CASE REPORT

Case report of rib polyostotic fibrous dysplasia

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Abstract

Polyostotic fibrous dysplasia is a rare benign asymptomatic tumour of the ribs not requiring surgery on most occasions. We present here a case with left 10th and 11th rib fibrous dysplasia which was causing a hinderance to the Urologist for renal stone extraction. Therefore the 10th and 11th ribs were excised followed by chest wall reconstruction.

Keywords: Fibrous dysplasia, polyostotic, thoracic wall, ribs, neoplasms

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Introduction

Lichtenstein was the first to describe fibrous dysplasia as a benign intramedullary tumour in 1938.[1] Fibrous dysplasia (FD) is a rare genetic bone disorder leading to normal bone replacement with abnormal fibro-osseous tissue. The newly formed abnormal skeleton is weak and more susceptible to deformity and fractures.[2] The FD epidemiology is not well understood. Many patients with asymptomatic fibrous dysplasia can be missed. In addition, fibro-osseous lesions may be misdiagnosed as monostotic FD due to the vague character of fibro-osseous bone disorders.[1] Monostotic fibrous dysplasia subtype patients may skip evaluation for endocrine disorders and fibrous dysplasia polyostotic subtypes, which leads to difficulties in determining the prevalence other subtype's prevalence.[3]

FD is the result of healthy bone tissue replacement with abnormal fibro-osseous tissue. The fibrous tissue has abundant fibroblast cells which can lead to excessive immature bone formation.[2] The clinical presentation of FD depends on the extent and location of the skeletal lesions. It may be a solitary bony lesion or can affect multiple bones across the whole body.[2] The case of a male patient with fibrous dysplasia of the 10th and 11th left ribs, diagnosed incidentally by the Urologist wanting to extract renal stones is presented.

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Case report

A 40-year-old male patient presented to the Kafrelsheikh University Hospital, Kafrelsheikh, Egypt with a left chest wall mass. There were no symptoms and the mass was accidentally discovered by chest computed tomography done one year back. There was an expansile rib lesion on the left 10th and 11th ribs and ultrasound-guided biopsy was performed. This showed no evidence of malignancy. He was given conservative treatment and advised a follow up.

As the patient was having recurrent renal colic caused by left renal stones, the urologist recommended laparoscopic lithotomy. As the rib mass was causing an obstacle to this procedure, the urologist referred the patient to the cardiothoracic surgery department.

Figure 1: chest computed tomography which show expansile osteolytic lesion on 10th and 11th ribs with ground glass matrix thin cortex with no cortical breaks

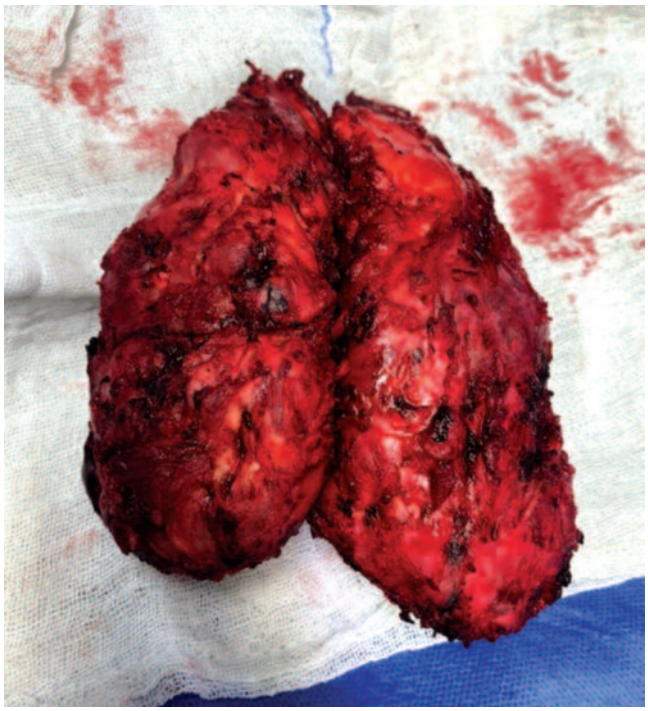
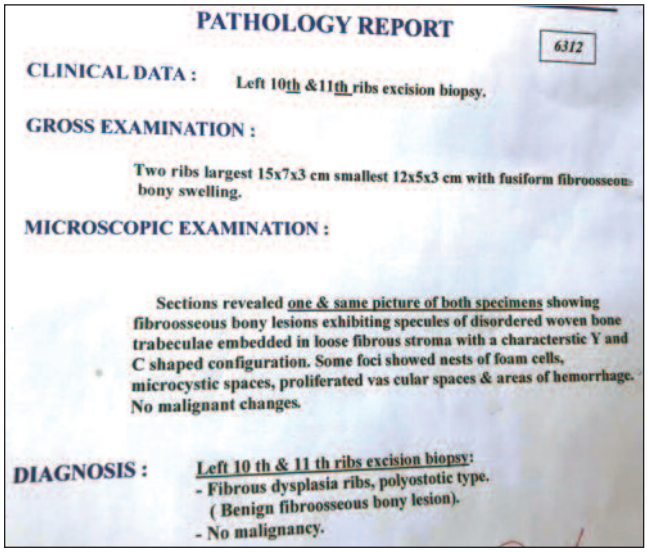


Figure 2: show the resected fibrous dysplasia rib



PATHOLOGY REPORT

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CLINICAL DATA : Left 10th &11th ribs excision biopsy.

