International Journal of Otolaryngology and Head & Neck Surgery, 2025, 14(3), 230-237
https://www.scirp.org/journal/ijohns
ISSN Online: 2168-5460
ISSN Print: 2168-5452

Neurofibrosarcoma of the Cervical Sympathetic Trunk: Report of a Clinical Case

N'Faly Konate[1*], Oumar Konate[1], Gaoussoou Simpara[1], Kassim Diarra[1], Coulibaly Kalifa[1], Soumaoro Siaka[1], Boubacary Guindo[1], Koné Fatogoma Issa[1], Moussa Bourama Keïta[1], Sidibé Mamadou[1], Gagny Dansoko[1], Singaré Kadiatou[1], Mahamane Django Djibo[2], Mohamed Mohamed Amadou Keita[1]

[1]ENT and Head and Neck Surgery Department, University Hospital Center "Gabriel Toure", Bamako, Mali
[2]Department of Anesthesiology, University Hospital Center "Gabriel Toure", Bamako, Mali
Email: *konatn2@gmail.com

How to cite this paper: Konate, N., Konate, O., Simpara, G., Diarra, K., Kalifa, C., Siaka, S., Guindo, B., Issa, K.F., Keïta, M.B., Mamadou, S., Dansoko, G., Kadiatou, S., Djibo, M.D. and Amadou Keita, M.M. (2025) Neurofibrosarcoma of the Cervical Sympathetic Trunk: Report of a Clinical Case. *International Journal of Otolaryngology and Head & Neck Surgery*, **14**, 230-237.
https://doi.org/10.4236/ijohns.2025.143025

Received: April 13, 2025
Accepted: May 24, 2025
Published: May 27, 2025

Abstract

The aim of this work is to review the management of compressive cervical neurofibrosarcoma. We report the case of a 45-year-old female housewife patient (DT) with no medical or surgical history admitted to the department for left laterocervical swelling. The onset of the disease dates to 6 months after admission, marked by a progressive left laterocervical swelling, with permanent evolution associated with left neck pain, torticollis, and dysphagia to solids. The cervical examination noted a left laterocervical swelling of approximately 8 cm/6 cm, hard, fixed, painful to palpation, with regular contours, non-blowing, non-beating, skin with an inflamed appearance, a bulge of the left lateral wall of the oropharynx pushing the left tonsil inward. A cervical CT scan revealed a weakly enhanced laterocervical tissue hypodensity pushing outward the jugulocarotid axis communicating with the conjugation foramina of C3, C4 and C5 with mass effect on the left vascular-nervous axis of the neck and the pharynx. The standard preoperative biological assessment noted a hemoglobin level of 11.04 g/dl with a hematocrit of 41.06%, and the chest X-ray, frontal and profile, was unremarkable. A transfusion of two bags of isogroup rhesus blood pre- and post-operatively was performed. Our approach used a wide cervicotomy by Paul Andre's "J" incision removing the skin, the subcutaneous cellular tissue and the platysma. The reclination of the anterior flap gave us access to the omohyoid and sternocleidomastoid muscles which we sectioned. The jugulocarotid and vagal neurovascular bundle were dissected from the capsule of the mass, and the spinal nerve was preserved. The tumor was freed from the cervical vertebrae and pharynx. The resection was complete with some ipsilateral cellulolymphatic flows. We established a 2 mm margin from the edges of the lesion. This required us to perform a resection of the transverse processes and the superior

articular processes of C3, C4 and C5. We performed a layer-by-layer closure on a drain. Histology revealed bundles of spindle cells with large and hyperchromatic nuclei; the nucleoli are visible and abnormal mitoses which are in favor of a neurofibrosarcoma. Immunohistochemistry allowed us to confirm the diagnosis of neurofibrosarcoma. The wound healed around the 15th day. The patient was referred for radiotherapy after a multidisciplinary consultation meeting. The clinical evolution after 6 months of post-radiotherapy monitoring was favorable. No functional discomfort was noted. No functional discomfort was noted. **Conclusion**: Neurofibrosarcoma is a malignant tumor that can develop throughout the peripheral nervous system. Its symptoms vary depending on the region. Surgery is the main therapeutic arsenal, often associated with adjuvant radiotherapy.

