SANAMED
Online First: October, 2024
ISSN-1452-662X

DOI: 10.5937/sanamed0-52736
Case report

SCHWANNOMA OF SCAPULA: CASE REPORT AND LITERATURE REVIEW

Stevanović Stefan,[1] Petrović Jovica,[1] Milićević Aleksandar,[2] Petrović Jelena,[3] Đorđević Nikola,[1] Nikolić Aleksandra[1]

[1] University Clinical Center Niš, Clinic for Thoracic Surgery, Niš, Serbia
[2] University Clinical Center Niš, Center for Pathology, Niš, Serbia
[3] University of Defence, Military Medical Academy, Belgrade, Serbia

Primljen/Received:13. 08. 2024. Prihvaćen/Accepted: 23. 09. 2024. Published online first: October 2024.

Abstract: **Introduction:** Schwannomas are benign peripheral nerve tumors, more often localized in soft tissues than bones. Out of about 200 recorded cases of schwannoma of bone, only three cases of schwannoma of scapula have been described to date.

Case report: We present the case of a 73-year-old female patient with an asymptomatic schwannoma of the scapula. Physical examination revealed a solid, fixed, well-defined walnut-size tumefaction in the right scapula area. CT of the chest confirmed a 2.33 x 0.96 cm diameter tumor at the junction of the upper-third and middle-third of the medial border of the right scapula. After discussion with the patient, it was decided to proceed with surgical removal of the tumor. The surgery involved partial resection of the tumor-affected part of the scapula. Histopathological examination confirmed it was a schwannoma of bone. No clinical or radiological signs of disease recurrence were observed during the one-year follow-up.

Conclusion: Schwannomas of bone are rare, slow-growing tumors. A definitive diagnosis is made based on histopathological and immunohistochemical findings. The main treatment modalities include curettage or "en bloc" resection. Recurrence is rare.

Keywords: neurilemmoma, tumor, surgical treatment, histopathology, radiology.

INTRODUCTION

Schwannomas (also known as neurilemmomas) are benign neurogenic tumors (1). They originate from Schwann cells forming the myelin sheath of the peripheral nervous system and account for approximately 5% of all soft tissue, benign tumors (2). They may develop in any period of life (most often in the fourth decade), with approximately equal incidence in males and females (2). Predilection sites for schwannomas include the head, neck, extremities and mediastinum (3, 4). They are generally slow-growing tumors with low malignant potential (5, 6).

Unlike schwannomas of soft tissue, which are rather common, schwannomas of bone are very rare tumors, with an incidence of less than 0.2% (7, 8). Considering that skeletal nerve fibers are mostly unmyelinated, such a low incidence of these tumors comes as no surprise (9). By 2021, about 200 cases of schwannomas of bone have been reported, with the most common localization in the maxillofacial region and the sacrum (10-13).

Our patient presents with an intraosseous schwannoma of the right scapula. According to the literature available in English, only three cases of schwannoma of scapula have been reported to date, the last being from 2021 (10). Before that, schwannoma of scapula was reported two more times, in 2014 and in 1967 (1, 7).

Given the rarity of this tumor and its even rarer occurrence in the scapula, we have chosen to present this case in detail.

CASE REPORT

A 73-year-old female patient was identified with a tumefaction in the right scapular region. The patient had no subjective complaints, and she discovered the change by accident, approximately one year before the surgery. She denied injuries in the region and previous surgeries. Her family history was negative.

She was referred to a preoperative examination by a general practitioner, with a posteroanterior and lateral chest X-ray. The surgical examination included

