Intraosseous Schwannoma of the Upper Extremity
A Single Institutional Experience and Review of Literature

Nandlal Bharwani¹, Abhijeet Ashok Salunke¹,³, Dhruv Patel¹, Ritesh Suthar², Keval Patel¹, Ishan Arora¹, Ashok Govada¹, Shashank Pandya¹

¹Department of Surgical Oncology, The Gujarat Cancer and Research Institute, Ahmedabad, India
²Department of Nuclear Medicine, The Gujarat Cancer and Research Institute, Ahmedabad, India

Abstract

Introduction
Schwannomas are benign soft tissue tumors of neural origin, predominantly occurring in the head and neck regions due to their rich innervation. Intraosseous Schwannoma (IOS) is an exceedingly rare form of Schwannoma. This study aims to enhance the understanding of intraosseous Schwannoma by reviewing cases affecting the upper extremity bones and providing a detailed analysis of their radiographic and magnetic resonance imaging (MRI) characteristics along with a review of the literature on these rare tumors.

Material and Methods
A total of three patients with IOS in upper extremity bones were identified and analyzed. Radiographs and MRI scans were available for all patients. A comprehensive literature review was conducted, including case reports, retrospective studies, and reviews of published data. The epidemiology, anatomical distribution, radiographic characteristics, histological findings, and therapeutic outcomes of intraosseous schwannoma were all investigated, identifying 31 documented cases of IOS involving extremity bones.

Results
Intraosseous Schwannomas primarily affect the mandible, followed by the sacrum and vertebrae. Patients frequently present with non-specific symptoms such as localized pain, swelling, or neurological impairments, which can lead to delayed diagnosis. Radiographic evaluation of IOS typically reveals lytic lesions with well-defined, expansile features and thin sclerotic rims. MRI findings showed that IOS lesions appeared low to iso-intense on T1-weighted images.
and hyper-intense on T2-weighted images. These imaging characteristics are crucial for differentiating IOS from other lytic bone lesions. Histologically, the presence of Antoni A and Antoni B patterns, as well as S-100 protein positivity, confirms the diagnosis. Surgical management, consisting of curettage, provides a favorable prognosis and low recurrence rates.

Conclusion
Despite its rarity, intraosseous Schwannoma should be considered in the differential diagnosis of well-defined, expansile lytic bone lesions, particularly those with thin sclerotic rims. This review provides the most comprehensive analysis to date of IOS affecting extremity bones, emphasizing the importance of recognizing this entity in clinical practice.

Keywords: Intra-osseous Schwannoma, IOS, Upper Extremity, Upper Limb Tumors, Schwannoma of Bones

Introduction
Schwannoma, also known as neurilemmoma, is a benign soft tissue tumor that arises from the Schwann cells of the neural sheath. (1) It accounts for nearly 5% of all benign soft tissue tumors. The most affected regions are the head and neck, which are rich in spinal and cranial nerve supply. (2,3)

Intraosseous Schwannoma (IOS), a rare form of schwannoma, constitutes only 0.2% of all primary bone tumors. (3) The proposed mechanism for its intraosseous origin is the proximity of nerve fibres to blood vessels within the medullary canal of the bone.

IOS can affect bones through three possible pathways:
(a) direct origin from within the bone
(b) origin from the nutrient canal
(c) from nearby soft tissue leading to bony erosion. (4,5)

The head and neck region, particularly the mandible, is most commonly involved, followed by the maxilla, petrous bone, spine, and, less frequently, long bones such as the tibia, fibula, radius, and ulna. (6,7)

IOS is often associated with conditions such as Carney syndrome or Neurofibromatosis-1 (NF-1) (8,9). Given the rarity of this entity, histopathological examination is crucial for a definitive diagnosis (9).

In this study, we present three cases involving the 5th metacarpal, proximal ulna, and scapula to demonstrate the clinical and radiological features and treatment options for Intraosseous schwannoma (IOS). To the best of our knowledge, the literature on IOS is limited to case reports and small case series. This case series aims to highlight the various bones of the extremities affected by this rare lesion, differentiate it from other bone lesions, and discuss the best available treatment options.

Case Presentation
Case 1:
A 20-year-old male presented to an outpatient clinic with pain and swelling around his right shoulder, persisting for six months. The swelling had progressively worsened over the past one and a half months. He had no family history of similar issues, no history of trauma, and no medical comorbidities.

Clinical Examination:
Clinical examination revealed swelling on the dorsum of the little finger with tenderness on pressure. There was no warmth, redness, or neurovascular deficit observed around the medial aspect of the hand.

