

# Concomitant Neurogenic and Vascular Thoracic Outlet Syndrome Due to Multiple Exostoses

## Abstract

We report a rare case of multiple hereditary exostosis where patient presented with bilateral base of neck exostoses with concurrent compression of brachial plexus and subclavian artery and vein. The patient was a young 26-year-old woman with chief complaints of pain in the left upper extremity, paresthesia in the left ring and little finger, and weakness in hand movement and grip. On referral, history, physical examination, radiological imaging, and electrodiagnostic tests evaluated the patient. Due to severe pain and disability in performing routine activities, surgical intervention was necessary. In the current case, the patient had thoracic outlet syndrome with concomitant venous, arterial, and neurogenic sub types. Radial pulse returned and pain associated with brachial plexus compression was resolved after the surgery.

**Keywords:** *Brachial plexus, multiple exostosis, thoracic outlet syndrome*

## Introduction

One of the compression neuropathies is thoracic outlet syndrome (TOS). There are two types of TOS, vascular, or neurogenic. Vascular type of TOS is divided into arterial and venous subtypes while neurogenic type of TOS is divided into true and disputed neurogenic.<sup>[1]</sup> Patients with vascular or neurogenic TOS present with an apparent mechanical obstruction to blood flow or with positive nerve conduction study tests.<sup>[2]</sup>

Multiple hereditary exostosis (MHE) is characterized by growths of multiple osteochondromas (benign cartilage-capped bone tumors that grow outward from the metaphyses of long bones). The prevalence of MHE is 1 in 50,000 in general population. Approximately, 15% of the exostoses are multiple and 62% cases have positive family history. The sporadic form is nearly 6 times more common than the MHE.<sup>[3]</sup>

Osteochondromas can cause reduction in skeletal growth, bony deformity, restricted joint motion, shortened stature, premature osteoarthritis, and compression of peripheral nerves.<sup>[4]</sup> In this case report, we present a case of MHE who developed TOS.

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## Case Report

The patient was a 26-year-old woman with chief complaints of pain in the left upper extremity, paresthesia in the left ring and little finger, and weakness in hand movement and hand grip. On physical examination, she was found to have clawing of the left hand, first web space atrophy, and interosseus muscles atrophy. Ulnar and radial pulses were absent. There were prominent venous collaterals around shoulder and upper chest [Figure 1]. Electrodiagnostic study was suggestive of compression of C8-T1 cervical nerves. X-ray neck and chest showed two large exostoses at the base of the neck in thoracic outlet bilaterally [Figure 2a].

The patient was taken for resection of exostoses under general anesthesia. Induction of anesthesia achieved with fentanyl 2 mcg/kg, midazolam 0.05 mg/kg, propofol 1.5 mg/kg, atracurium 0.5 mg/kg, and lidocaine 1 mg/kg. For maintenance of anesthesia, isoflurane 1 minimum alveolar concentration and infusion remifentanyl 0.1 mcg/kg/min was used. Surgical excision of mass with two-third part of the clavicle and the first rib was done [Figure 2b]. At the end of surgery, radial and ulnar artery pulsations were palpable. Patient was transferred to ICU and monitored for 24 h. Fortunately, in postoperative period,

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patient's pain had resolved and sensory deficit completely recovered [Figure 3a and b].

## Discussion

MHE is an autosomal dominant condition with multiple osteochondromas arising from the metaphysis of long tubular bones.<sup>[5]</sup> Mutations are seen in 1 and 2 exostosin genes. It is also called as multiple osteochondromatosis or diaphysealacclasis.<sup>[6]</sup> Exostoses are seen in all bones except skull, mandible, and facial bones. The common sites are around knee joint and humerus. The exostoses usually present as painless mass without any symptom, occasionally causes limb deformity or compression effects with vascular or neurological complications, and malignant transformation.<sup>[3]</sup>

TOS is a compression neuropathy. Diagnosis of TOS is clinical, but electrodiagnostic study plus routine imaging is helpful.<sup>[2]</sup> Arterial TOS is rare comprising around 1%–2% of all cases of TOS and the major etiology of arterial TOS is an osseous anomaly such as cervical rib. Subclavian artery may become thrombotic or aneurysmal and presentation may include fingertip ulceration, Raynaud's phenomenon, and claudication pain.<sup>[1,5]</sup> Venous TOS constitutes 2%–3% of all cases. These patients may develop thrombotic event, meanwhile there can be intermittent venous compression that after a long period of time leads to venous collateral development around the chest, breast, and shoulder.<sup>[2]</sup> True or electrically positive neurogenic TOS is uncommon; however, these patients have chronic nerve compression in C8-T1 territory. On physical examination, they have hypothenar muscle atrophy, decreased hand grip with sensory deficit. Electrically negative neurogenic TOS is very common and constitutes 95% of TOS cases. These patients have vague and nonspecific pain in the upper extremity, neck, arm and medial part of forearm, paresthesia in fingers plus they have negative electro diagnostic study.<sup>[7]</sup>

In the present case, there was involvement of both vascular and neurological component at the same time therefore, excision of the tumor bone was indicated and undertaken by vascular reconstructive surgeons. Indications for surgery in this patient were pain, absent peripheral pulse in the left upper limb, and weakness in the arm and hand movements. Surgical approach is classically by an incision through the neck with an extension on to the clavicle to access the subclavian artery and the brachial plexus.<sup>[3,6]</sup>

Surgery leads to relief of vascular and nerve compression resulting in relief of pain and recovery of sensory deficit in medial cord distribution area.

## Conclusion

The purpose of reporting this case is to highlight a rare case with concomitant occurrence of venous, arterial, neurogenic TOS due to huge exostosis. Postsurgery in the



Figure 1: Anterior aspect of patient showing prominent venous collaterals around upper chest

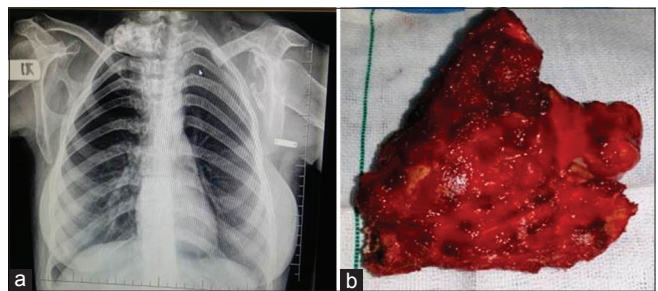


Figure 2: Postoperative X-ray imaging of two large exostoses at the base of neck in thoracic outlet (a) and exostosis mass after excision (b)

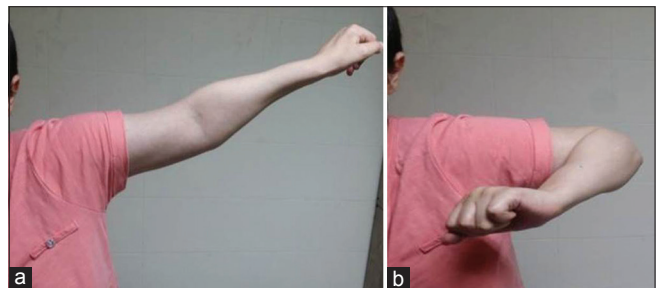


Figure 3: Shoulder abduction (a), Elbow flexion and finger motion (b) after successful surgery

follow-up period, patient did not have pain or neurological deficit.

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## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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