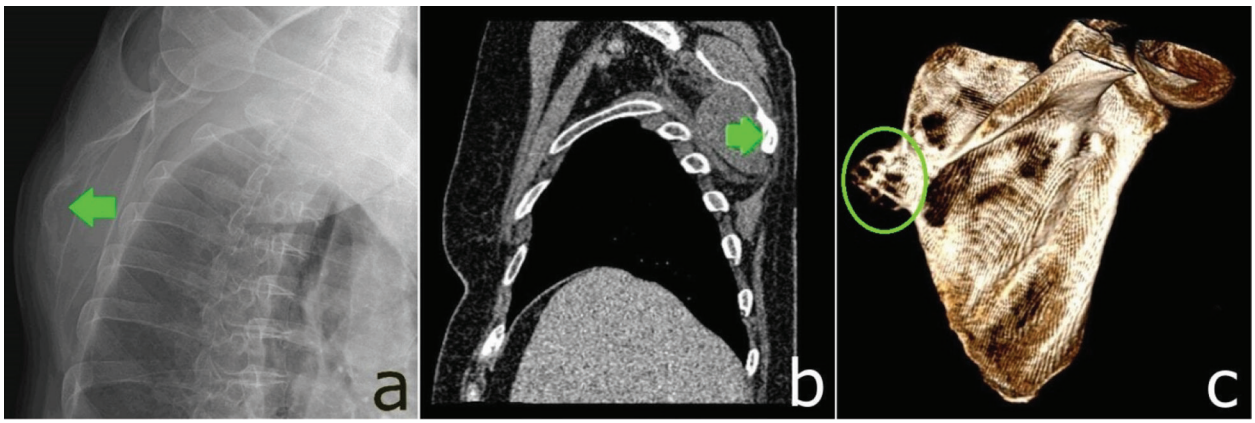

Figure 1. *Radiological features of the resected schwannoma*

Lateral chest X-ray (**1a**) shows a well-defined osteolytic tumor with a thin capsule and trabeculae. On sagittal chest tomogram (**1b**), green arrow shows a tumor on the medial border of the right scapula. Also, we can see a 3D reconstructed right scapula (**1c**) with an osteolytic lesion at the junction of the upper-third and middle-third of its medial border (by RadiAnt DICOM Viewer *v.2023.1*).

inspection and palpation of the right scapular region. Palpation revealed a walnut-size tumefaction in the medial border of the right scapula. The tumor was firm, well-defined, and fixed to the scapula, with no local swelling or skin changes. Mild tenderness to palpation was present. Lateral chest X-ray revealed a well-defined osteolytic lesion in the area of the medial border of the scapula, with a thin sclerotic capsule and trabeculae (Figure 1a). Computed tomography (CT) scan of the chest with contrast agent application was indicated and performed to achieve the most accurate diagnosis. The findings confirmed an osteolytic, well-defined tumor formation, 2.33 x 0.96 cm in diameter, located at the junction of the upper-third and middle-third of the medial border of the right scapula, without signs of pathological vascularization and infiltration of the surrounding soft tissues (Figures 1b and 1c).

After reviewing the entire medical documentation and obtaining the patient's consent, a decision was made to surgically remove the above-described change and refer it for a histopathological examination. After adequate preoperative preparation, the surgery was performed under general anesthesia. We approached the tumor through an incision in the skin and subcutaneous tissue of the right scapular region, with the patient being in the left lateral decubitus position. By preparing the surrounding soft tissues, we reached the tumor and performed a partial resection of the affected part of the scapula (Figure 2).We completely removed the change. Following tumor extirpation, we performed a revision of the area, established hemostasis, and drained the area using a four-channel drain. The postoperative course was normal and the patient was discharged home to self-care on the fifth postoperative day.

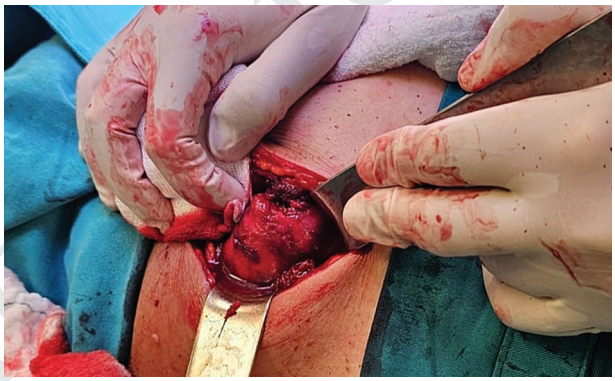

Figure 2. *Perioperative view of the tumor-affected part of the scapula (following preparation of the surrounding soft tissues)*