Imaging:
- X-rays (Anteroposterior and Lateral Views)
  Showed a well-defined lytic lesion with a cortical breach on the medial border of the shoulder. No periosteal reaction, calcification, or abnormal soft tissue was observed. (Fig.1 A)
- Magnetic Resonance Imaging (MRI):
  Revealed an intraosseous lesion involving the entire scapula, extending into the soft tissue on both the dorsal and volar aspects of the shoulder. The lesion displayed similar signal intensity to surrounding muscle on T1-weighted images, but brighter signal intensity on T2-weighted images.

Biopsy:
A core needle biopsy was performed in the planned incision line. Histopathological examination showed spindle-shaped cells arranged in fascicles. Both Antoni-A and Antoni-B cells,
characteristic of a schwannoma, were present. There were no signs of necrosis, excessive mitosis, or abnormal nuclei.

**Immuno-histochemistry (IHC):**
Immunohistochemical staining for S-100 and SOX10 proteins was performed, showing positive staining throughout the sample, further supporting the diagnosis of schwannoma.

**Treatment:**
Surgery was planned, and a posterior skin incision was made to perform a total scapulectomy. The proximal humerus was then fixed to the clavicle using prolene mesh and non-absorbable sutures (Fig.1B). Post-surgery, the shoulder was immobilised for 3-4 weeks, followed by physical therapy to gradually restore abduction, adduction at the shoulder joint, and flexion and extension movements at the elbow joint.

**Follow-up:**
The patient was monitored both clinically (through physical examination) and radiologically (with X-rays or other imaging). Follow-ups were conducted monthly for the first 3 months and then every 3 months for 1 year. At 60 months post-surgery, there were no signs of recurrence (Fig.1 C).

**Case 2:**
A 27-year-old woman presented to an outpatient clinic with a complaint of pain and swelling around her right elbow for six months. The symptoms had gradually worsened over the past month. She had no family history of similar issues, no history of trauma, and no medical comorbidities.

**Clinical Examination:**
The clinical examination revealed swelling on the posterior aspect of the elbow, with tenderness on pressure. There was no warmth, redness, or neurovascular deficit observed around the elbow joint.

**Imaging Studies:**
- X-ray (Anteroposterior and Lateral View):
  Showed a soap bubble-appearing lytic lesion with ill-defined margins in the proximal epiphysis of the ulna. (Fig.2A) There was no associated cortical breakdown or soft tissue involvement. Based on this finding, a giant cell tumor was a differential diagnosis.
- Magnetic Resonance Imaging (MRI):
  Showed a well-defined lesion with iso- to hypointense signal on T1-weighted images and hyper-intense signal on T2-weighted/STIR images. No cortical breakdown or soft tissue involvement was noted (Fig. 2B,C,D).
  - Radionuclide Sestamibi Scan: Performed to rule out brown’s tumor due to an elevated parathyroid hormone (PTH) level of 84.1 pg/ml. The scan was negative, and no parathyroid adenoma was found.

**Histopathological Examination:**
A core needle biopsy was performed along the line of a future incision. The histopathological examination revealed spindle cells arranged in bundles and fascicles, with the presence of both Antoni-A and Antoni-B cells. There was no necrosis, mitosis, or nuclear atypia. Immuno-histochemistry for S-100 and SOX10 showed diffuse positivity.

**Surgical Intervention:**
Surgery was planned using a posterior approach to the olecranon for extended curettage. The defect was filled with calcium hydroxyapatite crystals. The elbow was immobilised in a slab for four weeks. Physiotherapy was initiated to regain movement in the elbow joint, starting with flexion and extension, followed by pronation and supination.

**Follow-Up:**
Follow-up was conducted both clinically and radiologically. The patient was reviewed monthly for the first three months, followed by every three months for one year. At 15 months post-surgery, there were no signs of recurrence based on clinical examination and radiograph (Fig. 3 A,B).

**Case 3:**
A 24-year-old male presented to an outpatient clinic with complaints of pain and swelling around his right little finger for four months. The swelling had gradually worsened over the past two months. There was no family history of similar issues, no history of trauma, and no medical comorbidities.

**Clinical Examination:**
Clinical examination revealed swelling on the dorsum of the little finger, with tenderness on pressure. There was no warmth, redness, or neurovascular deficit observed around the medial aspect of the hand.

**Imaging Studies:**
- X-rays (Anteroposterior and Oblique Views):
  Showed a well-defined lytic lesion with cortical break involving the head of the fifth metacarpal bone. (Fig.4A) Additionally, there was a dislo-
cation of the joint between the metacarpal and the first phalanx. There was no periosteal reaction, calcification, or abnormal soft tissue observed on the X-ray.

