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Schwannoma of the Suprascapular Nerve Presenting With Atypical Neuralgia

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Abstract

The suprascapular nerve, being a mixed peripheral, supplies the supraspinatus and the infraspinatus muscles and posterior aspect of the capsule of the shoulder joint. Impairment of this nerve, therefore presents as weakness and atrophy of these muscles and a poorly defined aching pain along the posterior aspect of the shoulder joint and the adjacent scapula.Amongst the various non-traumatic suprascapular nerve lesions, entrapment neuropathy at the suprascapular notch is far more common than tumorous lesions. When considering the latter, ganglion cysts occurring in relation to the suprascapular nerve are more frequently seen. Whereas lipoma, other soft tissue and peripheral nerve sheath tumours [PNST] are rare. This is a case report of 24 year male presented with complaint of a poorly defined aching pain along the posterior aspect of the right shoulder joint and adjacent scapula, and difficulty in carrying out over head activities since 4 months. On examination weakness and atrophy of supraspinatus and infraspinatus noted. Tenderness elicited in the posterior aspect of the shoulder joint. Rest of the neurological examination including sensation over the shoulder and scapular regions was normal.

Antero-posterior radiograph and Scapular V-Y view of the right shoulder no obvious abnormality noted. MR imaging on T1weighted image showed an isointense mass in the substance of infraspinatus, normal labrum and articular cartilage. MRI on STIR image showed hyper intense signal due to cystic degeneration in the mass at the spinoglenoid notch with no communication of the lesion to the gleno-humeral joint. MR Coronal image showed a more peripheral location of the mass than usual at the spinoglenoid notch. ENMG showed delayed conduction velocities distal to suprascapular notch secondary to suprascapular nerve compression with denervation potentials in the supra- and infra-spinatus muscles. Managed with diagnostic shoulder arthroscopy and open surgical excision of the benign tumour. Histopathological examination revealed Spindle cells with elongated nuclei arranged in alternating hyper cellular palisading regions and hypo cellular loosely meshed regions. Immunohistochemical staining Positive for S 100 classical of Schwannoma. Post-operatively the pain subsided over a period of 5 months and the weakness recovered considerably. Three years following surgery the patient was symptom free and there is no recurrence of swelling.

Key words: *suprascapular nerve, Schwannomas, atypical neuralgia, Atrophy of the supra and infra-spinatus muscles, MRI, Nerve conduction study.*

INTRODUCTION

The suprascapular nerve, being a mixed peripheral, supplies the supraspinatus and theinfraspinatus muscles and posterior aspect of the capsule of the shoulder joint.Impairment of this nerve, therefore presents as weakness and atrophy of these musclesand a poorly defined aching pain along the posterior aspect of the shoulder joint and the adjacent scapula. Amongst the various non-traumatic suprascapular nerve lesions, entrapment neuropathy at the suprascapular notch is far more common than tumorous lesions [1,2,3,4,5]. When considering the latter, ganglion cysts occurring in relation to the suprascapular nerve are more frequently seen [6, 7, 8]. Whereas lipoma, other soft tissue and peripheral nerve sheath tumours [PNST] are rare [9,10,11]. In general, more than 50% of benign or malignant PNST's occur in association with neurofibromatosis-1 or following radiation therapy [12,13,14]. The origin of a de novo solitary schwannoma of the suprascapular nerve at neck of the glenoid and its presentation apart from the weakness and atrophy of the supraspinatus and infraspinatus muscles as atypical neuralgic pains over the posterior aspect of the shoulder joint and adjacent scapula is exceptional.

PRESENTATION OF THE CASE

A 24 year old male presented to out-patient department with complaint of poorly defined dull aching pain along the posterior aspect of the right shoulder joint and adjacent scapula associated with difficulty in carrying out activities involving abduction and external rotation of the shoulder of 4 months duration. He correlates the onset of these symptoms to injury to shoulder while pulling ropes at his farm 4 months back which was managed with rest and analgesics. Patient relatives noticed hollowness on the back of the right shoulder which is gradually increasing during this period.

