CASE REPORT

RJME Romanian Journal of Morphology & Embryology http://www.rjme.ro/

Cervical intranodal schwannoma – a rare diagnosis

DANIEL MIREA[1)], MARIUS CLAUDIU MANEA[1)], NONA IONELA BEJINARIU[1)], BOGDAN MOCANU[2)], ALINA MIHAELA CIOCÂLTEU[3,4)], MIRELA ŢIGLIŞ[5,6)], ILEANA PERIDE[7,8)], TIBERIU PAUL NEAGU[9,10)], LILIANA ELENA MIREA[5,6)], IOAN LASCĂR[9,10)]

[1)]*Department of Otolaryngology, Ponderas Academic Hospital, Bucharest, Romania*

[2)]*Department of Neurosurgery, Brain Institute, Monza Hospital, Bucharest, Romania*

[3)]*Clinical Department No. 12 – Ophthalmology, Otolaryngology, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[4)]*Ophthalmology Emergency Clinical Hospital, Bucharest, Romania*

[5)]*Clinical Department No. 14 – Orthopedics, Anesthesia and Intensive Care, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[6)]*Department of Anesthesia and Intensive Care, Emergency Clinical Hospital of Bucharest, Romania*

[7)]*Clinical Department No. 3 – Nephrology, Urology, Immunology, Transplant Immunology, Dermatology, Allergology, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[8)]*Department of Nephrology and Dialysis, St. John Emergency Clinical Hospital, Bucharest, Romania*

[9)]*Clinical Department No. 11 – Plastic and Reconstructive Surgery, Pediatric Surgery, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[10)]*Department of Plastic Surgery and Reconstructive Microsurgery, Emergency Clinical Hospital of Bucharest, Romania*

Abstract

Intranodal schwannoma is a rare benign tumor, which originates from the peripheral nerve sheath (Schwann cells), fewer cases being reported with lymphatic involvement. We present the case of a middle-aged female patient, with one-year growing mass in the lateral-cervical area, in intimate relation with the vascular package of the neck. Preoperative cervical computed tomography examination showed the tumor features. There was no intraoperative complication, with the piece being completely removed. The morphological examination revealed the structure of a lymph node, and after Hematoxylin–Eosin staining, there were eosinophilic cytoplasm, euchromatic nuclei, with round, elongated or slightly wavy form and reduced pleomorphism, rare degenerative nuclear atypia, and no mitotic activity nor necrosis. The expression of S100 protein on immunohistochemistry, along with negative results for smooth muscle actin and desmin sustained the diagnosis of intranodal schwannoma of the neck. With a low index of cellular proliferation (Ki67), this case is in line with the reported features of schwannoma having extremely rare malignant transformation.

Keywords: intranodal schwannoma, cervical region, S100 protein expression, low Ki67 index.

Introduction

Schwannoma (also known as neurilemmoma) is a rare encapsulated benign tumor that originates from the Schwann cells, within the peripheral nerve sheath. It can have any localization, 25–45% of reported cases being in the head or neck area. Other regions (where peripheral nerves are located) can be involved, like mediastinum, retroperitoneum, flexor aspects of extremities or cranial and spinal nerves [1–4]. About intranodal schwannoma (lymphatic involvement), there are only few presented cases, being considered as uncommon tumor [1, 4, 5]. Other benign tumors that originate from the peripheral nerve sheaths are neurofibroma and perineurioma [1].

Palisaded intranodal myofibroblastoma, metastatic spindle cell tumors (melanoma, Kaposi's sarcoma, dendritic cell sarcoma and spindle cell carcinoma) or rare metastasizing leiomyoma represent the main differential diagnosis [6–8]. Intranodal schwannomas are usually benign tumors, without metastasis, but in the face of rapid growing masses or pain development, malignancy should be considered [9].

Aim

Therefore, we report a case of a cervical schwannoma originating from the cervical area, in a middle-aged female patient, who presented painless but growing cervical mass.