GROSS EXAMINATION :

Two ribs largest 15x7x3 cm smallest 12x5x3 cm with fusiform fibroosseous bony swelling.

MICROSCOPIC EXAMINATION :

Sections revealed one & same picture of both specimens showing fibroosseous bony lesions exhibiting specules of disordered woven bone trabeculae embedded in loose fibrous stroma with a characterstic Y and C shaped configuration. Some foci showed nests of foam cells, microcystic spaces, proliferated vas cular spaces & areas of hemorrhage. No malignant changes.

DIAGNOSIS : Left 10 th & 11 th ribs excision biopsy:
- Fibrous dysplasia ribs, polyostotic type.
(Benign fibroosseous bony lesion).
- No malignancy.

Figure 3: show the histopathology result.

Physical examination revealed a diffuse firm indolent mass on the left lower chest wall over the 10th and 11th rib. An appendectomy scar was visible. shows No other abnormalities were noted on general examination.

Chest computed tomography showed an expansile osteolytic lesion on the 10th and 11th ribs with ground glass matrix thin cortex but no cortical breaks. (Figure 1)

The patient was intubated by a single-lumen endotracheal tube and was positioned in a lateral decubitus position. An incision was made along the 10th rib. The latissimus dorsi muscle and serratus anterior muscles were preserved. The latissimus dorsi muscle flap was prepared. The mass on both the 10th and 11th ribs was completely excised. The gross appearance was a hard, lobulated, grey-white expansile mass measuring 15×7×3 and 12 ×5×3 cm. (Figure 2). The defect was reconstructed with a double-layer prolene mesh and a Latissimus muscle flap. The patient was extubated intraoperatively and was transferred to the surgical intensive care unit. The intercostal chest tubes were removed on the 2nd and 3rd- post-operative. The patient was discharged from the hospital on the 6th post-operative day. Histologically, the lesion consisted of fibro-osseous bony lesions exhibiting spicules of disordered woven bones trabeculae embedded in the loose fibrous stroma. Some foci showed nests of foam cells, microcystic spaces, and areas of haemorrhage. Fibrous dysplasia ribs, polyostotic type were witnessed. (Figure 3). The patient attended the out-patients clinic for follow-up and to date no recurrence has been observed.

Discussion

Lichtenstein is the first one to describe fibrous dysplasia as a benign intramedullary tumour in 1938.[1] This neoplasm constitutes 6 % of benign bone tumours[4]. Fibrous dysplasia has two subtypes, monostotic and polyostotic, Monostotic type is more common and usually affects a single bone while the polyostotic type is rare and involves multiple bones.[5]

Fibrous dysplasia represents 30% of benign rib tumours with monostotic type being four times more prevalent than the polyostotic type.[6] The percentage of fibrous dysplasia subtypes is 75% for the monostotic subtype and 25% for the polyostotic subtype.[7] Patients of the age group, 20 to 40 years are more commonly seen to present with fibrous dysplasia with an equal male to female ratio.[5] Gsα gene mutation is thought to be the cause of fibrous dysplasia development.[8] Mazabraud's syndrome is characterized by the presence of fibrous dysplasia of polyostotic subtype and soft-tissue myxomas.[9] McCune-Albright syndrome is distinguished by the presence of fibrous dysplasia polyostotic subtype, and endocrine dysfunction. Both disorders are common among patients with polyostotic fibrous dysplasia.[10]

It is usually asymptomatic but it can present with pathological fractures or pain and has a low malignant transformation.[11] Occasionally multiple rib fibrous dysplasia causes restrictive lung disease which causes shortness of breath.[12] The gross appearance of fibrous dysplasia is a hard or firm mass replacing the bone cavity. The histopathology

of fibrous dysplasia is characterized by a well-circumscribed firm mass or lesion circumscribed by lamellar bone. The fibrous dysplasia tissue is composed of spindle cell fibroblasts with low cellularity embedded in a collagen matrix having a whorled appearance.[13]

The radiological findings of fibrous dysplasia are not specific but are characteristic, depending on the histopathology of the underlying lesion. Imaging shows medullary fusiform unilateral enlargement, cortical thickening with deformity, and increasing trabeculation. It appears as ground glass which is characteristic and caused by the fibrous and bone component which replaces the bone medullary cavity.[13] Stable and asymptomatic. FD lesions usually need no intervention. Surgery is indicated only for diagnostic purposes, confirmation, deformity management, and failure of conservative therapy. For patients with polyostotic fibrous dysplasia subtypes and not compatible with surgery, bisphosphonate is the drug of choice.[14,15]

In our case, it was necessary to excise the rib fibrous dysplasia mass to enable the urology surgeon to perform laparoscopic lithotomy.

