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Thoracic Outlet Syndrome Caused by Fibrous Dysplasia of the First Rib

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Abstract

Fibrous dysplasia causing thoracic outlet syndrome is rare. A 41-year-old woman presented with neurogenic thoracic outlet syndrome with imaging that demonstrated a large tumor of her proximal left first rib. Transaxillary excision was unsuccessful due to involvement of the subclavian vasculature and brachial plexus. Subsequent posterolateral thoracotomy and resection of her first rib revealed fibrous dysplasia. Thoracotomy should be considered in these cases for optimal vascular control and identification of thoracic outlet anatomy.

Thoracic outlet syndrome (TOS) refers to symptoms that arise from the compression of the subclavian artery, vein, or brachial plexus as they traverse the thoracic outlet [1]. Fibrous dysplasia (FD) is a benign tumor marked by the abnormal replacement of normal bone and marrow with a fibrous connective tissue stroma. There is a predilection for rib involvement [2]. FD and other benign tumors of the chest wall are uncommon, and those causing TOS are even more so. We report the fifth case to our knowledge of FD of the first rib causing TOS and describe a successful thoracotomy approach for rib resection following attempted transaxillary approach with resolution of neurologic symptoms.

A 41-year-old woman was referred to our hospital with neurogenic TOS. Two years prior to presentation, she had been involved in a motor vehicle collision that resulted in no injuries other than some cervical disc disease. Her past medical history was otherwise insignificant. Since this incident, she developed anterior chest wall discomfort as well as progressive, constant pain and paresthesias in her left arm and hand. Prior to evaluation she was taking both narcotics and muscle relaxants for her symptoms with little relief.

Physical examination revealed a fixed, nontender, palpable mass in the left infraclavicular fossa. Adson's test results were negative, and peripheral pulse examination results were normal. Laboratory studies were unremarkable. Computed tomography (CT) scanning revealed a 4.5-cm tumor in her proximal first left rib (Figs 1A, 1B). Differential diagnosis of this lesion included fracture with healing callus from her prior motor vehicle crash, a benign or malignant primary tumor, or metastatic tumor of the bone.

She was initially evaluated and taken to the operating room by the vascular service for resection of the tumor because her symptoms had not improved with medical therapy and malignancy could not be excluded. A transaxillary approach was attempted but was

unsuccessful due to involvement of the subclavian vessels and inability to identify the brachial plexus. Biopsy of the lesion revealed fibrosis without malignancy.

Due to the inability to resect the tumor through a transaxillary approach, the thoracic service was consulted, and 2 weeks later the patient underwent exploration through a muscle-sparing posterolateral thoracotomy through the third intercostal space. After the scapula was retracted off of the chest wall, the scalene muscles were divided from their attachments to the ribs. The subclavian vessels and brachial plexus were isolated and dissected free from the tumor. The first rib was transected and isolated from posterior to anterior, dividing intercostal muscles and adhesions from the prior transaxillary incision. The dissection carried out past the tumor to the manubrial attachment, where it was disarticulated at the costochondral junction and the rib with tumor removed in entirety. The chest was closed in standard fashion with anterior and posterior thoracostomy tubes.

The remainder of the hospital stay was unremarkable and the patient was discharged home in good condition on the fourth postoperative day. Final pathology revealed fibrous dysplasia (Figs 2A, 2B). She was seen in clinic 1 month postoperatively with marked improvement in her neuropraxic pain.

Comment

There are three subtypes of TOS, classified on the basis of the particular portion of the neurovascular bundle that is involved [1]. The surgical goal of treatment is decompression of these structures in the cervicoaxillary canal to restore normal neurovascular function to the upper extremity.

Neurogenic TOS is the most common subtype, manifesting as neuropraxia of the affected upper extremity from the compression of the brachial plexus between the anterior and middle scalene muscles. Venous TOS leads to effort thrombosis of the axillary or subclavian veins as they are compressed between the anterior scalene and the first rib and clavicle. Arterial TOS, often associated with a cervical rib or other skeletal abnormality, can lead to stenosis, arterial thrombosis/thromboembolism, and upper extremity ischemia.

Benign tumors of the chest wall are far less common than primary or metastatic malignant rib tumors [3]. FD is the most common benign tumor, occurring in approximately 30% to 50% of cases, followed by osteochondroma, chondroma, aneurysmal bone cyst, and eosinophilic granuloma [4]. FD is a developmental skeletal disorder found in both monostotic (70% to 80%) and polyostotic (20% to 30%) forms, the latter of which can be associated with McCune-Albright syndrome and Jaffe-Lichtenstein syndrome. The pathophysiology of FD is related to a somatic missense mutation in the *GNAS1* gene on chromosome 20, which leads to inhibition of GTPase activity and abnormal fibrous proliferation. Among patients with FD, up to 20% of monostotic cases occur in the ribs and approximately 55% of those with polyostotic disease will have chest wall disease with the most common location being the lateral, posterior aspect of the second rib [3].

TOS caused by a tumor of the rib is rare and has been reported only 12 times in the literature over the past one and one-half centuries, with the majority of cases due to osteochondroma [7]. A variety of operative approaches have been used, including the interscapulovertebral route for posterior tumors and the thoracotomy, supraclavicular, and transaxillary approaches for anterior tumors.

There have been only four reports of FD causing TOS, and the optimal management of these lesions is unknown [5-8]. The first report was of a 27-year-old man with neurogenic TOS from a first rib tumor [5]. The tumor was removed via the subclavicular approach with a

medial clavicular resection after a failed transaxillary excision. The second case was a 31-year-old woman with both neurogenic and venous TOS who underwent successful transaxillary excision of monostotic FD [6]. The third case was a 25-year-old man with neurogenic and arterial TOS who underwent supraclavicular excision of the medial clavicle and first rib [7]. The final case was a 43-year-old woman with large, polyostotic FD [8]. Her first rib was removed through a transcervical-thoracic approach with resection of the medial clavicle. In all cases, removal of the tumor was associated with amelioration of symptoms.

Ours is the second case of thoracotomy for resection of FD causing TOS and the third case where a standard transaxillary approach failed. In cases of TOS due to rib tumor, consideration should be given to a thoracotomy approach to obtain vascular control and safely identify the brachial plexus prior to resection of the tumor and involved first rib. Although rare, TOS due to FD is amenable to resection to relieve the compressive symptoms of TOS.

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Fig 1.
Computed tomography scan demonstrating a large tumor of the left first rib. (A) Coronal view. (B) Axial view.

Fig 2. Histopathologic section of the resected specimen demonstrating fibrous dysplasia. (A) Diagnostic curvilinear trabeculae of woven bone surrounded by a fibroblastic proliferation. (Hematoxylin and eosin; $\times 10$.) (B) Cytologically bland moderately cellular fibroblastic proliferation. (Hematoxylin and eosin; $\times 40$.)