European Annals of Otorhinolaryngology, Head and Neck diseases 136 (2019) 455–460

Available online at ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

Original article

Peripheral neck nerve tumor: A 73-case study and literature review

K. Boumaza [a], G. Michel [a], C. Salaud [b], C. Bossard [c], F. Espitalier [a], O. Malard [a,*]

[a] Service d'ORL et de Chirurgie Cervico-Faciale, CHU de Nantes Hôtel-Dieu, 1, place A. Ricordeau, BP 1005, 44093 Nantes Cedex 01, France
[b] Service de Neurotraumatologie/Neurochirugie, Centre Hospitalier Universitaire de Nantes, Hôtel-Dieu, 1, place A. Ricordeau, BP 1005, 44093 Nantes Cedex 01, France
[c] Service d'Anatomopathologie, Centre Hospitalier Universitaire de Nantes, Hôtel-Dieu, 1, place A. Ricordeau, BP 1005, 44093 Nantes Cedex 01, France

ARTICLE INFO

Keywords:
Neck nerve tumor
Schwannoma
Neurofibroma
Neuroblastic tumor
Malignant peripheral nerve sheath tumor

ABSTRACT

Objectives: Peripheral neck nerve tumors are rare and mostly benign neoplasms. The exceptional malignant forms are very aggressive, and diagnosis is difficult. The objective of this study was to evaluate diagnostic and therapeutic management and identify possible predictive factors.
Material and methods: A retrospective study was conducted of 73 patients treated for peripheral neck nerve tumor between 1995 and 2015.
Results: Mean age was 44 years. The main presenting symptom was a cervical mass, isolated or associated with signs related to the affected nerve structure. Diagnosis was suspected by slow progression of a firm mass, featuring T1 hyposignal and T2 hypersignal on magnetic resonance imaging. Surgery was performed in 99% of cases, completed by adjuvant chemotherapy in case of malignant neuroblastic tumor. Type 1 neurofibromatosis and sudden increase in mass with or without associated pain suggested malignant transformation. Age below 10 years suggested neuroblastic tumor.
Conclusion: Neck nerve tumors are very often benign with low degenerative potential. Surgery is the treatment of choice after risk/benefit analysis. However, there is no clearly defined consensus regarding the timing of surgery for these lesions.

1. Introduction

Neck nerve tumors are rare primary tumors, but have been known since 1742 when Haller discovered the carotid corpuscle; then in 1803 Rodier introduced the term "neuroma" for tumors of the peripheral nerves [1].

A neck mass is the usual clinical presentation of neck nerve tumor. The mass is often isolated and painless although it may be an esthetic blemish, but can also be associated with clinical signs related to the function of the affected nerve.

Clinical diagnosis is hampered by the numerous differential possibilities [2] in this location, but may be suggested by slow progression [2], by relative absence of symptoms, and on imaging. Recent advances in immunohistochemistry and imaging allow increasingly precise diagnosis [3–5]. MRI is the imaging examination of choice [6,7], although definitive diagnosis obviously depends on pathologic analysis [7].

The lesions originate in various neural elements of the neck, both major, such as cranial nerves IX, X, XI and sometimes XII or the cervical or brachial plexi, and their small branches [1]. Histology is heterogeneous and most often benign, except for exceptional malignant peripheral nerve sheath tumors (MPNST) and malignant neuroblastic tumors.

The various denominations necessitate a precise nosological classification according to histologic type and to origin: peripheral nerve, sympathetic ganglion or paraganglion [1].

No specific risk factors or population have been identified, except for phacomatoses, the most frequent of which are neurofibromatoses. Overall prognosis is excellent, except in rare malignant forms, and recurrence is relatively exceptional.

Treatment is mainly surgical. There is, however, no consensus as to timing or follow-up.

The aim of the present study was to assess diagnostic and therapeutic practices in these rare head and neck tumors.

2. Material and methods

A single-center retrospective study reviewed files for 76 patients treated for neck nerve tumor in the university hospital center of Nantes, France, between January 1995 and December 2015.