Keywords

Neurofibrosarcoma, Neurofibromatosis I, Cervical Mass, Peripheral Nerves

1. Introduction

Neurofibrosarcoma, also called malignant schwannoma, is a malignant tumor developed at the expense of the peripheral nerve sheath [1]-[3]. They represent approximately 5% to 10% of soft tissue sarcomas and their incidence in the general population is estimated at 0.001% [4]. However, certain locations such as the head and neck are exceptional because it is estimated that they represent less than 1% of malignant tumors in this region [1] [2]. Some may originate from peripheral nerve cells, develop de novo, or result from malignant transformation of pre-existing neurofibromas and 50% of neurofibrosarcomas are found in patients with neurofibromatosis type 1 or neurofibromatosis type 1 disease Von Recklinghausen's disease which is an autosomal dominant genetic disease [3]. Clinically, the tumor presents as a rapidly growing mass that is often associated with pain [2] [3]. Surgery is the main therapeutic modality. Radiotherapy is generally used as an adjuvant to achieve better local control. For some authors, radiotherapy as a main modality has been reported [5]. We report our diagnostic and therapeutic experience in relation to a case of isolated cervical neurofibrosarcoma developed at the expense of the cervical sympathetic trunk and then discuss our case with those found in the literature.

2. Clinical Case

We report the case of a 45-year-old female housewife patient (DT) with no medical or surgical history admitted to the department for left laterocervical swelling. The onset of the disease dates back to 6 months after admission, marked by a progressive left laterocervical swelling, with permanent evolution associated with left neck pain, torticollis, and dysphagia to solids. The cervical examination noted a left laterocervical swelling of approximately 8 cm/6 cm, hard, fixed, painful to palpa-

tion, with regular contours, non-blowing, non-beating, skin with an inflammatory appearance (Figure 1), poor oral hygiene with multiple carious teeth, wet and mobile tongue, a bulging of the left lateral wall of the oropharynx pushing the left tonsil inward. A Cervical CT scan revealed a weakly enhanced laterocervical tissue hypodensity pushing outward the jugulocarotid axis communicating with the conjugation foramina of C3, C4 and C5 with mass effect on the left vascular-nervous axis of the neck and the pharynx (Figure 2). The standard preoperative biological assessment noted a hemoglobin level of 11.04 g/dl with a hematocrit of 41.06%, and the chest X-ray, frontal and profile, was unremarkable. A transfusion of two bags of isogroup isorhesus blood pre- and postoperatively was performed. Our approach used a wide cervicotomy by Paul ANDRE "J" incision removing the skin, the subcutaneous cellular tissue and the platysma. The reclination of the anterior flap gave us access to the omohyoid and sternocleidomastoid muscles, which we sectioned (Figure 3). The jugulocarotid and vagal vasculonervous bundle were dissected from the capsule of the mass, and the spinal nerve was preserved (Figure 3). The tumor was freed from the cervical vertebrae and the pharynx. The resection was complete with some ipsilateral cellulolymphatic flows (Figure 4). We established a 2 mm margin from the edges of the lesion. This required us to perform a resection of the transverse processes and the superior articular processes of C3, C4 and C5. We performed a layer-by-layer closure on a drain (Figure 5). Histology revealed bundles of spindle cells with large and hyperchromatic nuclei; the nucleoli are visible and abnormal mitoses which are in favor of a neurofibrosarcoma (Figure 6). Immunohistochemistry allowed us to confirm the diagnosis of neurofibrosarcoma. The wound healed around the 15th day. The patient was referred for radiotherapy after a multidisciplinary consultation meeting. The clinical evolution after 6 months of post-radiotherapy monitoring was favorable. No functional discomfort was noted.

Figure 1. Front and profile view of the laterocervical mass.