- Magnetic Resonance Imaging (MRI):
  Revealed an intraosseous lesion involving the head of the fifth metacarpal. The lesion extended into the soft tissue on both the dorsal and volar aspects of the metacarpal. The lesion exhibited similar intensity to surrounding muscle on T1-weighted images and a brighter intensity on T2-weighted images (Fig.4B,C,D).

**Histopathological Examination:**
A core needle biopsy was performed along the planned incision line. Microscopic examination of the tissue showed spindle-shaped cells arranged in fascicles. Both Antoni-A and Antoni-B cells, characteristic of a specific type of tumor, were present. There were no signs of necrosis, mitosis, or abnormal nuclei. Immunohistochemical Staining (IHC) for S-100 and SOX10 proteins was performed, revealing positive staining throughout the sample, which supported the diagnosis.

**Treatment:**
Surgery was planned using a dorsal approach to access the metacarpal bone for extended curettage. Postoperatively, a cock-up splint was applied to the hand for four weeks to immobilise it. Physical therapy was then initiated to gradually restore flexion and extension movement of the metacarlo-phalangeal joint of the little finger.

**Follow-Up:**
The patient was monitored both clinically through physical examination and radiologically with imaging to track progress. Follow-up visits were conducted monthly for the first three months, followed by every three months for a year. At 13 months post-surgery, there were no signs of recurrence (Fig.5).

**Results**
In this study, three patients were diagnosed with intraosseous Schwannoma (2 males and 1 female). The study showed a male predilection, with the mean age of the group being 23.6 years (range 20-27 years). The presenting symptoms were pain and swelling in all cases. The mean follow-up was 17 months. Extended curettage was performed in 2 out of 3 cases (66.6%), and total scapulectomy was performed in 1 out of 3 cases (33.3%). There were no complications, and none of the patients had recurrence at the last follow-up.

**Review of Literature**
We conducted a search of PubMed and Google Scholar using the keywords "intraosseous schwannoma", "extremity bones", and "long and short bones". Our search covered publications from 2010 to 2023 and included all types of studies (1-41).

**Discussion**
Schwannoma, or neurilemmoma, is a benign tumor originating from Schwann cells, typically arising in sensory nerves due to the higher number of Schwann cells surrounding the axons. In 1923, Masson coined the term ‘Schwannoma’ to distinguish these from neumomas and other peripheral nerve sheath tumors. The involvement of intraosseous regions in Schwannomas is rare. A study by Knight et al. reviewed 234 cases and found 170 cases (73%) in the upper limbs, 64 cases (27%) in the lower limbs, and only six cases (2.6%) within bone or muscle.

This case series presents the clinical and imaging findings of three cases. (Table 1) We combined our findings with those from existing literature to enhance understanding of the clinical and radiological features and to guide the management of these rare entities. The most common presenting symptoms are pain and swelling around the affected area, with a long history and slow onset. Some reports also mention sensory or motor involvement and pathological fractures as presenting complaints. The usual age of presentation is between the 2nd and 5th decade of life, with females more commonly affected. In our study, we evaluated 35 patients with a female-to-male ratio of approximately 3:1 (25:9).

We conducted a search of PubMed and Google Scholar using the keywords "intraosseous Schwannoma", "extremity bones", and "long and short bones" and our search showed 31 studies on this topic (Table 2). The most commonly affected long bones in our study were the femur, followed by the tibia, and then the humerus and scapula. Radiographically, intraosseous schwannoma appear as well-defined osteolytic lesions with a lobulated appearance, a thin rim of sclerosis, a narrow zone of transition, and cortical expansion or erosion. MRI findings show that these lesions are hypointense to isointense relative to surrounding tissues on T1-weighted images and hyperintense on T2-weighted images. A characteristic MRI feature is the "target sign", where Antoni-B cells (hypo-cellular) surround Antoni-A cells (hyper-cellular), though this is more common in soft tissue schwannoma. None of our cases exhibited the target sign.
Due to their nonspecific symptoms and rarity, diagnosing these tumors can be challenging. Histopathological examination (HPE) and immunohistochemistry (IHC) are crucial for diagnosis. HPE typically reveals Antoni-A and Antoni-B areas with spindle cells, and IHC shows S-100 protein positivity, which is characteristic of Schwannoma.

The treatment of choice for these tumors is curettage, followed by filling the defect with bone graft, allograft, or bone cement. In cases presenting with pathological fractures, fixation is also required along with curettage and defect filling. These tumors generally have a good prognosis, with only one case of malignant transformation reported to date. Local recurrence is commonly due to incomplete removal, whereas complete removal usually prevents recurrence.

The present study has limitations due to its retrospective design and small sample size; however, it can aid researchers in developing a multicenter investigation.