The specimen sent for histopathological examination contained the resected part of the scapula with tumefaction and surrounding soft tissues. Upon section, it had a colorful appearance and tough-elastic consistency, and it was partially encapsulated, measuring 8.3 x 6.5 x 4.5 cm (Figure 3a). The histopathological finding of the excised change was morphologically and immunohistochemically consistent with the schwannoma of bone. Examination of the material revealed a proliferation of spindle cells (with bent nuclei and intranuclear inclusions) arranged in the *Antoni A* (Figures 3b and 3c) and *Antoni B* (Figures 3d and 3e) patterns. The tumor parenchyma was vascularized by thickened, hyalinized blood vessels and occasionally permeated with degeneration and bleeding zones. There were no atypical cells or other signs of malignancy. *Ki67* proliferative index was < 2% (Figure 3f). Most tumor cells showed positive immunohistochemical staining with *S-100* protein (Figures 4a and 4b), *CD56* (Figures 4c and 4d), *bcl2* and *TLE1*.

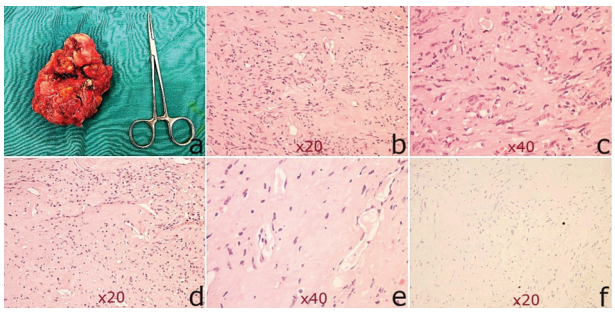

Figure 3. *Patohistological features of the resected schwannoma*

The figure shows the surgical specimen (**3a**), with a colorful appearance, tough-elastic consistency, composed of spindle cells arranged in *Antoni A* (**3b et 3c**) and *Antoni B* (**3d et 3e**) patterns (*H&E staining*). *Ki67* proliferative index was < 2% (**3f**).

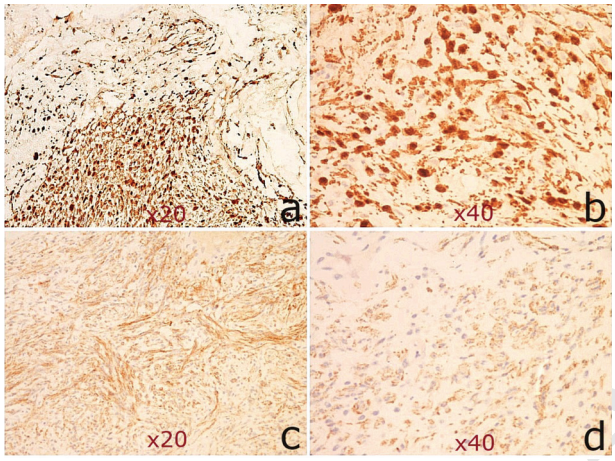

Figure 4. *Immunohistochemical features of the resected schwannoma*

Most tumor cells showed strong immunohistochemical staining positivity with *S-100* (**4a et 4b**) and *CD56* (**4c et 4d**).

The patient did not receive additional therapy. The surgical wound was healing *per primam*. After a one-year follow-up, there were no clinical or radiological signs of disease recurrence.

DISCUSSION

Neurilemmoma, now commonly known as schwannoma, was first described by Verocay in 1908 and elaborated upon by Stout in 1935 (13, 14). The disease etiology has not been fully elucidated, but the formation of this tumor seems to be caused by a mutation of *NF2* gene (15, 16). *NF2* is a tumor-suppressor gene (chromosome 22), which encodes the structure and synthesis of the *schwannomin* protein. Schwannomin is synthesized in the nervous system, predominantly in Schwann cells, and plays a significant role in the transduction of signals responsible for their structure, growth, division and adhesion. The loss of *NF2* gene function leads to disruption of these functions and the formation of tumors (16).

