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Case Report

Surgical management of ventral scapular osteochondroma in a pediatric patient with multiple hereditary exostoses: a case report

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ABSTRACT

Multiple hereditary exostoses (MHE) is a rare autosomal dominant disorder caused by EXT1/EXT2 gene mutations, characterized by multiple osteochondromas arising from the metaphyses of long and flat bones. Scapular osteochondromas are uncommon, representing only 3-4.6% of all cases, and ventral (subscapular) lesions are exceedingly rare with limited surgical literature. A 12-year-old male with no family history of MHE presented with a one-year history of progressive, hard swelling over the right scapular region along with bilateral wrist, foot, and right arm swellings. He reported pain on overhead activities and scapular movement, with normal scapulohumeral rhythm and no neurological deficits on examination. Imaging confirmed a pedunculated osteochondroma on the ventral surface of the right scapula and multiple additional lesions consistent with MHE. He underwent complete surgical excision via a posterior approach through the triangle of auscultation, with rhomboid muscle splitting. Histopathology confirmed benign osteochondroma. At 1-year follow-up, the patient was completely asymptomatic with full shoulder range of motion and a QuickDASH score of 0. No recurrence was detected on follow-up imaging. This case demonstrates that symptomatic ventral scapular osteochondromas in MHE can be successfully managed via a posterior approach, with excellent functional outcomes.

Keywords: MHE, Ventral scapular osteochondroma , Paediatric

INTRODUCTION

Multiple hereditary exostoses (MHE) is a rare genetic disorder affecting approximately 1 in 50,000 individuals, inherited through mutations in the EXT1/EXT2 genes with nearly 100% penetrance.[1-3] Osteochondromas typically arise from long bone metaphyses and are detected in early childhood.

While shoulder osteochondromas occur in approximately 85% of MHE patients, true scapular involvement is rare, representing only 3-4.6% of all cases.[1,2] Patients with scapular lesions frequently carry the EXT1 mutation and have a heavier exostosis burden.[1] Ventral scapular osteochondromas are particularly uncommon, presenting with painful shoulder motion and mechanical snapping.[5,6] They carry a 2-5% risk of malignant transformation, reportedly higher than long bone lesions.[4]

Given the paucity of literature on ventral scapular osteochondromas, we report a case of successful surgical excision in a 12-year-old male with MHE, with excellent functional outcome at one year.[1]

CASE REPORT

History

A 12-year-old male presented with a one-year history of insidious onset, gradually progressive, hard swelling over the right scapular region, first noticed by his parents. He reported pain localized to the right scapula, aggravated by

overhead activities and weight-bearing and relieved by rest. Additional bony swellings were noted over the bilateral wrists, feet, and right proximal arm. There was no history of trauma, fever, or constitutional symptoms. Notably, there was no family history of MHE or similar skeletal disorders.

Physical examination

Multiple firm, non-tender bony swellings were palpated over the bilateral wrists, feet, and right arm. No mass was palpable over the dorsal aspect of the right scapula. Shoulder range of motion was full in all planes; however, pain was reproduced on overhead abduction and forced scapular protraction and retraction. Scapulohumeral rhythm was intact throughout the range of motion and was associated snapping of scapula and crepitus (Figure 1 A and B). There was no scapular winging, overlying skin changes, or neurovascular deficit in the upper extremity.

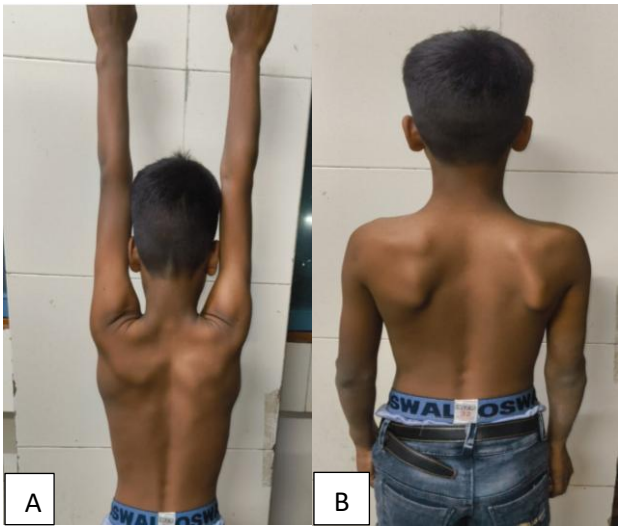

Figure 1 (A and B): Clinical picture of preoperative shoulder range of motion.