The study population was based on the Nantes university hospital pathology department database. Inclusion criteria comprised

* Corresponding author.
E-mail address: omalard@chu-nantes.fr (O. Malard).

https://doi.org/10.1016/j.anorl.2019.09.005
1879-7296/

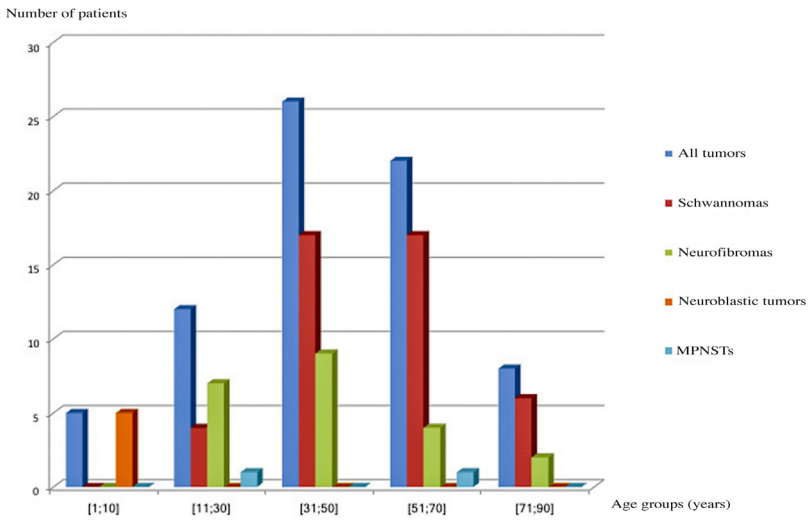

Fig. 1. Age in overall population and by tumor type.

neural tumor confirmed on pathology examination of specimen or biopsy. The requisite information was extracted from the clinical and imaging files.

Data were epidemiological (age, gender), clinical (revelation, symptoms, location, monitoring, progression), pathological (tumor type, resected volume, resection margins) and paraclinical (ultrasound, CT, MRI). These data identified the affected population, tumor mass and local extension, clinical impact and treatment decision. The histologic data established the precise diagnosis of tumor type.

Paraganglioma was excluded: although originating in nerve crest paraganglions, the cells migrate beyond the sympathetic chain and acquire a glandular character as neuroendocrine tumor. Vestibular nerve schwannoma and acoustic neuroma were also excluded, being located outside the study region.

The study objective was to assess diagnostic and therapeutic practices for these rare neck tumors, specify population characteristics in terms of histologic type, and screen for pathologic and prognostic predictive factors according to age, location and diathesis.

Statistical analysis used Chi^2 test for qualitative variables and Student t test for quantitative variables.

3. Results

3.1. Epidemiology

Mean age was 44 years (range, 1–90 years), varying according to histologic type: 51 years for schwannoma, 41 years for neurofibroma, 34.5 years for MPNST (with only 2 patients), and 4 years for neuroblastic tumor (Fig. 1). There was slight female predominance overall, with 40 females and 33 males (non-significant: $P = 0.79$); there was, however, a significant sex difference for two forms: neuroblastic tumor, concerning only under-10 year-olds, with 4:1 female predominance; and MPNST, with 2 patients, both male.

Overall, the patients had no particular ENT history: notably, no radiation therapy or cervical trauma. In 2 of the 3 cases of serendipitous discovery, the tumor was found during neck dissection following treatment of a malignant tumor: squamous cell carcinoma of the tongue, and thyroid carcinoma, respectively; in the third case, discovery followed treatment of a multinodular goiter. Only 1 patient had history of multiple schwannoma.

All patients with neurofibroma had at least 1 other non-ENT location and/or history of type-1 neurofibromatosis de type 1.

Half of the patients (49%) were managed in the ENT department, and half (51%) in other departments: neurosurgery, maxillofacial surgery, plastic surgery, visceral surgery or pediatric surgery.

Files were incomplete for 3 patients: 1 in neurosurgery and 2 in ENT. These patients were treated in 1995, when computer files were still little used; the relevant data had not been supplied on the paper document.

3.2. Clinical presentation

In most cases (84%; 61 patients), discovery followed at least 1 clinical symptom. In 12% of cases ($n = 9$), discovery was serendipitous, and in 4% ($n = 3$) not recorded in the file.

In 43% of cases (26 patients), the presenting symptom was simply an isolated neck mass, painless but causing an esthetic blemish. In 8% (5 patients), there were 1 or more associated symptoms related either to compression by the mass or to the function of the affected nerve. In 49% of cases (30 patients), there was no neck mass and the presenting symptom was pharyngeal discomfort or pain, dysphagia, dysphonia with hemilaryngeal paresis, or painful upper limb deficit related to thoracic outlet syndrome or Bernard-Horner syndrome.

There were no cases of impaired general health or superinfection.

The interval between first symptom onset and first consultation was specified in only 10 files, and was very variable: from 3 days to several years, without further detail.

The interval between first consultation and surgery ranged between 7 days and 18 months. In 3 cases of serendipitous discovery, the tumor (lingual or thyroid carcinoma) was resected within a neck-dissection specimen, or around a multinodular goiter.