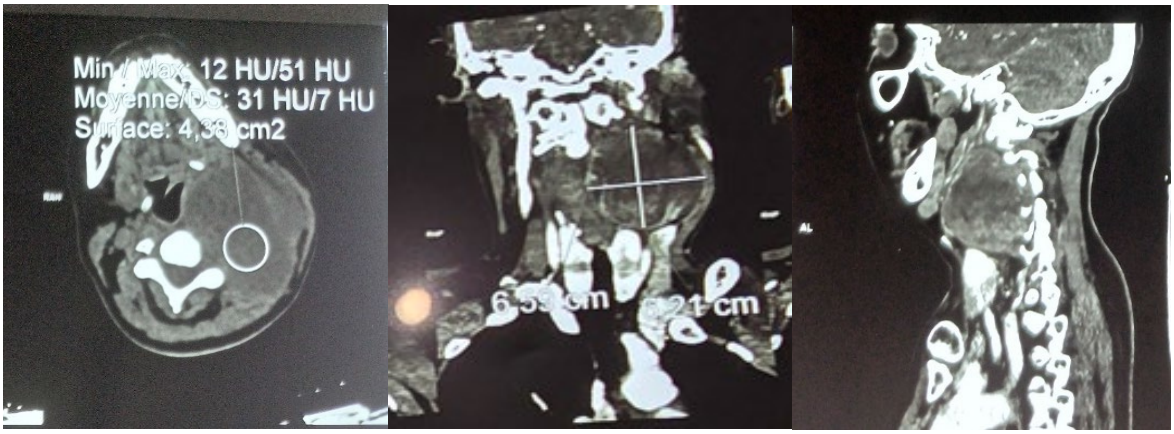

Figure 2. Axial, coronal and sagittal CT image showing a large, weakly enhanced tissue hypodensity occupying the laterocervical and paravertebral region pushing outwards the jugulocarotid axis with mass effect on the pharynx.

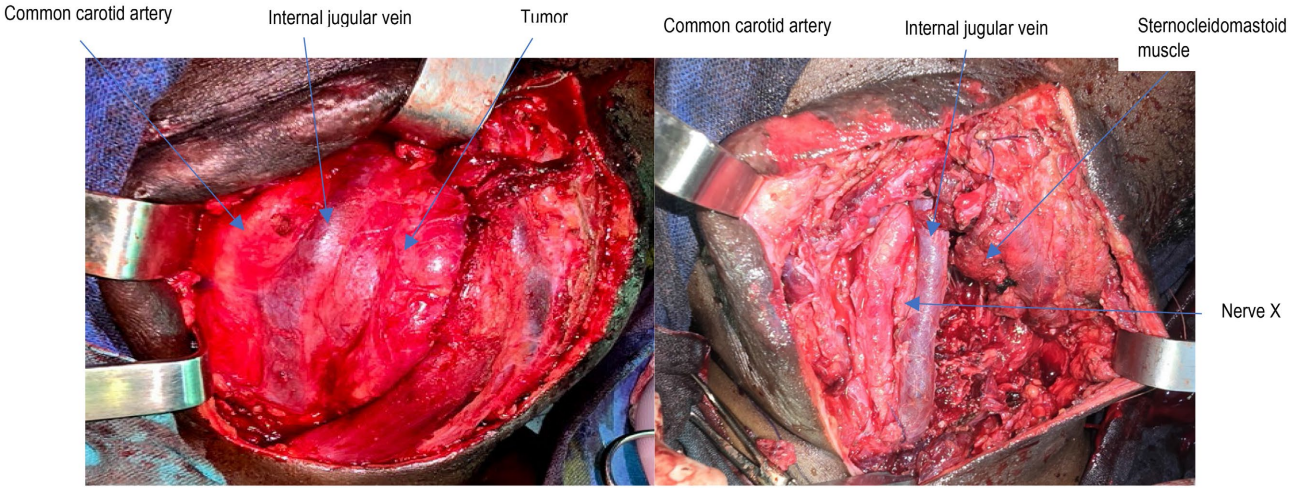

Figure 3. Exposure of the mass during surgery.

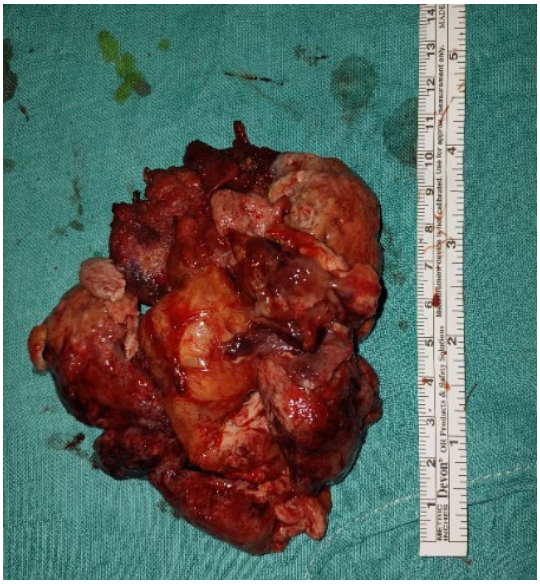

Figure 4. surgical part.

Figure 5. Postoperative wound closure.