Case presentation

A 50-year-old female patient, with no significant medical history, presented for a one-year painless, slowly growing right lateral-cervical swelling. On physical examination, the mass was solid, up in the 2/3 superior of the cervical region, with profound localization under the sternocleidomastoid muscle. The long axis of the tumor was longitudinally, with 5×3 cm diameter and no adherence to the surrounding structures. No other pathological modification on clinical examination nor during fiberoptic nasopharyngoscopy were identified. Cervical computed tomography (CT) examination revealed a well-circumscribed, homogenous tissue mass, with modest contrast enhancement, 2.5×4.5 cm in size, localized side of right lateral-cervical vascular

package, which imprints and moves the internal carotid artery to the ventral part of the neck (Figure 1, A–C). Laboratory tests showed slightly increase of erythrocyte sedimentation rate (ESR), hemoglobin and hematocrit values, without any other modifications. Methicillin-resistant *Staphylococcus aureus* (MRSA) screening tests were performed, with negative results. Intraoperative, the tumor was completely removed, without any complications (Figure 2), with a favorable short and long-term post-operative evolution. The formalin-fixed paraffin-embedded resected mass was conventionally stained with Hematoxylin–Eosin (HE). The morphological examination revealed the structure of a lymph node, with rare peripheral lymphoid tissue (Figure 3). The mass proliferation does not exceed the lymph node capsule, with normal architecture being replaced by a nodular mesenchymal proliferation, with elongated cells, with indistinct limits (Figure 4), eosinophilic cytoplasm, euchromatic nuclei, with round, elongated or slightly wavy form and reduced pleomorphism, rare degenerative nuclear atypia, and no mitotic activity nor necrosis. The cells had a fascicular proliferation, in a collagenous background. In addition, immunohistochemistry (IHC) showed expression of S100 protein – useful for establishing a positive diagnosis (Figure 5), with negative results for smooth muscle actin (SMA) and desmin (Figure 6, A and B). The cellular proliferation index (Ki67) was low (5%) (Figure 7). Therefore, the diagnosis of intranodal schwannoma was established.

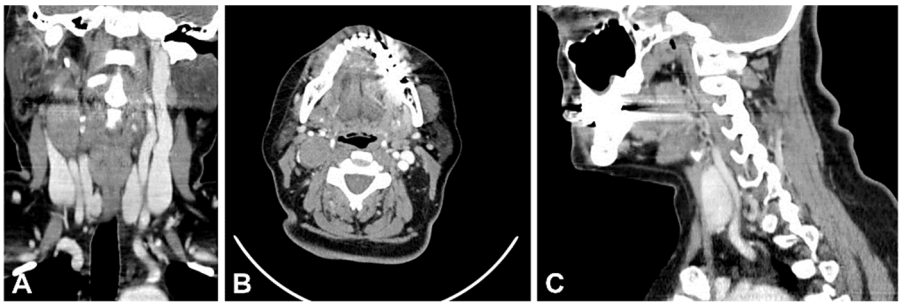

Figure 1 – ***Computed tomography examination, with enhanced contrast: (A) Coronal image presenting a well-circumscribed, homogenous tissue mass, with modest contrast enhancement, of 2.5×4.5 cm, in the right lateral-cervical area, displacing ventrally the carotid artery; (B) Axial image of the same tumor mass; (C) Sagittal image of the same tumor mass.***

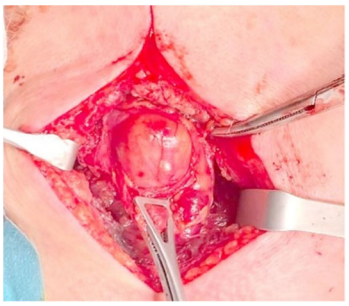

Figure 2 – ***Intraoperative aspect of the intranodal schwannoma showing a solid, well delimited mass.***