On examination

Inspection

Wasting of the supraspinatus and infraspinatus muscles with more prominent spine of the scapula on right side compared to left side as shown in figure 1a and 1b. No obvious swelling noted.

Palpation

No local rise of temperature, tenderness present on deep palpation over the posterior aspect of the shoulder joint. There definite wasting of supra- and infra-spinatus muscles and weakness of right shoulder abduction and external rotation. The rest of the neurological examination including sensation over the shoulder and scapular regions was normal. All peripheral pulses were normal.

A careful search for possible stigmata of neurofibromatosis was negative. No obvious abnormality noted on antero-posterior and scapula V-Y view plain radiographs of right shoulder as shown in figure 2. HRUS of right shoulder revealed aniso-anechoic mass with cystic component in the substance of infraspinatus extending to neck of the spinoglenoid notch. Ultrasound guided aspiration of the cystic component of the lesion revealed thick gelatinous material with brownish tinge. MR imaging on

T1weighted image showed a isointense mass in the substance of infraspinatus (arrow head), normal labrum ,articular cartilage(small arrow) as shown in figure 3 . MR imaging on STIR image showed hyperintense signal due to cystic degeneration in the mass at the spinoglenoid notch with no communiation of the lesion to the gleno-humeral joint as shown in figure 4a and 4b. MR Coronal image showed a more peripheral location of the massthan usual at the spinoglenoid notch (arrow) as shown in figure 5a and 5b.ENMG showed delayed conduction velocities distal to suprascapular notch secondary to suprascapular nerve compression with denervation potentials in the supra- and infra-spinatus muscles.

Management

We planned for diagnostic arthroscopy and further management. Diagnosticarthroscopy was negative for labral pathology or communication of the lesion with gleno-humeral joint cavity.Incision marked along the inferior margin of the spine ofthe scapula, following this deep dissection carried out, trapezius retracted upwards along with periosteum and dissection carried out in the substance of infraspinatus muscle.

A mass measuring 4×5 centimetre extending along the spinoglenoid notch into the supraspinatus region was identified as shown in figure 6a and 6b. During further dissection a fine attachment of this mass with suprascapular nerve was identified as shown in figure 7a and 7b and then the nerve was traced to suprascapular notch. On palpation the mass was soft in consistency with areas of cystic degeneration. A complete excision of the mass measuring 4×5×3 centimeter was achieved with microsurgical technique preserving the integrity of the suprascapular nerve as shown in figure 8. Wound closed after anchoring trapezius back through drill holes made in the spine of the scapula as shown in figure 9. Postoperatively the pain subsided completely and the patient showed distinct improvement in muscle strength despite persistent muscle mass reduction during follow-up.

Gross picture

Solitary well defined mass with smooth glistening surface measuring 4 x 5 x 3 cm, Soft in consistency with areas of cystic degeneration containing gelatinous material with brownish tinge as shown in figure 8.

Histopathological examination

Spindle cells with elongated nuclei arranged in alternating hyper cellularpalisading regions and hypo cellular loosely meshed regions as shown in figure 11.

Immunohistochemistry

Staining positive for S100 classical of schwannoma as shown in figure 10.

Follow up

Three years following surgery the patient was symptom free and there is no recurrence of swelling.