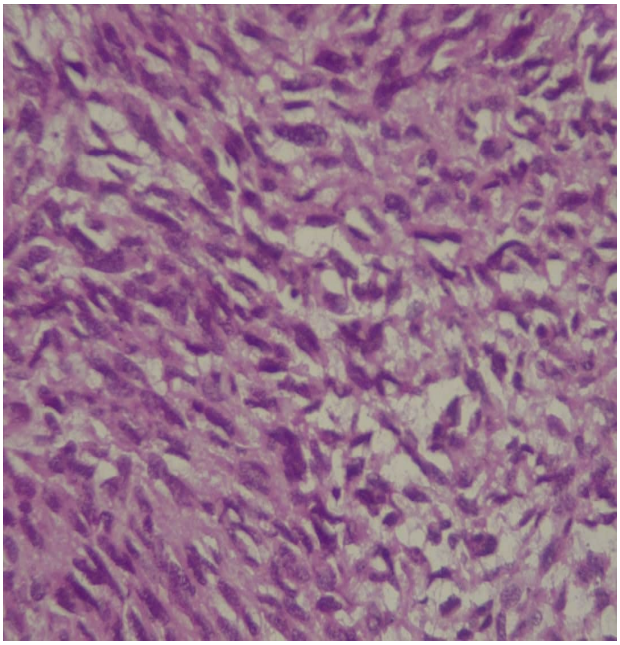

Figure 6. Bundles of spindle-shaped cells with large, hyperchromatic nuclei, visible nucleoli, and abnormal mitoses.

3. Discussion

Neurofibrosarcoma, also called malignant Schwannoma or neurogenic sarcoma, is a tumor of the peripheral nervous system that develops in the nerve sheath [1] [2]. It is a rare clinical entity, most often reported as a clinical case. In the last 50 years, only about a hundred cases located in the head and neck have been reported [2]. The incidence of this tumor is higher in patients with Von Recklinghausen disease or type 1 neurofibromatosis, in whom it can reach 5% [1] [6]. The history of

cervical irradiation is also recognized by several authors as a risk factor for the occurrence of neurofibrosarcoma [1] [6]. In the literature, we have the link between tobacco and alcohol as the etiopathogenesis of neurofibrosarcoma. Our patient had no notion of cervical irradiation, nor signs of neurofibromatosis type 1 which allowed us to conclude that it was de novo neurofibrosarcoma. This tumor affects all ages, the sex ratio is equal to one, and it develops on nervous structures such as the brachial plexus, the cervical sympathetic chain and the cranial nerves and their branches [2]. In our patient, the tumor was left laterocervical and communicated with the conjugation foramen of the cervical vertebrae C3, C4 and C5 and was in continuity with the left cervical sympathetic trunk.

Patients with neurofibrosarcoma most often present with a rapidly increasing laterocervical mass that may be painless or cause severe but often intermittent pain initially. A notion of recent trauma to the anatomical region concerned is sometimes found and the associated signs are a function of compression and infiltration of the affected neighboring structures [7]. The circumstance of discovery in our patient was a rapidly evolving left laterocervical mass at 6 months associated with neck pain and torticollis, and dysphagia to solids. These signs were linked to compression because after surgery, they disappeared.

Computed tomography (CT) and magnetic resonance imaging (MRI) are both important in the exploration of this tumor to determine its location, its locoregional extension or metastases, and the relationships of the tumor with the noble organs [3]. Our patient did not present signs of distant metastasis. On CT, neurofibrosarcoma presents as a hypodense mass that enhances intensely and heterogeneously after injection, unlike neurofibromas [2]. Our patient's mass was hypodense before the injection of the contrast agent and after the injection of the latter it took the contrast agent which favors diagnosis. Our patient did not undergo MRI because of a limited technical platform. MRI is more efficient than CT in the exploration of soft tissues because it allows us to give more details about the locoregional relationships of the tumor. Significant signs of malignancy on MRI are four in number, which are an increase in the largest dimension of the mass, presence of an enhanced peripheral pattern, perilesional edema and an intratumoral cystic lesion [2] [8]. In our case, the MRI could not be performed due to the high cost and the lack of coverage of medical expenses by health insurance.