As mentioned in the introduction, schwannomas account for less than 0.2% of all bone tumors (7, 8). Generally, these tumors occur more in sensory than motor nerves. This is probably the reason why the tumor process most often affects the mandible and sacrum (8, 13). Unlike these bones, scapula is a rather rare site for schwannoma and has been reported only three times to date. Fawcett *et al*. were the first to describe schwannoma of the left scapula in a 24-year-old female patient, a little over 50 years ago (7). Tian et al. reported the same tumor in a 42-year-old woman in 2014 (1). The last case of schwannoma of scapula was reported by Reyniers et al. in 2021 (10). In all three cases, there were female patients with schwannoma localized on the left and affecting the glenoid cavity of the scapula, and the only symptom they had was pain. Following *in toto* tumor removal, there was no disease recurrence. In our case, the tumor was localized on the right, did not involve the glenoid cavity, and was asymptomatic. Following tumor resection and one-year follow-up, there were no signs of recurrence. So far, the recurrence of schwannoma of bone has been reported twice, probably due to incomplete resection (8, 17).

Schwannomas can involve bone in three ways: 1) by arising in the medullary cavity of the bone; 2) by arising in the nutrient canal of the bone or 3) by arising in the immediate vicinity of the bone, causing its secondary erosion (18).

The disease can be asymptomatic (25% of cases) or manifested by symptoms such as pain, swelling, and redness at the tumor site (1). Pain is reported in nearly half of the patients and typically worsens with tumor palpation. Fewer patients may experience loss of function of the affected nerve, due to its prolonged compression by the tumor (1, 19).

The diagnostic algorithm includes detailed history taking, physical examination, and additional radiological tests (ultrasound, radiography, CT, MRI). A definitive diagnosis is made solely based on the histopathological findings of the examined material. The classical radiological presentation of schwannoma of bone implies a well-defined osteolytic lesion with a thin sclerotic capsule and trabeculae (20). This radiological description largely corresponds to our case. The main histological feature of schwannoma is the presence of spindle cells arranged in the *Antoni A* and *Antoni B* patterns. Tumor cells typically show *S-100* protein positivity (*ddx.* neurofibroma) (1, 21, 22). Al-

though there are several different histological types of schwannoma (classical, epithelioid, cellular, microcystic, neuroblastoma-like, etc.), this classification has no major clinical significance in itself (23).

Schwannomas are benign tumors. Malignant alteration is rare and thus far only recorded in soft tissue, not in skeletal forms (8). Curettage or "en bloc" resection are the methods of choice for treating schwannoma of bone (8, 24). Major bone defects can be compensated with grafting (25). In our case, the patient was treated with the "en bloc" resection method without additional therapy.

CONCLUSION

Schwannomas of bone are rare tumors. They are usually slow-growing and have mild symptoms, most commonly pain. One-quarter of patients have no complaints at all. A definitive diagnosis is made based on histopathological and immunohistochemical findings. The main treatment modalities include curettage or "en bloc" resection. Recurrence is rare.

Abbreviations

Bcl2 – B-cell leukemia/lymphoma 2 protein
CD56 – Neural cell adhesion molecule
NF2 – Neurofibromatosis type 2 gene
TLE1 – Transducin-like enhancer protein 1

Conflict of Interests: The authors declare no conflicts of interest related to this article.

Funding: No.

Author contribution: All authors have contributed equally

Note: Artificial intelligence was not utilized as a tool in this study.

Sažetak

ŠVANOM LOPATICE: PRIKAZ SLUČAJA I PREGLED LITERATURE

Stevanović Stefan,[1] Petrović Jovica,[1] Milićević Aleksandar,[2]
Petrović Jelena,[3] Đorđević Nikola,[1] Nikolić Aleksandra[1]

[1] Univerzitetski klinički centar Niš, Klinika za grudnu hirurgiju, Niš, Srbija
[2] Univerzitetski klinički centar Niš, Centar za patologiju, Niš, Srbija
[3] Univerzitet odbrane, Vojnomedicinska akademija, Beograd, Srbija

Uvod: Švanomi su benigni tumori perifernih nerava, sa češćom lokalizacijom u mekim tkivima nego u kostima. Od oko 200 zabeleženih slučajeva koštanog švanoma, švanom lopatice je do danas svega tri puta opisan.

