Abstract: Osteochondromas are the most common benign bone tumors occurring mostly in adolescent age group. Solitary Osteochondroma of proximal ulna are extremely rare, only a single case of Osteochondroma of proximal ulna with ulnar neuropathy was reported in the literature. We are presenting a rare case of Osteochondroma of proximal ulna with cubitus valgus deformity and tardy ulnar nerve palsy in a 12 year old girl. A provisional diagnosis was made based on clinical findings, x-ray, CT and MRI. The patient was operated, the osteochondroma was excised extraperiosteally and anterior transposition of ulnar nerve done. The sample was sent to histopathological examination and the diagnosis was confirmed. The patient was improved symptomatically at 6 months follow up with no residual clawing, no motor or sensory deficit but Nerve conduction study showed residual ulnar axonopathy.

Keyword: Osteochondroma, ulnar neuropathy, Extraperiosteal excision, anterior transposition.

Introduction: An osteochondroma is the most common type of benign bone tumor and accounts for 10-15% of all bone tumors and 20-50% of all benign bone tumors.1 Osteochondroma is a developmental lesion rather than a true neoplasm. "Cartilaginous exostosis" or "Osteocartilaginous exostosis", it is considered synonymous with osteochondroma. The tumor is often diagnosed as an incidental finding. Most are asymptomatic, but they can cause mechanical symptoms depending on their location and size. It is a benign cartilage-capped outgrowth, It arises from cartilage tissue in children and adolescents between the ages of 10 and 20. They generally occur at the end of the growth plates of the long bones, often around the joints. They most commonly form at the shoulder or the knee but have been known to occur in the long bones of the forearm (i.e. the radius and ulna). Malignant transformation occurs in the cartilage cap, and is uncommon in sporadic solitary osteochondromas (~ 1 %), whereas in the setting of hereditary multiple exostosis the rate is much higher (5-25%). Once a child has reached skeletal maturity, the osteochondroma typically stops growing. Ongoing growth and or pain after skeletal maturity has been reached is suspicious for malignant degeneration. We are presenting a rare case of osteochondroma of proximal ulna with cubitus valgus deformity and tardy ulnar nerve palsy.

Case report: A 12 year old girl presented with swelling of left elbow for one month duration. The swelling was insidious in onset, non progressive, not painful, no restriction of movements. There was no history of trauma. On examination diffuse bony swelling of size 3x 2 cm palpable over the medial aspect of proximal ulna below the medial epicondyle. Swelling was not warm, non tender, not mobile, bony hard in consistency. Positive findings include Valgus deformity of the left elbow with a carrying angle of 25 degrees ( Rt – 16o)(fig.1a,1b); No movement restriction with 0 to 130o of flexion ,85o pronation and 85o supination. Ulnar clawing was present with hypothenar muscle wasting(fig.1c). Weakness of interrosei, adductor pollicis were noted and the power was 4/5. The sensation was decreased over the little and ring fingers; Froment's sign, Card test and Egawa’s tests were positive.

Fig 1.pre operative x-ray 2a. AP view 2b. Lateral view
Blood investigations including Complete hemogram, ESR, CRP, ASO, RFT and Mantoux were within normal limits. X-ray of left elbow AP (fig.2a) and Lateral (fig.2b) showing an abnormal bony mass seen in the medial aspect of left proximal ulna. Exostoses of other sites were ruled out by skeletal survey (fig.2c, 2d, 2e). Nerve conduction study showed Left ulnar nerve axonal neuropathy (Distal amplitude = 1.43 micro volts and proximal amplitude = 1.09 microvolts) and the nerve conduction velocity was 64.25 m/s. During NCS, the stimulating and recording electrodes were kept at identical distances on both sides. The proximal stimulating electrode was kept at the level of ulnar groove and distal stimulating electrode at the level of wrist. The distance between them was 21 cm on both sides. The responses were measured by recording electrodes kept over the Abductor digiti minimi. Hence, the distance between the proximal stimulating electrode to recording electrodes was greater than that of distal stimulating electrode to the recording electrodes. The lesion was confirmed at the level of the elbow. The lesion was Sunderland Type 1 nerve injury (corresponding to Neurapraxia - Seddon classification). MRI with CT screening of left elbow showed Evidence of dysplastic or hypoplastic abnormal bone of size 1.4 X 1.3 cm (fig.3a, 3d) noted in the medial aspect of proximal left ulna with surrounding cartilaginous cap. The maximum cartilage cap thickness was 0.7 cm without any dispersed calcification or surrounding soft tissue mass (fig.3b, 3c). Cortex and medulla of the abnormal bone appears to be continuous with the host bone.