Fifty-six percent of tumors (41 patients) were right cervical and 30% (22 patients) left, although the difference was non-significant ($P = 0.90$); 3% (2 patients) were bilateral, and 8% (6 patients) medial. Medial tumors were either nuchal or intracanal medullary. In 3% of cases (2 patients: 2 of the 3 incomplete files), the file did not specify laterality (Table 1).

One schwannoma patient died of a gastric cancer diagnosed shortly after the neural tumor.

Table 1
Epidemiological and clinical data.

Characteristics	*N* = 73	%
Age: mean (range)	45 (1–90)	
Gender (M/F)	33/40	
Revelation		
Serendipitous	9	12%
MD	3	4%
Clinical symptoms	61	84%
Isolated neck mass	26	43%
Discomfort/Pain	20	33%
Neck mass + other symptom	5	8%
Sensorimotor deficit in a limb	4	6%
Dysphonia	3	5%
Dysphagia	2	3%
Bernard-Horner syndrome	1	2%
Location		
Neck, non-specified	33	45%
Neck, specified:	40	55%
Posterior	7	17%
Parapharyngeal	5	13%
Supra-clavicular	5	13%
10th cranial nerve	4	10%
Cervical sympathetic plexus	4	10%
11th cranial nerve	3	7%
Medullary	3	7%
Retrostyloid	3	7%
Brachial plexus	2	4%
12th cranial nerve	1	3%
Lingual nerve	1	3%
Retropharyngeal	1	3%
Submandibular	1	3%
Laterality		
Right	41	56%
Left	22	30%
Medial	6	8%
Bilateral	2	3%
MD	2	3%

MD: missing data.

3.3. Paraclinical data

Imaging was performed in 51 patients (70%) while 19 (26%) had no morphologic work-up. Two of the latter cases were schwannomas: 1 patient with history of multiple schwannoma, and a 90 year-old patient who had biopsy alone. Another case was of neuroblastic tumor in a 1 year-old, diagnosed on resection-biopsy, leading to full morphologic work-up. The other patients without imaging had neurofibroma, mainly subcutaneous and causing blemish; diagnosis was clinical.

Thirty-three of the patients with imaging (65%) had neck MRI, alone or associated to neck CT, revealing an aspect typical of neural origin, with T1 hyposignal and T2 hypersignal. Fourteen (27%) had neck CT without MRI; 3 (6%) had ultrasound alone; and 1 (2%), with history of thyroid neoplasia, had PET-scan.

Fifty patients (68%) had no preoperative pathology analysis. Of the other 20 (28%), 8 (40%) had fine-needle aspiration, 7 (35%) simple biopsy, and 5 (25%) resection-biopsy. For the other 3 patients, data were missing.

Fine-needle aspiration was consistently non-contributive, whereas biopsy consistently established definitive diagnosis, except in 1 patient in whom biopsy suggested neurofibroma while definitive specimen analysis showed ganglioneuroma.

Pathology identified 5 tumor types: 44 schwannomas (60%), 22 neurofibromas (30%), 5 neuroblastic tumors (7%) including 3 neuroblastomas (4%), 2 ganglioneuromas (3%), and 2 MPNSTs (3%) (1 sporadic neurofibrosarcoma and 1 low-grade malignant transformation of neurofibroma in a patient with type-1 neurofibromatosis) (Fig. 2).

Histology in patients undergoing resection found safe margins in 89% of cases (62 patients). Resected tumor size ranged from 1 to 11.4 cm (median, 3.8 cm).

The affected neural structure was not identified in 55 patients (75%) and identified in 18 (25%): cranial nerve X in 4 patients, cervical sympathetic plexus in 4 patients, cranial nerve XI in 3, brachial plexus in 2, lingual nerve in 1, and cranial nerve XII in 1. Three patients had medullary locations.

3.4. Treatment

Treatment was surgical in 99% of cases (72 patients with nerve tumor diagnosed on pathology). The 3 patients in whom pathology diagnosed neuroblastoma received complementary chemotherapy; 1, with residual tumor 6 months after chemotherapy, underwent salvage jugulo-carotid neck dissection.

The 1 patient not operated on had simple annual clinical and radiological surveillance with neck CT, due to advanced age and symptomatology comprising only pharyngeal discomfort and intermittent reflex otalgia.