In the histology of neurofibrosarcoma we generally find the presence of spindle cells, often arranged in bundles, alternating cellular zones and more myxoid zones [1] [3] [4]. In our patient, we found bundles of spindle cells with large and hyperchromatic nuclei, the nucleoli are visible and abnormal mitoses which are the signs of histological malignancy. Electron microscopy and immunohistochemistry are very accurate in the diagnosis and the S-100 protein, the biological marker used, is positive in 17% to 56% of cases of neurofibrosarcoma [1] [3]. Immunohistochemistry allowed us to make the diagnosis in our patient.

Surgical excision of the tumor is the standard treatment for sarcomas. Radiotherapy is important in the therapeutic arsenal of sarcomas; it is exclusive for in-

operable sarcomas. In the postoperative period, it is systematic for soft tissue sarcomas except for low-grade tumors with wide resection [3] [7]. We performed a cervicotomy for total excision of the mass and the patient was referred for radiotherapy after a multidisciplinary consultation meeting.

The 5-year survival rate is 50% in patients with de novo neurofibrosarcoma. This figure falls to 15% in cases of association with neurofibromatosis [2].

Following this study, we propose three factors that may influence the recurrence of peripheral nerve sarcomas: the stage of development of the lesion, the margins of surgical excision and the early start of radiotherapy. Our clinical case demonstrates the occurrence of neurofibrosarcomas in a subject with no known factors, in particular, a family history of von Recklinghausen's disease or a history of alcohol and tobacco use. Hope rests on future research into the etiopathogenesis of this condition.

4. Conclusion

Neurofibrosarcoma is a malignant tumor that can develop throughout the peripheral nervous system. Its symptoms vary depending on the region involved, its progression is rapid, and its treatment is multidisciplinary. Surgery is the main therapeutic arsenal, often associated with adjuvant radiotherapy.

Ethical Approval Statement

Informed Consent: It was purely scientific work aimed at improving the grip of the in charge in the field of head and neck surgery, the ethical standards have been strictly for the study participant with their informed consent, and respect for anonymity.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Tekaya, R., Hamdi, W., Azzouz, D., Bouaziz, M., Jaafoura, M.H., Ladeb, M.F., *et al.* (2008) Névralgie cervicobrachiale révélatrice d'un neurofibrosarcome (MPNST) cervical. *Revue Neurologique*, **164**, 82-86. https://doi.org/10.1016/j.neurol.2007.07.006

[2] Dalil, A., Fokouo Fogha, V., Evehe Vokwely, J., Sougou, E. and Miloundja, J. (2018) A Case Report of Cervical Neurofibrosarcoma: Clinical Presentation, Treatment and Outcome. *International Journal of Surgery Case Reports*, **42**, 175-178. https://doi.org/10.1016/j.ijscr.2017.12.019

[3] Lamhar, L.M.M. (2022) Sarcimatous Degeneration of Von Recklinghausen's Disease Regarding a Case. Ph.D. Thesis, Faculty of Medicine and Odontostomatology.

[4] Benevello, C. (2013) A Rare Case of Malignant Schwannoma of the Brachial Plexus. *World Journal of Surgical Procedures*, **3**, 1-3. https://doi.org/10.5412/wjsp.v3.i1.1

[5] Pandey, M., Chandramohan, K., Thomas, G., Mathew, A., Sebastian, P., Somanathan, T., *et al.* (2003) Soft Tissue Sarcoma of the Head and Neck Region in Adults. *Interna-*

tional Journal of Oral and Maxillofacial Surgery, **32**, 43-48.
https://doi.org/10.1054/ijom.2001.0218

[6] Touil, H., Briki, S., Karray, F. and Bahri, I. (2015) Malignant Peripheral Nerve Sheath Tumor of the Superficial Cervical Plexus with Parotid Extension. *European Annals of Otorhinolaryngology, Head and Neck Diseases*, **132**, 93-95.
https://doi.org/10.1016/j.anorl.2013.11.012

[7] Julieron, M., Robin, Y., Penel, N. and Chevalier, D. (2013) Sarcomi del capo e del collo. *EMC—Otorinolaringoiatria*, **12**, 1-19.
https://doi.org/10.1016/s1639-870x(13)66027-4

[8] Wasa, J., Nishida, Y., Tsukushi, S., Shido, Y., Sugiura, H., Nakashima, H., *et al.* (2010) MRI Features in the Differentiation of Malignant Peripheral Nerve Sheath Tumors and Neurofibromas. *American Journal of Roentgenology*, **194**, 1568-1574.
https://doi.org/10.2214/ajr.09.2724