The above mentioned features suggestive of osteochondroma of left proximal ulna. Based on the clinical, x-ray & MRI we considered it as a benign lesion and did an extraperiosteal excision. We planned to do the deformity correction later, if the deformity increases. Moreover, the difference between the carrying angles of the normal side (16°) to that of affected side (25°) was less than 10 degrees without any movement restriction. The patient was operated under GA, with tourniquet control. The patient was placed supine; the affected limb was positioned on a forearm table with elbow in flexion and forearm in supination. Through medial approach, extra periosteal excision of osteochondroma (fig.4b, 4c, 4d) with anterior transposition of ulnar nerve (fig.4a) in the subcutaneous plane was done. We ensured adequate removal of the intended lesion intra-operatively. Hence, we sent it directly to Histopathology examination without any further imaging of the specimen. Post operative x-rays showed adequate removal of the lesion (fig.5a, 5b). The post operative period was uneventful and the patient was discharged on the 10th post-op day. Postoperatively, the elbow was immobilized in a above elbow slab at 90 degrees of flexion for 3 weeks. Histopathological examination (fig.6a, 6b) confirmed the diagnosis as osteochondroma (lobules of hyaline cartilage enclosing the bony trabeculae with intervening bone marrow elements). The ulnar clawing and intrinsic muscle strength improved at 2 months follow up (fig.7). At 6 months follow up, x-rays were normal (fig.8a, 8b), the patient had full range of movements (fig.9b), no residual clawing (fig.9a), no residual motor / sensory deficit, Froment's sign (fig.9c), card test (fig.9d) and Tinel's sign were negative. Six months post-operative nerve conduction study shows persistent left ulnar axonal neuropathy. There was a mild improvement in the nerve conduction study. The pre-operative distal amplitude value of left ulnar nerve was 1.43 microvolts which improved to 3.0 microvolts. Similarly, the pre-operative proximal amplitude of left ulnar nerve was 1.09 microvolts which improved to 2.0 microvolts. The post-operative Nerve conduction velocity was 62.14 m/s.
Discussion: In a study conducted by porter et al among 172 patients with HME, the incidence of exostosis in proximal ulna or olecranon was found to be 0.2%. In rare cases osteochondromas of proximal ulna or radius cause proximal radio ulnar diastasis.3  

radial head dislocation 4 or ulnar nerve neuropathy. Solitary osteochondroma of proximal ulna causing ulnar neuropathy is rare, only one case was reported in literature.5  

Symptomatic presentation is either due to mechanical effects of the lesion, fracture, or malignant transformation. The mechanical symptoms includes impingement upon the nearby structures causing nerve compromise, vascular compromise, reactive myositis. Other symptoms are palpable lump, bursal formation and bursitis. Fractures occur typically through the neck of the pedunculated lesions. In fewer than 1% of solitary osteochondromas, malignant degeneration of the cartilage cap into secondary chondrosarcoma has been described and is usually heralded by new onset of growth of the lesion, new onset of pain, or rapid growth of the lesion.

The average thickness of cartilage cap is about 0.6 cm, rare for it to exceed 1 cm. 6 Features of malignant degeneration of osteochondroma in CT and MRI include Development of thick, bulkiy cartilaginous cap in CT > 1 cm, MRI > 2 cm.7 There is a dispersed calcification within the cartilaginous cap and soft tissue mass development will be evident. It is very difficult to differentiate low graded well differentiated chondrosarcoma from benign osteochondroma even from histopathology. It subjects to a distressingly high degree of interobserver variability. The diagnosis rests on combination of clinical, radiological and cytologic features. In doubtful cases with suspicion of malignancy, Bone scan and Immunohistochemistry can be done. Bone scan will show increased uptake and IHC shows positivity for S-100, Estrogen receptors, Sox-9 and there is upregulation of PTHrP & BCL2.6,8