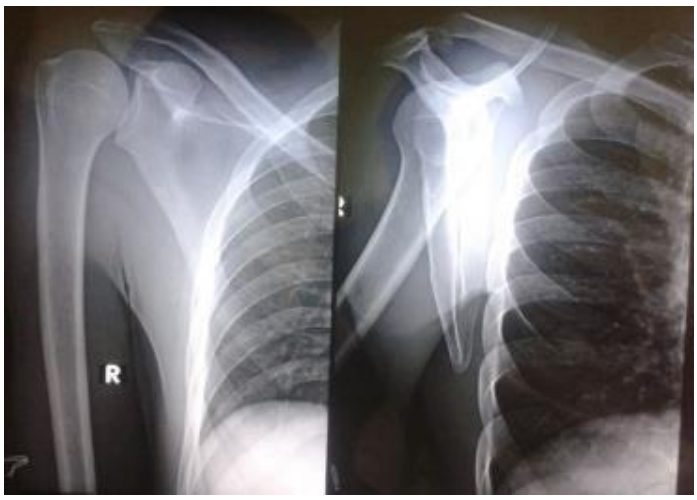
Figure 1a

Back of the shoulder
Atrophy of infraspinatus (long arrow)
Atrophy of supraspinatus (small arrow)

Figure 1b

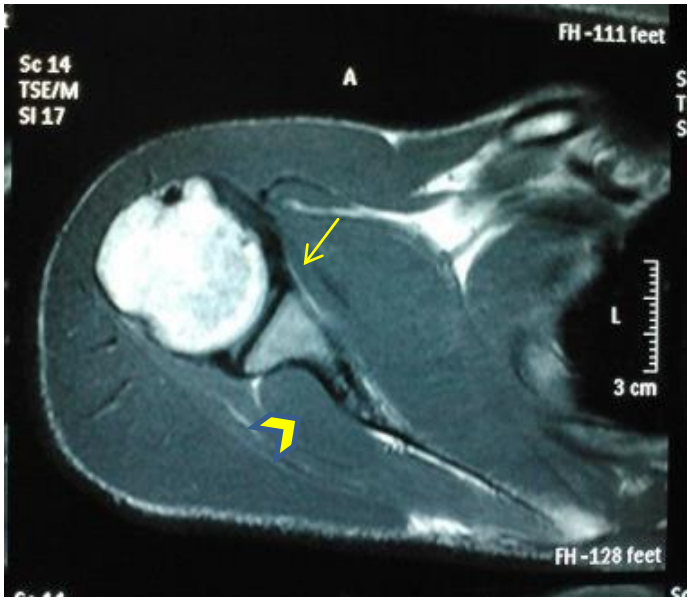
Side view
Atrophy of supraspinatus and infraspinatus muscle (arrow)

Figure 2



Plain Radiograph
Antero-posterior & Scapula V-Y view Normal

Figure 3



MR imaging - T1weighted image
Isointense mass in the substance of infraspinatus (arrow head), Normal labrum, Articular cartilage(small arrow) .

Figure 4a

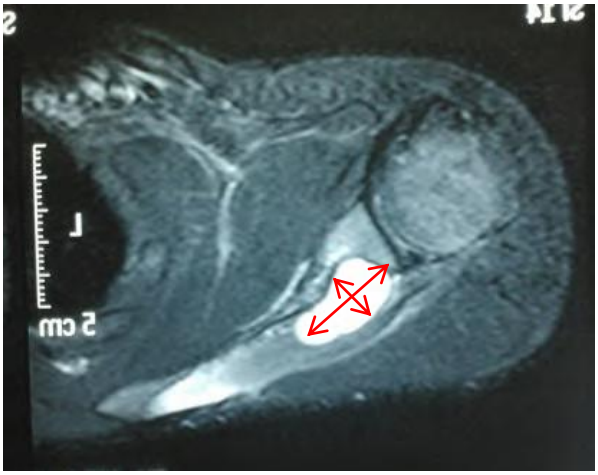
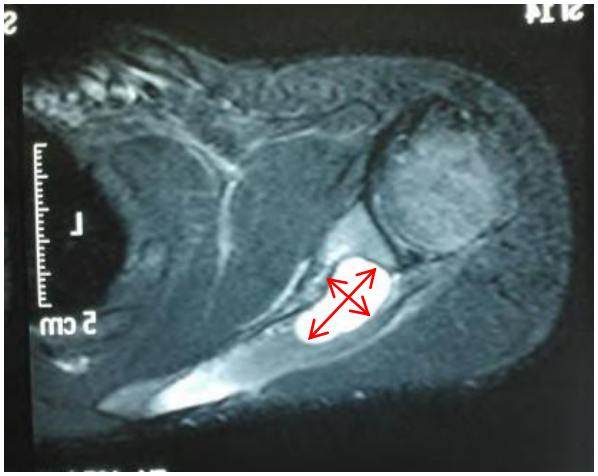