Investigations

Plain radiographs of the right shoulder revealed a pedunculated bony lesion on the ventral scapular surface (Figure 2 A and B).

Magnetic resonance imaging (MRI) confirmed a pedunculated osteochondroma projecting anteriorly from the inferoventral surface of the right scapula with medullary continuity between the lesion and the native scapula. Cartilaginous cap thickness on MRI was within normal limits, and there were no features of malignant transformation.

Imaging of the bilateral extremities demonstrated additional pedunculated and sessile osteochondromas at the metaphyses of the distal femur, proximal tibia, and distal radius bilaterally, establishing the diagnosis of MHE.

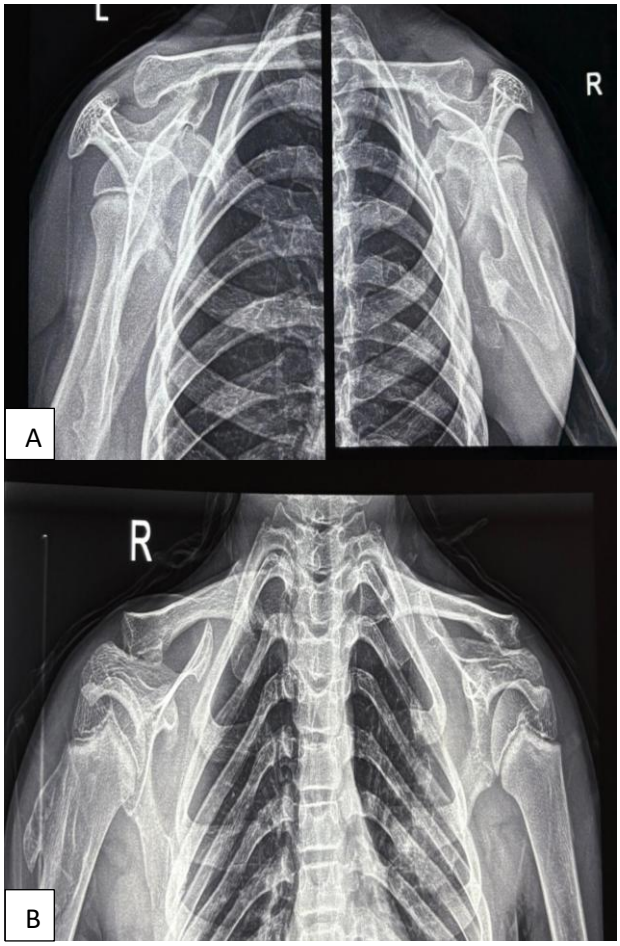

Figure 2: Plain radiographs of bilateral shoulders with scapila (A) scapular Y view and (B) anteroposterior view.

Surgical treatment

Given the symptomatic presentation with functional limitation, surgical excision was planned. Under general anesthesia in the prone position, a longitudinal incision was marked over the triangle of auscultation, bounded by the trapezius superiorly, latissimus dorsi inferiorly, and the medial border of the scapula laterally (Figure 3). The incision was deepened through subcutaneous tissue, and the rhomboid major muscle was gently split along its fiber direction to expose the subscapular surface. Neurovascular structures were carefully protected throughout the dissection.

The pedunculated osteochondroma was identified on the inferoventral scapular surface (Figure 4). The mass was completely excised using an osteotome and bone nibblers, ensuring removal flush with the native scapular cortex. The base was smoothened to reduce the risk of recurrence. Hemostasis was achieved, the wound irrigated with normal saline, and the rhomboid fibers reapproximated with absorbable sutures. Layered closure of subcutaneous tissue and skin was performed. The excised specimen was sent for histopathological examination.

Figure 3: Intraoperative surface marking of incision.

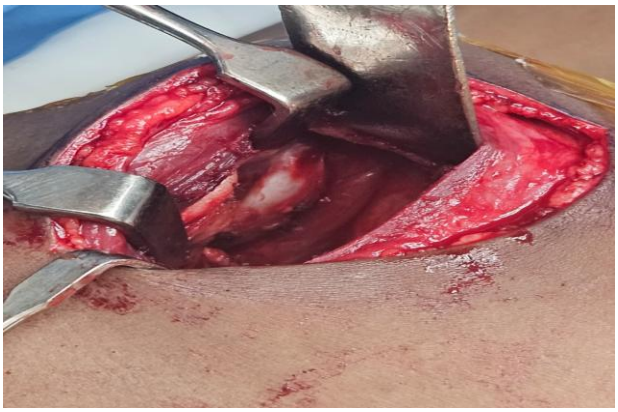

Figure 4: Intraoperative image showing osteochondroma on the ventral aspect of the scapula.