**Conclusion**
In this series, we evaluated three patients along with 31 cases involving upper and lower limb bones. To the best of our knowledge, this is the most comprehensive collection of isolated appendicular skeleton bones affected by IOS.

Whenever radiographs reveal a lobulated area with an osteolytic lesion surrounded by a sclerotic rim, IOS should be looked into as a potential differential diagnosis, nonetheless it is uncommon. On magnetic resonance imaging, the lesion appears isointense to hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences in comparison to the surrounding tissue.

Histopathological examination (HPE) must be performed for a definitive diagnosis, which reveals typical Antoni A and B cells positive for S-100 protein. The most appropriate treatment for this rare tumor is curettage followed by filling the defect with bone graft, bone graft alternatives, allografts, or bone cement.

**Conflict of interest:**
The authors declare that there were no conflicts of interest within the meaning of the recommendations of the International Committee of Medical Journal Editors when the article was written.

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Fig. 2: (A) Anteroposterior and lateral views of the elbow joint show a lytic lesion with a sclerotic margin involving the proximal part of the ulna. (B, C, D) Sagittal and axial sections of magnetic resonance imaging of the elbow joint show an expansile lytic lesion involving the proximal part of the ulna with a medial cortical breach.

Fig. 3: (A) Radiograph showing antero-posterior view at 1 year follow-up following curettage of right proximal ulna intraosseous schwannoma. (B) Radiograph showing lateral view at 1 year follow-up following curettage of right proximal ulna intraosseous schwannoma. (C) At 1 year followup functional outcome following curettage and bone graft substitute surgery for right proximal ulna intraosseous schwannoma.
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Fig. 4: (A) Anteposterior and oblique views of the hand with wrist show a osteolytic lesion involving the fifth metacarpal head with dislocation on the dorsal aspect. (B, C, D) Coronal and axial sections of magnetic resonance imaging of the hand show an expansile lytic lesion involving the fifth metacarpal with soft tissue extension on both the ventral and dorsal aspects of the metacarpal bone.

Fig. 5: Radiograph of right hand at 1 year follow-up following curettage of 5th metacarpal bone intraosseous schwannoma.
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<table>
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<tr>
<th>No</th>
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<th>Affected Bone</th>
<th>Radiograph</th>
<th>Computed Tomography (CT-Scan)</th>
<th>Magnetic Resonance Imaging (MRI)</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20 yrs M</td>
<td>Scapula</td>
<td>Shows well defined lytic lesion with thin sclerotic rim involving genoid</td>
<td>Shows expansive lytic lesion with cortical breach at multiple sites involving genoid part of scapula</td>
<td>Shows low to isointense lesion on T1-weighted images, hyperintense lesion on T2-weighted images</td>
<td>Total Scapuectomy</td>
<td>60 months</td>
</tr>
<tr>
<td>2</td>
<td>27 yrs F</td>
<td>Proximal ulna</td>
<td>Well defined lytic lesion with thin rim of sclerosis involving proximal ulna</td>
<td></td>
<td>Shows low to isointense lesion on T1-weighted images, hyperintense lesion on T2-weighted images</td>
<td>Extended curettage + hydroxyapatite crystals</td>
<td>15 months</td>
</tr>
<tr>
<td>3</td>
<td>24 yrs M</td>
<td>Fifth metacarpal</td>
<td>Shows expansive lytic lesion involving metacarpal head with dislocation of 5th metacarpophalangeal joint</td>
<td></td>
<td>Shows low to isointense lesion on T1-weighted images, hyperintense lesion on T2-weighted images</td>
<td>Extended curettage</td>
<td>13 months</td>
</tr>
</tbody>
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Table 1: Study characteristics of patients with upper extremity intraosseous Schwannoma (IOS)

<table>
<thead>
<tr>
<th>No</th>
<th>Sex</th>
<th>Age</th>
<th>Ethnic</th>
<th>Affected Bone</th>
<th>Radiograph</th>
<th>Computed Tomography (CT-Scan)</th>
<th>Magnetic Resonance Imaging (MRI)</th>
<th>Treatment</th>
<th>Follow-up</th>
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<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>20</td>
<td>White</td>
<td>Scapula</td>
<td>Shows well defined lytic lesion with thin sclerotic rim involving genoid</td>
<td>Shows expansive lytic lesion with cortical breach at multiple sites involving genoid part of scapula</td>
<td>Shows low to isointense lesion on T1-weighted images, hyperintense lesion on T2-weighted images</td>
<td>Total Scapuectomy</td>
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<td>13 months</td>
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Table 2: Review of literature on intra-osseous Schwannoma (IOS) in extremity bones
References