Prikaz slučaja: U našem slučaju je prikazana 73-godišnja pacijentkinja sa asimptomatskim švanomom lopatice. Fizikalnim pregledom je evidentiran čvrst, fiksiran, jasno ograničen tumefakt, veličine oraha, u predelu desne lopatice. CT-om grudnog koša je potvrđeno da na spoju gornje i srednje trećine unutrašnje ivice desne lopatice postoji tumorska promena dijametra 2,33 x 0,96 cm. Nakon razgovora sa pacijentkinjom doneta je odluka da se navedena promena hirurški ukloni. Operacija je obuhvatila parcijalnu resekciju tumorom zahvaćenog dela lopatice. Patohistološkim pregledom je utvrđeno da se radi o koštanom švanomu. Tokom jednogodišnjeg praćenja nisu zabeleženi klinički i radiološki znaci recidiva bolesti.

Zaključak: Koštani švanomi su retki, sporo rastući tumori. Definitivna dijagnoza se zasniva na rezultatima patohistoloških i imunohistohemijskih pretraga. Glavni modaliteti lečenja uključuju kiretažu ili "en bloc" resekciju. Recidivi su retki.

Ključne reči: neurilemom, tumor, hirurško lečenje, patohistologija, radiologija.

REFERENCES

1. Tian YW, Zhang LY, Liu ZQ. Giant intraosseous schwannoma of scapula: a rare case report and review of the literature. Diagn Pathol. 2014; 9:31. doi: 10.1186/1746-1596-9-31.

2. Albert P, Patel J, Badawy K, Weissinger W, Brenner M, Bourhill I, et al. Peripheral nerve schwannoma: a review of varying clinical presentations and imaging findings. J Foot Ankle Surg. 2017; 56(3): 632–7. doi: 10.1053/j.jfas.2016.12.003.

3. Guerrissi JO. Solitary benign schwannomas in major nerve systems of the head and neck. J Craniofac Surg. 2009; 20(3): 957–61. doi: 10.1097/SCS.0b013e3181a14cbc.

4. Fierro N, D'ermo G, Di Cola G, Gallinaro LS, Galassi G, Galassi G. Posterior mediastinal schwannoma. Asian Cardiovasc Thorac Ann. 2003; 11(1): 72–3. doi: 10.1177/021849230301100118.

5. McMenamin ME, Fletcher CD. Expanding the spectrum of malignant change in schwannomas: epithelioid ma-

lignant change, epithelioid malignant peripheral nerve sheath tumor, and epithelioid angiosarcoma: a study of 17 cases. Am J Surg Pathol. 2001; 25(1): 13-25. doi: 10.1097/00000478-200101000-00002.

6. Woodruff JM, Selig AM, Crowley K, Allen PW. Schwannoma (neurilemoma) with malignant transformation. A rare, distinctive peripheral nerve tumor. Am J Surg Pathol. 1994; 18(9): 882-95. doi: 10.1097/00000478-199409000-00003.

7. Fawcett KJ, Dahlin DC. Neurilemmoma of bone. Am J Clin Pathol. 1967; 47(6): 759–66. doi: 10.1093/ajcp/47.6.759.

8. Ida CM, Scheithauer BW, Yapicier O, Carney JA, Wenger DE, Inwards CY, et al. Primary schwannoma of the bone: a clinicopathologic and radiologic study of 17 cases. Am J Surg Pathol. 2011; 35(7): 989-97. doi: 10.1097/PAS.0b013e31821fcc0c.

9. Gordon EJ. Solitary intraosseous neurilemmoma of the tibia: review of intraosseous neurilemmoma and neurofibroma. Clin Orthop Relat Res. 1976; 117: 271-82.

10. Reyniers P, Wafa H, Sinnaeve F, Debeer P, Sciot R. Intraosseous schwannoma of the glenoid: case report and literature review. SICOT-J. 2021; 7:2. doi: 10.1051/sicotj/2020048.