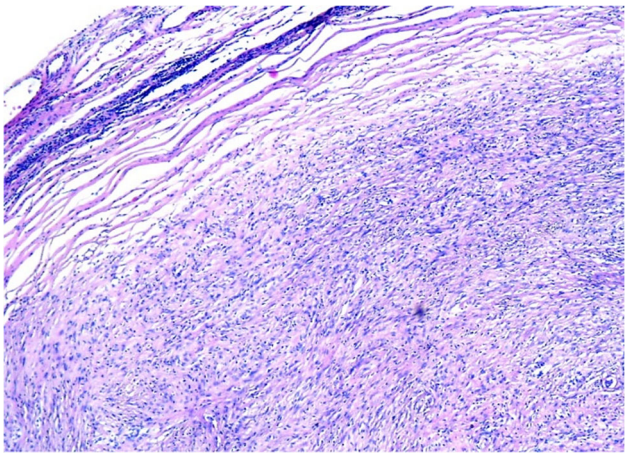

Figure 3 – ***Morphological examination: residual lymph node tissue at the periphery of the tumor and part of the lymph node capsule [Hematoxylin–Eosin (HE) staining, ×40].***

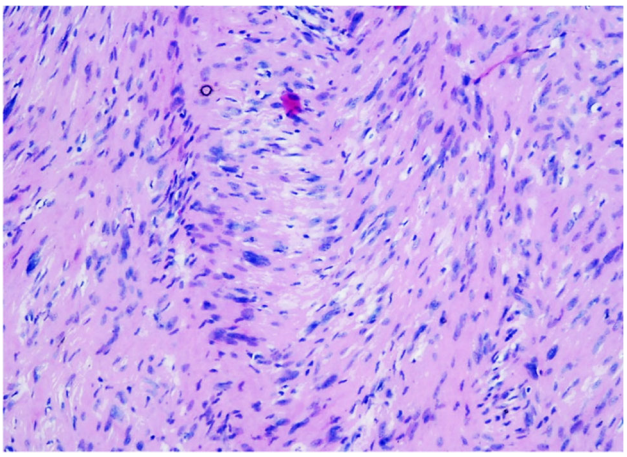

Figure 4 – ***Morphological examination: nodular mesenchymal proliferation, with elongated cells, arranged in palisade (HE staining, ×200).***

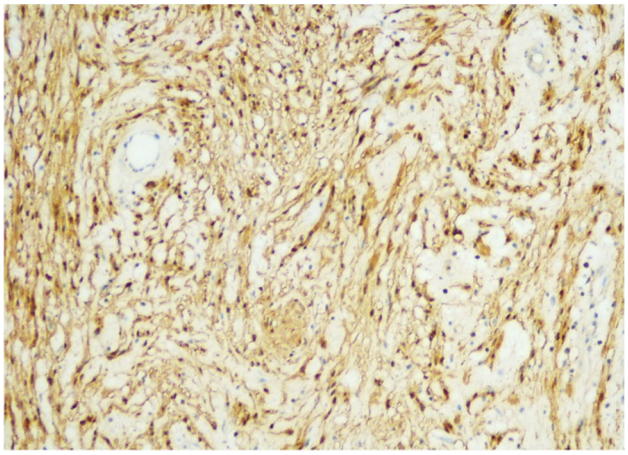

Figure 5 – ***Immunohistochemistry (IHC) analysis (×200): tumor cells with increased expression of S100 protein.***