Outcome

Histopathology confirmed benign osteochondroma with no evidence of malignant transformation. The arm was immobilized in a sling for 2 weeks, followed by physiotherapy with progressive range-of-motion and strengthening exercises. At the 6-month follow-up, the patient was completely asymptomatic. Shoulder range of motion was full and symmetric bilaterally. The QuickDASH score was 0, indicating no functional limitations. Follow-up radiographs confirmed complete excision with no evidence of recurrence.

DISCUSSION

MHE is driven by loss-of-function mutations in EXT1 or EXT2, leading to aberrant endochondral ossification and formation of cartilage-capped bony projections at metaphyseal regions.[3] Patients with scapular involvement tend to carry EXT1 mutations and have a greater overall burden of exostoses.[1] Scapular osteochondromas account for only 3% of all lesions in MHE cohorts, and ventral lesions are even rarer, limiting surgical experience.[1,5]

The clinical presentation of ventral scapular osteochondroma is distinct from dorsal lesions and can be diagnostically challenging. Because the lesion lies on the subscapular surface, there is typically no visible or palpable dorsal mass as observed in our patient. Instead, the bony projection mechanically disrupts normal scapulothoracic gliding, producing the hallmark feature of snapping scapula syndrome (scapulothoracic crepitus).[2,7] This snapping is often audible and palpable at a reproducible arc of shoulder elevation, during protraction and retraction, and may be painful or initially painless. As the lesion enlarges or secondary bursitis develops, pain typically becomes the predominant complaint. Our patient exhibited pain on overhead abduction and scapular protraction/retraction with preserved scapulohumeral rhythm, a presentation characteristic of a ventral mechanical obstruction.[5-7]

A secondary subscapular bursa frequently develops around the lesion as a protective response to chronic mechanical irritation. This bursa can enlarge independently, become inflamed, and contribute to pain and restricted scapulothoracic motion, even when the underlying lesion is relatively small.[1,10] Mouradian et al found a large bursa that had effectively isolated the osteochondroma from surrounding structures, including the adjacent second rib, facilitating safe resection.[1] Pseudowinging, posterior displacement of the scapula by the underlying mass may also be observed and must be distinguished from true neurogenic winging due to long thoracic or accessory nerve involvement.[7,9]

In rare cases, larger or anteriorly projecting lesions can cause neurovascular complications, including long thoracic nerve compression, which can lead to true scapular winging; suprascapular nerve entrapment, which can lead to rotator cuff weakness; and brachial plexus irritation.[9] Thoracic complications such as rib erosion, hemothorax, or pneumothorax have been reported in very large lesions that cause mass effect on the chest wall.[1,6] Delayed diagnosis therefore risks progression to these complications, reinforcing the value of early imaging and intervention.[2]

The posterior approach through the triangle of auscultation, as utilized in this case, provides reliable access to the ventral scapular surface by splitting the rhomboid muscles along their fibers, preserving function and vascularity.[1,2] This avoids division of major muscle bellies and reduces postoperative morbidity. Mouradian et al similarly used a dorsal approach with trapezius and rhomboid division for a ventral scapular osteochondroma in an adult MHE patient, reporting a short hospital stay and full functional recovery.[1]

Regarding malignant transformation, scapular osteochondromas carry a higher risk than long bone lesions. Concerning features include new pain after skeletal maturity, rapid growth, cartilage cap >2 cm in adults, and irregular margins.[4,8]

Confirmed benign histopathology was reassuring; however, long-term surveillance remains essential given his young age.[3,4]

Notably, our patient had no family history of MHE despite its autosomal dominant inheritance pattern, suggesting a de novo mutation. This underscores the importance of considering MHE in any pediatric patient presenting with multiple osteochondromas, even in the absence of a positive family history.[3,12]

CONCLUSION

Ventral scapular osteochondroma is a rare but clinically significant manifestation of MHE. Symptomatic lesions with functional limitation warrant surgical excision. The posterior approach through the triangle of auscultation provides safe, effective exposure with minimal morbidity. Complete excision with base smoothening is essential to prevent recurrence. With appropriate technique and postoperative rehabilitation, excellent functional outcomes are achievable. Long-term surveillance is recommended given the elevated risk of malignant transformation in scapular lesions.

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