planned for the diagnosis and treatment of the patient. Anesthesia was induced using 7 mg/kg pentobarbital, 0.6 mg/kg rocuronium, and 2 μg/kg fentanyl. The right lateral decubitus position was achieved by intubation with a single-lumen endotracheal tube. The operation was started with an oblique incision from the left rear axillary line. When the thorax was opened with left lateral thoracotomy, a mass, 12×8.5×7 cm^3 in size, sticking to the fifth costa, and having no connection with either the lung or the heart, but creating pressure on the lungs, was detected in the anterior mediastinum. Then, 5 cm of the mass at the fifth costa, which reached an extremely large size, was resected by leaving a safe surgical margin against the possibility of malignancy. The resulting costa defect was reconstructed with a mercury mesh patch and a methyl methacrylate sandwich graft to prevent paradoxical movement. The graft was fixed with double-sided prolene sutures into a mersilene mesh patch. Despite surgical cauterization, a thoracic tube was placed in the base and apex of the lung due to bleeding from the thoracic wall in the form of leak. The bleeding follow-up was done through postoperative drains. The pathological examination of the excised capsular mass, weighing 401 g (Figure 2), showed a capsular view containing vascular areas macroscopically. Moreover, some irregular bone structures in the fibrous stroma containing fusiform cells were also detected microscopically. The diagnosis was made as fibrous dysplasia (Mazabraud syndrome, McCune–Albright syndrome). As a result of the pathology, no additional clinical findings were found in the patient in terms of predicted syndromes.

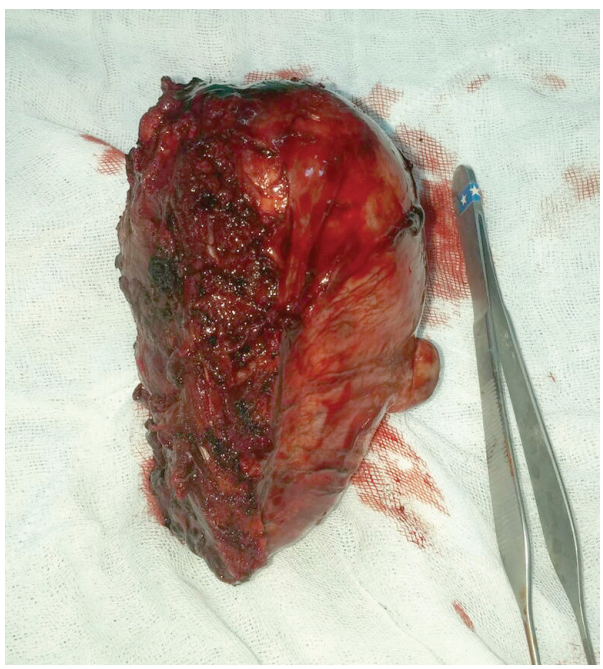

Figure 2. Resected surgical mass.

The patient was followed up at the first step anesthesia intensive care unit in the first postoperative 24 h and then transferred at the end of 24 h. His repeated PFT revealed the following: FEV1, 88%; FVC, 93%; FEV1/FVC, 94%; FEF 25%–75 %, 109%; PEF, 74%; FEF 50, 109%. Since the vital signs of the patient were stable, the thorax tube placed in his chest basal region was removed on the third postoperative day and the other thorax tube placed in the apex region was removed on the fourth postoperative day. The patient was discharged on the seventh postoperative day and followed up periodically.

DISCUSSION

Fibrous dysplasia is characterized by the pathology in which resorbing normal bone tissue is replaced by fibrous tissue and some immature bone structures. Histopathologically, it can be seen in various stages of bone metaplasia. Endocrine anomalies, trauma, bone growth defects, and hamartomatous malformation have been proposed for the pathogenesis of this disease. However, no well-accepted theories have been adopted so far. It constitutes approximately 5%–7% of benign bone tumors. [3] Monostatic fibrous dysplasia, which is defined as the most commonly monocentric form, accounts for 80% of cases. Femur, tibia, mandibula, and maxilla are the bones

most frequently involved. The costa involvements were rarely reported.[8,9]

Fibrous dysplasia is usually detected incidentally during radiological examinations due to other causes such as sports injuries or trauma.[3] However, they may become symptomatic by causing pain, mechanical loss of strength, neuropathy, pathological fractures due to thinning of bone cortex, and asymmetric deformities depending on the location and extent of the involvement.[10] Aydogdu et al. studied dysplasia cases retrospectively, 85% of the cases were in monostatic form and involved one costa. All of these patients complained of pain; only one had swelling. [11] The patient in the present case had atypical symptoms such as dyspnea and pain with effort.