Figure 4b



STIR images showing hyper intense signal due to cystic degeneration in the mass at Spinoglenoid notch without any communication to the Gleno-humeral joint

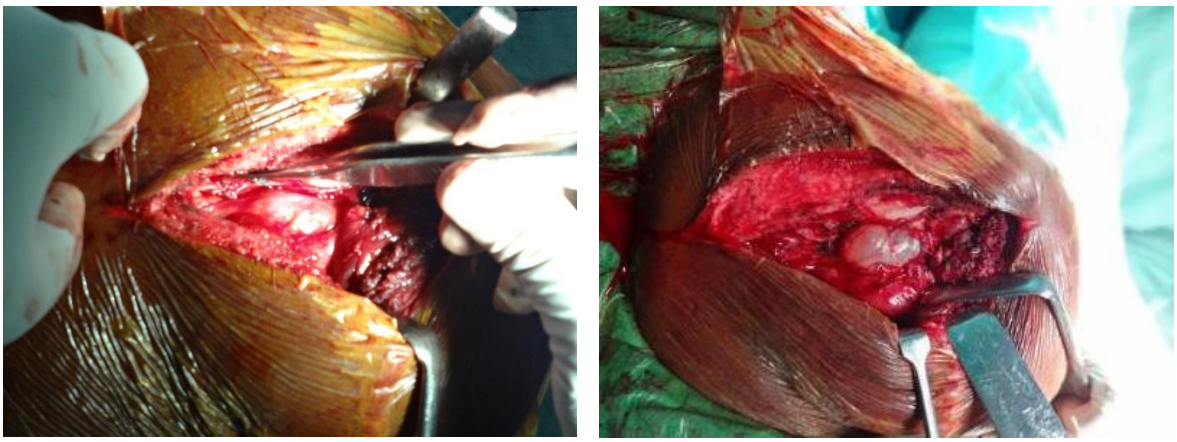
Figure 5a

Figure 5b

Coronal image showing more peripheral location of the mass than usual at the spinoglenoid notch (arrow)

Figure 6a

Figure 6b



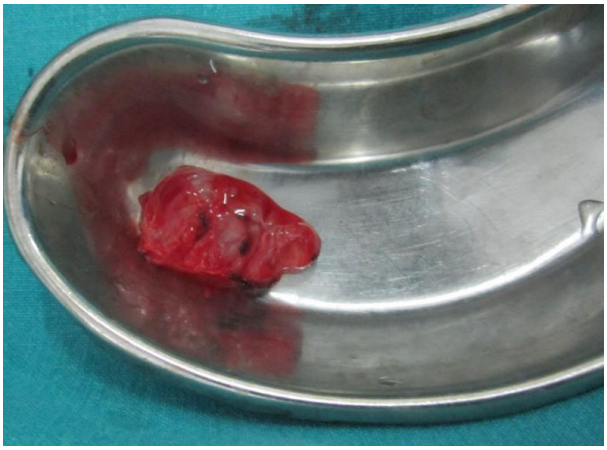
Intra-operative image showing mass in the substance of infraspinatus muscle below spine of scapula after sub-periosteal elevation of trapezius