11. Suzuki K, Yasuda T, Watanabe K, Kanamori M, Kimura T. Association between intraosseous schwannoma occurrence and the position of the intraosseous nutrient vessel: a case report. Oncol Lett. 2016; 11(5): 3185–8. doi: 10.3892/ol.2016.4363.

12. Zhang L, Xia BQ, Sun H, Wang LZ, Zhao ZL, Li B, et al. Intraosseous schwannomas of the jaws: 2 case reports and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol. 2012; 114(6): e13–7. doi: 10.1016/j.oooo.2011.09.026.

13. Buric N, Jovanovic G, Pesic Z, Krasic D, Radovanovic Z, Mihailovic D, et al. Mandible schwannoma (neurilemmoma) presenting as periapical lesion. Dentomaxillofac Radiol. 2009; 38(3): 178-81. doi: 10.1259/dmfr/59344498.

14. Stout AP. The peripheral manifestations of the specific nerve sheath tumor (neurilemoma). Am J Cancer. 1935; 24: 751–96.

15. Kline DG, Donner TR, Happel L, Smith B, Richter HP. Intraforaminal repair of plexus spinal nerves by a posterior approach: an experimental study. J Neurosurg. 1992; 76(3): 459-70. doi: 10.3171/jns.1992.76.3.0459.

16. Helbing DL, Schulz A, Morrison H. Pathomechanisms in schwannoma development and progression. Oncogene. 2020; 39(32): 5421-9. doi: 10.1038/s41388-020-1374-5.

17. Wirth WA, Bray CB Jr. Intra-osseous neurilemoma. Case report and review of thirty-one cases from the literature. J Bone Joint Surg Am. 1977; 59(2): 252-5. PMID: **845213.**

18. de la Monte SM, Dorfman HD, Chandra R, Malawer M. Intraosseous schwannoma: histologic features, ultrastructure, and review of the literature. Hum Pathol. 1984; 15(6): 551-8. doi: 10.1016/s0046-8177(84)80009-x.

19. Huajun J, Wei Q, Yuxuan W, Jingjing Y. Intraosseous schwannoma of the proximal humerus with pathologic fracture. Eur J Med Res. 2021; 26(1): 72. doi: 10.1186/s40001-021-00541-7.

20. Isaac J, Shyamkumar NK, Karnik SV. Intraosseus schwannoma. J Postgrad Med. 2004; 50(2): 127-8.

21. Kindl RP, Patel K, Trivedi RA. Supraclavicular brachial plexus approach for excision of C8 nerve root schwannoma: 3-dimensional operative video. Oper Neurosurg (Hagerstown). 2019; 16(5): 634-5. doi: 10.1093/ons/opy209.

22. Terzic Z, Radonjic D, Paunovic M, Ljaljevic A, Bojic M. Large solitary encapsulated neurofibroma of upper arm-a case report. Sanamed. 2022; 17(1): 33-6. doi: 10.5937/sanamed17-36810.

23. Meyer A, Billings SD. What's new in nerve sheath tumors. Virchows Arch. 2020; 476(1): 65-80. doi: 10.1007/s00428-019-02671-0.

24. Palocaren T, Walter NM, Madhuri V, Gibikote S. Schwannoma of the fibula. J Bone Joint Surg Br. 2008; 90(6): 803-5. doi: 10.1302/0301-620X.90B6.19901.

25. Mutema G, Sorger J. Intraosseous schwannoma of the humerus. Skeletal Radiol. 2002; 31(7): 419–21. doi: 10.1007/s00256-002-0493-0.

***How to cite this article**:* Stevanović S, Petrović J, Milićević A, Petrović J, Đorđević N, Nikolić A. Schwannoma of scapula: case report and literature review**.** Sanamed. Online First, October 2024. doi: 10.5937/sanamed0-52736.

Correspondence to/Autor za korespondenciju
Stefan Stevanović, M.D.
Address: 48 Mlinska St., 16240 Medveđa, Serbia
Phone: +381 61 66 58 218
E-mail: dr.stefan.stevanovic@gmail.com
ORCID: 0009-0007-8026-7168