Conclusion

Polyostotic fibrous dysplasia is a rare benign tumour that is usually treated conservatively and the patient is kept under follow up. In other cases, the best treatment choice is surgical excision if the mass is causing deformity or unbearable symptoms as pain. In a case scenario as ours, where the fibrous dysplasia rib mass was causing a hinderance for the Urologist in renal stone extraction surgery, it had to be excised.

Disclaimer: The abstract has not been previously presented or published in a conference.

Conflict of interest: No Conflict of interest

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Consent of the patient was obtained for publishing his case for promoting science.

References

1. Lichtenstein L. Polyostotic fibrous dysplasia. Arch Surg 1938;36:874-98. doi:10.1001/archsurg.1938.01190230153012.
2. Hartley I, Zhadina M, Collins MT, Boyce AM. Fibrous Dysplasia of Bone and McCune-Albright Syndrome: A Bench to Bedside Review. Calcif Tissue Int 2019;104:517-29. doi: 10.1007/s00223-019-00550-z.
3. Yang L, Wu H, Lu J, Teng L. Prevalence of Different Forms and Involved Bones of Craniofacial Fibrous Dysplasia. J Craniofac Surg 2017;28:21-5. doi: 10.1097/SCS.0000000000002830.
4. Campanacci M, Enneking WF. Bone and soft tissue tumors: clinical features, imaging, pathology and treatment, 2nd ed. New York, USA: Springer-Verlag Wien; 1999.
5. DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. J Bone Joint Surg Am 2005;87:1848-64. doi: 10.2106/JBJS.D.02942.
6. Hughes EK, James SL, Butt S, Davies AM, Saifuddin A. Benign primary tumours of the ribs. Clin Radiol 2006;61:314-22. doi: 10.1016/j.crad.2005.12.005.
7. Remotti F, Feldman F. Nonneoplastic lesions that simulate primary tumors of bone. Arch Pathol Lab Med 2012;136:772-88. doi: 10.5858/arpa.2011-0557-RA.
8. Weinstein LS, Chen M, Liu J. Gs(alpha) mutations and imprinting defects in human disease. Ann N Y Acad Sci 2002;968:173-97. doi: 10.1111/j.1749-6632.2002.tb04335.x.
9. van der Wal WA, Unal H, de Rooy JW, Flucke U, Veth RP. Fibrous dysplasia of bone associated with soft-tissue myxomas as well as an intra-osseous myxoma in a woman with Mazabraud's syndrome: a case report. J Med Case Rep 2011;5:239. doi: 10.1186/1752-1947-5-239.
10. Boyce AM, Turner A, Watts L, Forestier-Zhang L, Underhill A, Pinedo-Villanueva R, et al. Improving patient outcomes in fibrous dysplasia/McCune-Albright syndrome: an international multidisciplinary workshop to inform an international partnership. Arch Osteoporos 2017;12:21. doi: 10.1007/s11657-016-0271-6.
11. Snieders MN, van Kemenade FJ, van Royen BJ. Monostotic fibrous dysplasia of a lumbar vertebral body with secondary aneurysmal bone cyst formation: a case report. J Med Case Rep 2009;3:7227. doi: 10.4076/1752-1947-3-7227.
12. King RM, Payne WS, Olafsson S, Unni KK. Surgical palliation of respiratory insufficiency secondary to massive exuberant polyostotic fibrous dysplasia of the ribs. Ann Thorac Surg 1985;39:185-7. doi: 10.1016/s0003-4975(10)62564-7.
13. Mahadevappa A, Patel S, Ravishankar S, Manjunath GV. Monostotic fibrous dysplasia of the rib: a case report. Case Rep Orthop 2012;2012:690914. doi: 10.1155/2012/690914.
14. Traibi A, El Oueriachi F, El Hammoumi M, Al Bouzidi A, Kabiri el H. Monostotic fibrous dysplasia of the ribs. Interact Cardiovasc Thorac Surg 2012;14:41-3. doi: 10.1093/icvts/ivr048.
15. Ruggieri P, Sim FH, Bond JR, Unni KK. Malignancies in fibrous dysplasia. Cancer 1994;73:1411-24. doi: 10.1002/1097-0142(19940301)73:5<1411::aid-cncr2820730516>3.0.co;2-t.