Neural involvement is rarely seen in solitary osteochondromas, most of the cases were reported in hereditary multiple exostoses. Neural involvement is due to pressure or stretching of the nerve fibres by the lesion. The commonly involved nerves are Peroneal nerve due to fibular head osteochondromas,9 Sciatric nerve due to femoral neck osteochondromas10 Ulnar nerve due to distal humerus or proximal ulnar osteochondromas5,3 Spinal nerve roots due to vertebral osteochondromas. 11 Ulnar nerve has no branches proximal to elbow and it has both fast and slow conducting nerve fibres. The reasons for low amplitude recordings in the proximal electrodes compared to the distal are, 1) The distance between the nerve and the skin surface greatly influences the amplitude of the evoked potential. Thus, a small potential may simply indicate a deeper location of the nerve over the area in question. Unless one is aware of this possibility, amplitude variability from one site of recording to another may lead to the erroneous conclusion that a conduction block exists.12 2) If all axons conducted at the same velocity, the amplitude would remain unchanged; but because they differ, slower-conducting axons activate the muscle progressively later with more proximal stimulation so that the CMAP becomes longer and lower. Partial CMAP cancellation of negative components by overlapping positive components results in only minimal reduction in area compared to amplitude at more proximal sites of stimulation.13 3) During routine motor studies, the CMAPs recorded by proximal and distal stimulations are nearly identical in configuration. If measured carefully, the proximal CMAP duration may increase slightly, and both the area and amplitude may fall slightly. In general, the larger fibres depolarize before the smaller ones. Likewise, there is a normal variation in the size of individual fibre action potentials, with larger fibres generally having larger amplitudes. Temporal dispersion occurs as this individual nerve fibres fire at slightly different times (i.e., larger, faster fibres depolarize before smaller, slower ones). Temporal dispersion normally becomes more prominent at proximal stimulation sites because the slower fibres progressively lag behind the faster fibres.14

Valgus deformity of elbow may cause ulnar nerve neuropathy, most commonly due to previous distal humeral fracture, growth abnormality of lateral condylar physis and rarely due to mass formation. Lesions like synovial chondromatosis, osteochondromatosis.15 In this condition the nerve is stretched along the distal end of the humerus, known as tardy ulnar nerve palsy. The entrapment of ulnar nerve in the cubital tunnel also causes palsy.5 This causes numbness and tingling, loss of sensation in ulnar nerve distribution. Also clawing, hypothenar wasting, Froment’s sign, card test, Egawa test may be present.

Symptomatic osteochondroma’s and cosmesis are the major indications for surgery. Extraperiosteal excision16 along with removal of cartilage cap and perichondrium is recommended to prevent recurrence. Ideally, the line of resection should be through the base of the stalk; thus, the entire lesion is removed en bloc with its fibrous covering. Atypical or very large lesions should be investigated fully to exclude the remote possibility of malignancy. In the skeletally immature patient, care must be taken to avoid damage to the growth plate during the exposure and resection of the lesion. In case of suspicion of malignancy wide local resection should be done. The ulnar nerve runs just posterior to the medial epicondyly in the cubital tunnel. Decompression with anterior transposition17 usually is the operation of choice for ulnar nerve compression at the elbow because it removes the nerve from its compressive bed and puts it in one that is more suitable. By transferring the nerve anteriorly, it effectively lengthens the nerve, decreasing tension on it in flexion. An incision begins 5 cm above the elbow joint centered over the medial epicondyly and extending downwards. Once the nerve has been visualized the distal portion of the medial intermuscular septum, the fibroaponeurotic roof of the epicondylar groove, the Osborne ligament, and the fascia of flexor carpi ulnaris are incised, freeing the ulnar nerve. The nerve is positioned beneath the subcutaneous tissue and held to the muscle fascia with a few sutures through the epineurium. Postoperatively, the elbow is immobilized in a cast or splint at 90 degrees of flexion for 3 weeks.

Conclusion: Solitary osteochondroma of proximal ulna causing pressure on the ulnar nerve should be promptly removed. Before any surgery, plain X-Ray skeletal survey is a must and if suspected further imaging with CT or MRI is to be ordered.

Bibliography

3. Peterson HA. Physieal Injury Other Than Fracture; 2012, P 133.