Fibrous dysplasia is classified as a benign bone tumor; sarcomatous degeneration may be seen in rare cases (1%), especially in patients with increased ionizing radiation exposure.[10]

The diagnosis of fibrous dysplasia is made by clinical, radiological, and histopathological examinations. They usually progress asymptomatically before reaching a large size and causing pathological bone fractures and neuropathic pain. Radiologically, they are seen in direct radiographs as medullary localized, lytic, with frosted glass density, and forming expansive bones and sharp sclerotic margins.

Calcifications can be observed if a significant amount of cartilage is present in the lesion.[9] CT is the most important imaging method in terms of showing the details of the lesions. According to the mineralization of the excised tissue, CT densities may have different looks such as radiolucent (difficult to distinguish from a simple bone cyst), frosted glass (uniform fibrous and bony structures), and sclerotic (distinct bone tissues).[12]

Even if the radiological imaging methods assist in diagnosis, the general approach to tumoral masses is to take biopsy material by performing surgical excision for both excluding the malignancy and classifying and making a histopathological diagnosis. Histopathologically, it is composed of proliferating fibroblasts and abundant collagen, which surrounds the irregular and narrow maturation defect (similar to the fishhook or Chinese letters). The density and cellularity of the fibrous tissue vary.[13]

Early diagnosis and surgical resection of primer chest wall tumors play a key role in the treatment. Surgical excision is sufficient in benign lesions. Basically, correction of the deformity, avoidance of possible pathological fractures, and regression of the clinical symptoms should be aimed. Reconstruction of thoracic wall defects after resection should not impair respiratory physiology. Reconstruction involves stabilizing the bone thorax and grafting soft tissue defects depending on the size of the defect and the area covered. Grafts are of two types: autologous grafts and synthetic grafts.[6,10,11] In the present case, a polypropylene patch methyl methacrylate sandwich graft was used as a synthetic graft for thoracic bone stabilization due to the destruction of mass on the costa. No neurological deficits developed due to the localization of the lesion. The progression of the mass was followed up periodically for 6 months.

Conclusions

Although fibrous dysplasia is frequently detected incidentally, is characterized by chest disease symptoms, such as dyspnea and apnea, depending on the location and extent of involvement. In symptomatic cases, the mass should be surgically excised. Excision should be followed by the reconstruction technique depending on the size of the pathological mass. It is good to follow up periodically due to the risk of developing a low-grade malignancy.

Authorship contributions

Consept: S.K., S.B.; Design: F.K., S.B.; Data collection or processing: S.K., F.K.; Analsis and/or interpretation: S.K., S.B.; Literature search: F.K., S.B.; Writing: S.K., F.K.; Critical review: S.K., S.B.

Conflict of interest

None declared.

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Dev Kostal Fibröz Displazi: Olgu Sunumu

Fibröz displazi normal kemik dokusunun rezorbe olarak yerini fibröz doku ve immatür kemik yapıların aldığı benign bir hastalıktır. Benign kemik tümörlerinin %5–7'sini oluşturduğu bildirilmiştir ve kosta tutulumu oldukça nadirdir. Semptomatik olgularda kitle cerrahi olarak eksize edilmeli ve oluşan defektin büyüklüğüne göre rekonstrüksiyon yapılmalıdır. Rekonstrüksiyon tekniği kemik toraksın stabilize edilmesi ve yumuşak doku defektlerinin greft ile kapatılmasını içermektedir. Bu yazıda, 18 yaşında nefes darlığı ve ağrı gibi atipik semptomlar ile fibröz displazi tanısı alan ve sandviç greft ile rekonstrüksiyon yapılan olgunun sunulması amaçlandı.

Anahtar Sözcükler: Fibröz displazi; kosta; rekonstrüksiyon.