Case Report

Secondary Chondrosarcoma of Clavicle Due to Multiple Hereditary Exostosis in Adolescent

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Abstract

Approximately 0.5% bone tumor occurs in clavicle. Primary chondrosarcoma is very rare even among clavicle tumors. Secondary Chondrosarcoma is reported with variable incidence. It is the third most common primary tumor arising from bone. Surgery is the only treatment option in Chondrosarcomas with excellent prognosis. We report a case of 14 years old male patient history of multiple hereditary exostosis largest over clavicle with restricted shoulder and neck movements previously operated elsewhere two and half years back. We evaluated the patient with plain radiographs of shoulder, chest, neck, CT scan of chest, neck, and shoulder MRI with contrast and technetium scan for skeletal metastasis. CT angiography was done for vascular involvement. Multidisciplinary team involved before the surgery. Total claviculectomy was done. Postoperatively patient recovered uneventful. After one year follow-up his shoulder movements flexion, extension, adduction and internal rotation normal except abduction up to 90 degree.

Keywords: clavicle; chondrosarcoma; multiple hereditary exostosis; secondary.

Introduction

Tumors of clavicle are rare; most of them are primary malignant lesions [1-2]. Pelvis and trunk bones are common sites for Chondrosarcomas; while secondary chest wall chondrosarcoma are relatively common as compared to primary Chondrosarcomas [3-4]. Chondrosarcomas are more prevalent in adults than in children (being more common in males than females), they are more common in people older than 40. They occur mainly in pelvis, femur, humerus and scapula. Chest wall Chondrosarcomas originate from ribs in 80% of cases, the rest arises from sternum. Clavicle and scapula are less likely to be the origin of chondrosarcoma [5-6]. In multiple hereditary exostosis, multiple cartilage-capped Exostoses develop during childhood and ossify when skeletal growth is complete [7]. Osteochondroma is a cartilage capped bony projection arising on the external surface of bone containing a marrow cavity that is continuous with that of the underlying bone [8]. Since Chondrosarcoma do not respond well to radiotherapy or chemotherapy, surgery is mainstay of treatment. Although intermediate- and high-grade tumors have a survival rate of 40% to 60%, low-grade tumors result in long-term survival in as much as 90% of patients [9].

Case Report

A 14 year boy resident of swat presented with swelling in right supraclavicular region for last two and half years. He also had multiple bony swelling with deformity around both wrists and around both knee since childhood. Initially he treated at swat than referred to tertiary care public sector hospital where he underwent trucut biopsy and then intrasional surgery was done along with biopsy which revealed Chondrosarcoma. After the surgery, swelling gradually increased in size. Examination of this patient revealed swelling of about 20x12 cm on right supraclavicular region extending from sternoclavicular joint medially to right shoulder joint laterally and suprascavicular fossa posteriorly, there also was presence of linear scar of previous surgery. On palpation, there was a lobulated swelling in right clavicular region firm to hard inconsistency, scar was adherent to underlining structures and he had restricted movements at right shoulder mainly abduction and restricted neck lateral bending on right side. Distal neurovascular intact.
Hematological investigations were in normal limits. Plain radiograph of chest showed osteoblastic lesion in right clavicle extending from right sternoclavicular joint to lateral end of clavicle with narrow zone of transition, punctuate calcification and soft tissue extension. MRI done for extension of lesion and neurovascular involvement. CT CHEST done for metastasis which turns out negative. The nuclear medicine bone scan reveals active bone pathology in right clavicle and multiple exostosis around both knees, wrist and ribs. CT angiography done shows mass not invading major vessels.

**Figure 1:** Clinical radiograph of patient with massive swelling over right clavicle with previous transverse surgical scar mark

**Figure 2:** Plain radiograph of chest (a) showing swelling over right shoulder with mixed density lesion and punctuate calcification, (b) plain radiograph of both wrist joint and radius ulna showing exostosis in distal radius and ulna.

**Figure 3:** CT scan shoulder and chest showing extent of lesion
Surgery was planned after detailed discussion with multiple departments including E.N.T., Cardiothoracic, Plastic and Vascular Surgery. We did marginal resection of tumor with total clavicectomy and specimen was sent for frozen section and histopathology. Frozen section revealed tumor free margins and final histopathology shows moderately differentiated Chondrosarcoma.

Postoperatively recovery was smooth. Patient developed seroma over the wound which was drained. Patient had improved range of motion of right shoulder and neck. After one year follow-up his shoulder movements flexion, extension, adduction and internal rotation normal except abduction up to 90 degree.

Figure 6: clinical photograph of patient (a) preoperative with incision marked (b) per operative photograph (c) specimen photograph after excision.

Figure 7: post operative plain radiograph of patient after total claviculectomy

Figure 8: Post operative clinical photograph of patient after total claviculectomy showing full range of motion at shoulder joint.
Discussion

Chondrosarcoma is the most common primary malignant bone tumor in patients older than 25 years. Treatment of Chondrosarcoma is predominantly surgical because adjuvant chemotherapy and radiotherapy are generally ineffective with the exception of dedifferentiated subtypes [9]. Risk of secondary chondrosarcoma is variable from 1% to 25% due to multiple hereditary exostosis. [10] Sudden increase in size of tumor and pain in previously asymptomatic lesion are signs of malignant transformation. An increase in the thickness of the cartilaginous cap of more than 1cm on MRI should raise a suspicion of Chondrosarcoma. The prognosis of secondary Chondrosarcoma depends on its histological grade. The 10 year survival rates are 83% for chondrosarcoma grade I as compared to 29% for grade III chondrosarcoma [11]. About 95% present insidiously with recent increase in the size of previous present mass or recent pain, radiographically Chondrosarcomas is expansile, mixed sclerotic (represent chondroid matrix) and centrally lucent lesion, with a narrow zone of transition, and possible thin sclerotic margin. CT scan better defines better detects subtle tumor matrix and MRI is the preferred imaging modality and determine extent of marrow involvement and soft tissue invasion [12]. Wide excision is the treatment of choice for grades 2 and 3 chondrosarcoma. For grade 1 chondrosarcoma, intralesional curettage followed by local adjuvant therapy (phenol, liquid nitrogen) and filling the cavity with bone grafts achieves satisfactory outcome [13].

References