3.5. Progression and follow-up

Follow-up varied greatly between individual patients and specialties. Apart from the 3 incomplete files and 1 patient who died of other cause (gastric cancer), 38% of patients (26 patients) had no follow-up in the initial department; follow-up here might have identified secondary neurologic deficits indicating the nerve of origin. Neurofibroma, which was predominantly subcutaneous, was associated with few neurologic deficits.

The other 43 patients (62%) had follow-up ranging between one 6-month consultation to consultations every 2 or 3 months then once yearly, notably for schwannoma and MPNST.

All 5 neuroblastic tumors concerned under-10 year-olds. The 2 patients with ganglioneuroma were seen only once, at 2 and 6 months respectively, and were not followed up in pediatrics either. The 3 patients with neuroblastoma did not have a postoperative check-up, but were followed in oncopediatrics after initiation of chemotherapy.

One of the 2 patients with MPNST, aged 17 years, underwent surgical revision at less than 3 weeks after primary surgery, due to insufficient resection margins; he developed ipsilateral left neck recurrence at 6 years, treated again by resection. Twenty-two years after the initial diagnosis of left brachial plexus MPNST, he developed an ipsilateral synovial sarcoma, once again treated by resection.

The second patient, aged 52 years, was managed by surveillance in the Nantes reference center, as he had type-1 neurofibromatosis, with longstanding right cervical neurofibroma. The volume rapidly increased to 6 cm on the long axis, without change in MRI signal intensity. MRI found features of nerve tumor (T1 iso-hyposignal T1, T2 hypersignal and gadolinium uptake), without definitively diagnosing malignancy.

The 22 patients with neurofibroma (30%) showed type-1 neurofibromatosis. Slightly more than half ($n = 13$) were followed in the Nantes neurofibromatosis reference center. The diagnosis was known ahead of surgery in more than half of the cases. Surgery was indicated for discomfort associated with pain in 11 cases, blemish in 6, and neurologic deficit in 5.

The 3 patients with serendipitous diagnosis after treatment for another pathology had treatment for the first disease (lingual neoplasia, thyroid neoplasia, multinodular goiter, respectively), and histologic specimen analysis revealed the nerve tumor.

One patient died 3 months after resection-biopsy of the neck lesion, which was found to be a schwannoma; death followed a gastric cancer diagnosed shortly after the nerve tumor.

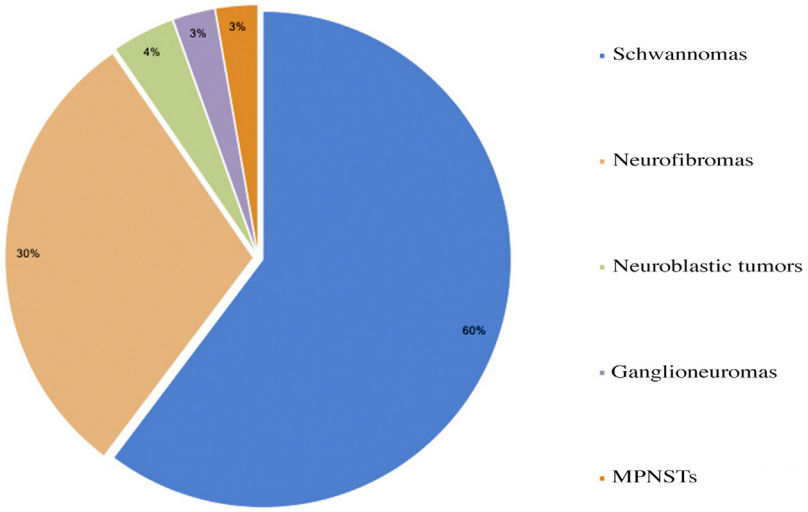

Fig. 2. Histologic sub-types of head and neck nerve tumors.

Postoperative consultation for the 43 patients followed up in the same department found sequelae in 63% of cases (27 patients). Eight of these patients showed Bernard-Horner syndrome; 7 had cranial nerve X palsy, 3 of whom underwent vocal-fold medialization; 5 showed limb motor deficit; 5 showed postoperative neuralgia; 2 showed first-bite syndrome [8]; 2 showed dysphagia; 1 showed cranial nerve XII paresis; and 1 showed TMADS (temporomandibular joint algo-dysfunctional syndrome).

Given that 38% of patients were not followed up in the original department, deficit duration is difficult to estimate. Most deficits seemed to be transient, as patients did not return to consultation. However, the 2 sequelae persisting at postoperative consultation in the 62% of patients followed up were 3 of the 8 cases of Bernard-Horner syndrome, and 3 of the 7 recurrent nerve palsies, 2 of which were treated by vocal-fold medialization at about 1 year.