Figure 7a

Figure 7b

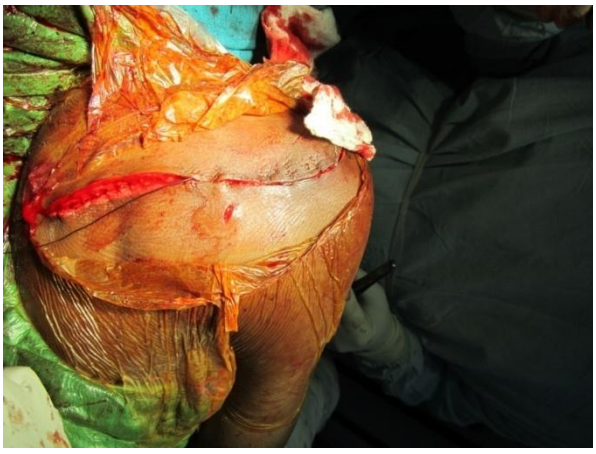
During further dissection a fine attachment of mass to the suprascapular nerve is found as it is traced proximally towards the spinoglenoid notch

Figure 8



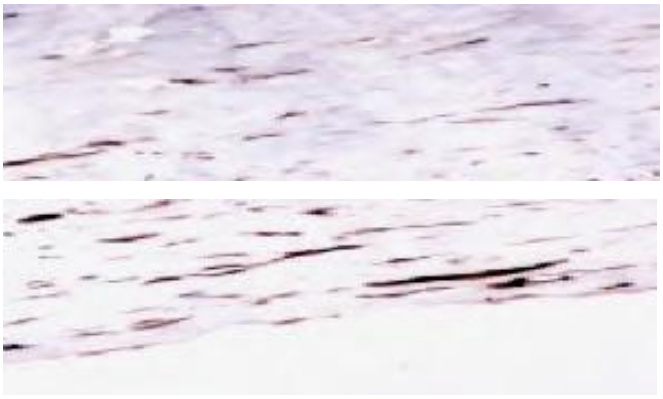
A glistening mass measuring 4×5×3 cm, soft on palpation with areas of cystic degeneration

Figure 9



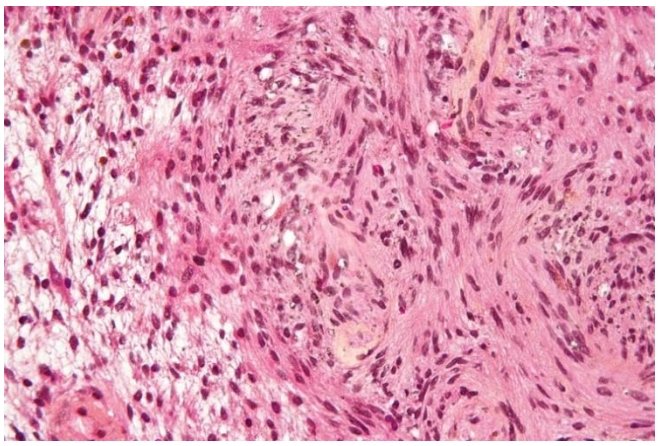
Wound closed after anchoring trapezius back through drill holes made in the spine of the scapula

Figure 10



Immuno-histo-chemical staining
Positive for **S 100** classical of Schwannoma

Figure 11



Histopathological examination
Spindle cells with elongated nuclei arranged in alternating hypercellular palisading regions and hypocellular loosely meshed regions.

DISCUSSION

Among the lesions of the suprascapular nerve, traumatic suprascapular neuropathy in the posterior triangle of the neck and suprascapular entrapment neuropathy at the suprascapular are two well described pathologies[5,12], whereas neoplastic lesions involving the suprascapular nerve are relatively rare [9,11,15]. Ganglion cysts involving this nerve are now more frequently reported because of MR imaging and ultrasonography [6,9,10], whereas solitary benign or malignant peripheral nerve sheath tumour arising `de novo' from suprascapular nerve and unassociated with neurofibromatosis-1 or prior radiation therapy is extremely rare [16,17,18]. In the case under discussion the lesion was a schwannoma of the suprascapular nerve. Such a lesion is more often seen in association with the spinal accessory nerve [19,20], or other nerves related to the cervical and brachial plexuses [21,22].