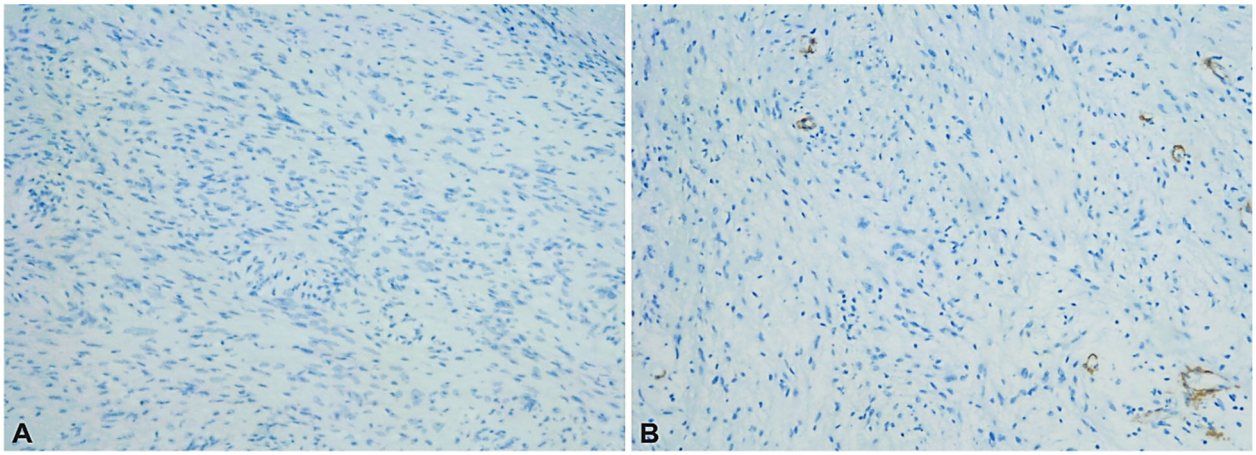

Figure 6 – ***IHC analysis (×200): tumor cells with negative results for smooth muscle actin (SMA) (A) and desmin (B).***

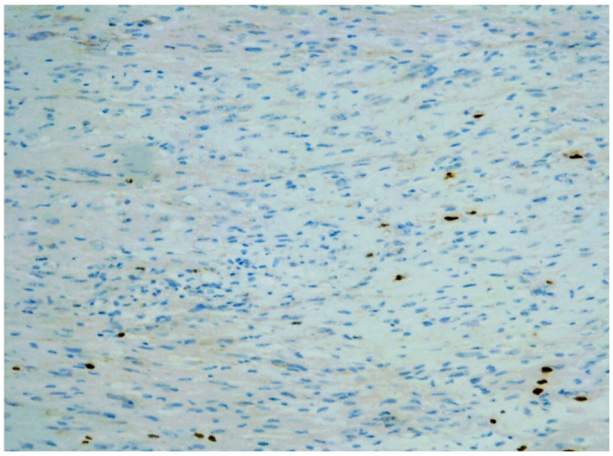

Figure 7 – ***IHC analysis (×200): the cellular proliferation index (Ki67) was around 5%.***

Discussions

Regarding epidemiological aspects, schwannoma has equal incidence in men and women, most reports involving patients between 20–50 years old [10]. The same trend was also observed for the intranodal schwannoma, affecting patients between two to 79 years old, as our case, with no gender preponderance [1, 3–5, 8, 11].

Grant *et al*. reported the first case of an intranodal schwannoma emerging from a parotid lymph node, in a 67-year-old female patient, emphasizing the need to define this as a specific histopathological (HP) entity [12]. A few years later, Hayes & O'Sullivan reported a form of hybrid schwannoma and perineurioma tumor [13].

Normally, after clinical suspicion has been raised, these tumors are diagnosed by CT examination of the incriminated area, followed by surgical excision and HP examination. Depending on mass localization, other diagnosis tools can be used. Kang *et al*. presented a mediastinal lymph node schwannoma, diagnosed using endobronchial ultrasound [14].

Most often, this tumor has <5 cm, with rare reported cases of intranodal schwannoma dimensions of up to 10 cm, having a regular form, with no adherence to other tissues. No necrosis, calcification or cystic modification are normally seen within this type of tumor [1, 3–5, 15, 16].

Another important aspect about schwannoma is that it can be part of schwannomatosis (that can associate different visceral cysts, including in the kidney, according to Merker *et al*. study [17]) or neurofibromatosis type 2 (NF2) disease. Therefore, some patients may be predisposed in presenting with multiple schwannomas over the years, sparing the vestibular nerve (particular to NF2), a disease which can be sporadic or inherited [18, 19]. Due to single localization in our patient and the lack of familial history, no genetic tests were necessary. Nevertheless, a longer follow-period is required in such cases.

On morphological examination, intranodal schwannoma consists of spindle cells, with rare or even absent mitosis [5, 10]. There are two growth patterns reported, Antoni type A, with elongated spindle cells and compact aspect, and Antoni type B, with a careless organization, frequently presenting cystic areas [16]. The exposed case shows classical morphological aspect, without intratumoral necrosis, and an Antoni type A growth pattern. Worth to be mentioned is the absence of stellate zones of collagen deposition (also known as "amianthoid fibers"), which are highly specific for palisaded myofibroblastoma [6, 16].

Regarding IHC features, these tumors present S100 protein, which are low-molecular weight proteins of cells originating from the neural crest [10, 15]. Therefore, in our case, the strong positive expression of S100 protein on IHC analysis, along with the absence of SMA or desmin were useful in making the main differential diagnosis with intranodal palisaded myofibroblastoma and leiomyoma [5, 7].

Conclusions

From our knowledge, this is the third reported case of an intranodal schwannoma located in the cervical area. In face of this tumor, complete resection is associated with favorable evolution. Establishing a correct diagnosis is essential due to the high-risk similar tumors with an aggressive clinical course and fatal outcomes.

Conflict of interests

The authors declare no conflict of interests.

Ethics Statement

All the procedures of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained.

Funding

This research received no external funding.

References

[1] Griffiths AP, Ironside JW, Gray C. True neurilemmoma arising in a lymph node in infancy. Histopathology, 1991, 18(2):180–183. https://doi.org/10.1111/j.1365-2559.1991.tb01465.x PMID: 2010185

[2] Nath P, Sharma H, Chatterjee T. A rare case of hilar lymph node schwannoma. Indian J Surg, 2013, 75(Suppl 1):233–235. https://doi.org/10.1007/s12262-012-0652-z PMID: 24426575 PMCID: PMC3693224

[3] Silvestre CF, Tavares JA, López-Presa D, Dos Santos VR, Rocha J, João Bugalho M. Cervical lymph node schwannoma – an unexpected diagnosis. Clin Pathol, 2019, 12:2632010X19829239. https://doi.org/10.1177/2632010X19829239 PMID: 30915425 PMCID: PMC6429639

[4] Medina-Gallardo A, Curbelo-Peña Y, Molinero-Polo J, Saladich-Cubero M, De Castro-Gutierrez X, Vallverdú-Cartie H. Mesenteric intranodal schwannoma: uncommon case of neurogenic benign tumor. J Surg Case Rep, 2017, 2017(2):rjx008. https://doi.org/10.1093/jscr/rjx008 PMID: 28458819 PMCID: PMC5400457

[5] Black JO, Zhai QJ, Varona OB, Ordonez NG, Luna MA. Primary schwannoma in a cervical lymph node. Head Neck, 2010, 32(7):964–969. https://doi.org/10.1002/hed.21160 PMID: 19536856

[6] Weiss SW, Gnepp DR, Bratthauer GL. Palisaded myofibroblastoma. A benign mesenchymal tumor of lymph node. Am J Surg Pathol, 1989, 13(5):341–346. PMID: 2712186

[7] Suster S, Rosai J. Intranodal hemorrhagic spindle-cell tumor with "amianthoid" fibers. Report of six cases of a distinctive mesenchymal neoplasm of the inguinal region that simulates Kaposi's sarcoma. Am J Surg Pathol, 1989, 13(5):347–357. PMID: 2712187

[8] Reinus C, Shutin O, Reissman P. Intranodal schwannoma presenting as an adrenal mass. Histopathology, 2004, 45(1):95–96. https://doi.org/10.1111/j.1365-2559.2004.01823.x PMID: 15228455

[9] Woodruff JM, Kourea HP, Louis DN, Scheithauer BW. Schwannoma. In: Kleihues P, Cavenee WK (eds). Pathology and genetics of tumors of the nervous system. 3rd edition, vol. 1, World Health Organization (WHO) Classification of Tumours, International Agency for Research on Cancer (IARC) Press, Washington, DC, USA, 2000, 164–166. https://publications.iarc.fr/Book-And-Report-Series/Who-Classification-Of-Tumours/Pathology-And-Genetics-Of-Tumours-Of-The-Nervous-System-2000