The suprascapular nerve arises from the upper trunk (C5–C6)of the brachial plexus. It crosses theposterior triangle of the neck deep and parallel to theinferior belly of the omohyoid muscle and runs under the trapezius and through the suprascapular notch,below the suprascapular ligament and into the supraspinous fossa to supply the supraspinatus muscle. The nerve curves around thelateral border of scapular spine to enter infraspinous fossa to supply infraspinatus muscle; it alsosupplies articular branches to the shoulder and acromioclavicular joints. Pain over the posterior aspect of the shoulder joint and adjacent scapula is the most common presenting symptom of suprascapular nerve impairment followed by weakness and atrophy of the supra- and infraspinatus muscles. The absence of detectable cutaneous sensory loss relates to the fact that the suprascapular nerve normally has no cutaneous distribution. Lesions of the anterior horn cells,

cervical rootlets or upper trunk of the brachial plexus involving the fifth cervical segmentalso result in weakness of the rhomboid and the deltoid muscles in addition to weakness of the supra- and infraspinatus muscles as these neural structures are proximal to the origin of the suprascapular nerve[5,12].

Denervation potentials confined to the supra- and infraspinatus muscles and delayed conduction from Erb's point to the supraspinatus muscle are helpful diagnostic tests of suprascapular neuropathy. The latency from Erb's point to the supraspinatus muscle is in the range of 1.7 to 3.7 ms. In rotator cuff injuries, which it is sometimes difficult to differentiate from suprascapularnerve lesions, there are no denervation potentials in the supra- and infraspinatus muscles[1,3,5].

Schwannomas are benign, slowly growing lesions. They are distinguished from other peripheral nerve sheath tumours by the presence of only Schwann cells. The spindle cells with elongated nuclei are arranged either in alternating hyper cellular palisading (Antoni A) regions and/or hypo cellular loosely meshed (Antoni B) regions[22,23,24]. The stromal blood vessels are often dilated with sclerotic walls and can undergo thrombosis. The resultantischaemia leads to haemorrhage, necrosis and cystic change and rarely the cells may produce melanin and become pigmented[6,7,10]. These tumours become well encapsulated by the epineurium and push aside adjacent fascicles; they are soft to palpation and may be cystic and therefore they may mimic a ganglion cyst or lipoma[10,12,13]. CT and MRI clearly demonstrate their location, extent, size and relationship to the surrounding structures; adequate imaging is therefore necessary to plan the microsurgery [7,10,12]. The suprascapular nerve is at risk of being compressed in the suprascapular notch by the suprascapular ligament and also in the posterior triangle of the neck under the firm deep cervical fascia especially when a large swelling is attached to it [4,6,7]. In areas where a schwannomaarises from a peripheral nerve under firm fascia there is a higher likelihood of pain and motor deficits [6,7,10]. Smaller tumours are found on occasions when operating for entrapment syndromes[1,2].

Conclusion

The origin of a de novo solitary schwannoma of the suprascapular nerve at neck of the glenoid and its presentation apart from the weakness and atrophy of the supraspinatus and infraspinatus muscles as atypical neuralgic pains over the posterior aspect of the shoulder joint and adjacent scapula is exceptional. However thorough evaluation is needed to diagnose the various conditions such as suprascapular nerve entrapment, synovial ganglion cysts with or without associated labral tear, traction injury to suprascapular nerve, lipoma at the spinoglenoid notch and other rare causes having similar presentation. MR imaging is highly valuable in differentiating these conditions and assessment of neoplastic pathologies associated with this nerve, by demonstrating their location, extent, size and relation to the surrounding structures. Role of microsurgical techniques during excision of these lesions are highly rewarding.

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