[10] Barnes L, Tse LLY, Hunt JL. Tumours of the nasal cavity and paranasal sinuses. In: Barnes L, Eveson JW, Reichart P, Sidransky D (eds). Pathology and genetics of head and neck tumours. 3rd edition, vol. 9, World Health Organization (WHO) Classification of Tumours, International Agency for Research on Cancer (IARC) Press, Lyon, France, 2005. https://publications.iarc.fr/Book-And-Report-Series/Who-Classification-Of-Tumours/Pathology-And-Genetics-Of-Head-And-Neck-Tumours-2005

[11] Rivas F, Ureña A, Macia I, Rosado G, Toñanez J, Penin R, Ramos R. Intranodal thoracic schwannoma, an unusual type of neurogenic tumor. Gen Thorac Cardiovasc Surg, 2011, 59(12):819–821. https://doi.org/10.1007/s11748-010-0773-x PMID: 22173682

[12] Grant DG, Breitenfeldt N, Shepherd NA, Thomas DM. Intranodal neurilemmoma presenting as parotid mass. J Laryngol Otol, 2009, 123(8):912–914. https://doi.org/10.1017/S0022215108003496 PMID: 18940018

[13] Hayes BD, O'Sullivan MJ. Intranodal hybrid benign nerve sheath tumor. Pediatr Dev Pathol, 2011, 14(4):313–317. https://doi.org/10.2350/10-06-0843-CR.1 PMID: 21208040

[14] Kang LH, Shin DH, Yoon SH. Schwannoma arising in mediastinal lymph node diagnosed by endobronchial ultrasound. Respirol Case Rep, 2019, 7(8):e00481. https://doi.org/10.1002/rcr2.481 PMID: 31452898 PMCID: PMC6702317

[15] Nam KB, Namkung S, Kim HC, Kim HS, Ryu BY, Choi YH. Intranodal schwannoma mimicking a gastrointestinal stromal tumor of the stomach: a case report. J Korean Soc Radiol, 2011, 65(4):395–398. https://doi.org/10.3348/jksr.2011.65.4.395 https://jksronline.org/search.php?where=aview&id=10.3348/jksr.2011.65.4.395&code=2016JKSR&vmode=FULL

[16] Piana S, Gelli MC, Cavazza A, Serra L, Gardini G. Ancient schwannoma arising in a lymph node: report of a case and review of the literature. Pathol Res Pract, 2002, 198(1):51–54. https://doi.org/10.1078/0344-0338-00184 PMID: 11866211

[17] Merker VL, Esparza S, Smith MJ, Stemmer-Rachamimov A, Plotkin SR. Clinical features of schwannomatosis: a retrospective analysis of 87 patients. Oncologist, 2012, 17(10):1317–1322. https://doi.org/10.1634/theoncologist.2012-0162 PMID: 22927469 PMCID: PMC3481897

[18] Ferner RE. The neurofibromatoses. Pract Neurol, 2010, 10(2):82–93. https://doi.org/10.1136/jnnp.2010.206532 PMID: 20308235

[19] Mansukhani SA, Butala RPR, Shetty SH, Khedekar RG. Familial schwannomatosis: a diagnostic challenge. J Clin Diagn Res, 2017, 11(2):RD01–RD03. https://doi.org/10.7860/JCDR/2017/20929.9307 PMID: 28384950 PMCID: PMC5376892

Corresponding authors

Tiberiu Paul Neagu, Assistant Professor, MD, PhD, Department of Plastic Surgery and Reconstructive Microsurgery, Emergency Clinical Hospital of Bucharest, 8 Floreasca Avenue, Sector 1, 014461 Bucharest, Romania; Phone +4021–599 23 00, e-mail: dr.neagupaul@gmail.com

Liliana Elena Mirea, Associate Professor, MD, PhD, Department of Anesthesia and Intensive Care, Emergency Clinical Hospital of Bucharest, 8 Floreasca Avenue, Sector 1, 014461 Bucharest, Romania; Phone +4021-599 23 00, e-mail: llmirea@yahoo.com

Received: May 15, 2022

Accepted: August 10, 2022