3.6. Recurrence and progression

Three of the 43 patients with follow-up (7%) had recurrence: 1 multiple schwannoma at 3 years, 1 MPNST at 6 years, and 1 malignant neuroblastic tumor 6 months after chemotherapy, requiring neck dissection. Two patients (5%) showed disease progression: the 1 non-operated patient, showing slowly progressive retropharyngeal schwannoma, and a patient with medullary neurofibroma with positive resection margins.

3.7. Predictive factors

Mean age was significantly greater in schwannoma (51 years; $P = 0.0009$), and youngest in neuroblastic tumor (4 years; $P < 0.001$).

There were no sex differences, except in MPNST, with only 2 patients, both male ($P = 0$; statistical analysis non-applicable).

Mean tumor size was greater in neuroblastic tumor (5.25 cm) than elsewhere (2.93–3.89 cm), but differences were non-significant, with *P*-values ranging between 0.20 and 0.65 for comparison of mean size between the 4 tumor groups.

A neck mass was found in 29% of cases of schwannoma, 72% of neurofibroma, 80% of neuroblastic tumor, and 100% of MPNST. Pain was reported in 59%, 32%, 20% and 50% of cases, respectively, but again these differences were non-significant, due to small sample sizes.

4. Discussion

Neck nerve tumor is rare, whence the paucity of prospective studies assessing treatment.

The present study analyzed all forms except paraganglioma. They can be distinguished only on histology and immunohistochemistry, imaging findings being similar for all.

Most nerve tumors involve hyperexpression of S100 protein [6,9], determining their neurectodermal character. In schwannoma, however, the cells are spindle-shaped with palisading nuclei forming Verocay bodies. In neurofibroma, cells are also spindle-shaped, but with long nuclei detached from a fibrillar myxoid ground. Neuroblastic tumor comprises neuroblasts, the maturity of which determines malignancy according to mitosis-karyorrhexis index (MKI: high levels indicating malignancy). In MPNST, the histologic aspect is a fairly dense fibroblastic tissue with sparse cellular cytoplasm and ovoid nuclei, S100 expression being low or absent.

Each tumor type shows specific epidemiological, clinical and progression features.

4.1. Schwannoma

According to the literature, 25% to 45% of schwannomas are located in the head and neck region [1,6,10]. Age at diagnosis ranges between 30 and 60 years [9]. Likewise, in the present series, mean age was 51 years. Like in the present series, there seems to be no gender predominance [6,9,10].

In published series, schwannoma is usually isolated [6], as in the present series, with only 1 case of multiple schwannoma, without phacomatosis and notably without type-2 neurofibromatosis.

Findings for the nerve of origin matched the literature: < 5% brachial plexus, 2–5% vagus nerve, and > 5% other cranial motor nerves such as the hypoglossal nerve [1,6,11]. On the other hand, 11% of the present locations were parapharyngeal, compared to < 5% in the literature [3,4]; further studies of this location would be worthwhile.

The literature reports 12% preoperative vocal-fold palsy in vagus schwannoma [6,10]; in the present series, preoperative hemilaryngeal paresis was found in 1 of the 4 vagus schwannomas (25%); the postoperative rate was 3 out of 4 (75%), compared to 55% in the literature [10], testifying to the importance of early vocal rehabilitation. The deficit seems to be transient, especially if the vagus nerve is conserved; if deficit proves definitive, later vocal-fold medialization may be proposed.

Schwannoma is well-encapsulated [4], and resection is often extra-capsular, sparing the nerve of origin [3]. Three of the present series hade intra-capsular resection: i.e., 7% of schwannomas.

Sequelae were found in 100% of intra- and 37% of extra-capsular resections, whereas rates are identical in both cases in the literature [2], although lack of power precludes any firm conclusion.

Progression is very slow, at 2.5–3 mm per year [2,9]. Malignant transformation is rare, at 1.5–10% [1,2]. Associated neurofibromatosis is reported in 27–70% of cases [1]. In the present series, no schwannomas showed degeneration.

Surgery is the unanimous treatment of choice, although optimal timing is undetermined. As these tumors are benign, conservative management with clinical and/or radiological surveillance may be indicated in first line in the absence of functional or esthetic impairment or risk of compression [10].

4.2. Neurofibroma

Thirty-five percent of neurofibromas are in the head and neck region [1]. Mean age at diagnosis was 41 years in the present study, versus 20–30 years in the literature [7].

All patients in the present series presented type-l neurofibromatosis (NF1), versus 10–40% rates in the literature [1,7]. The discrepancy may be due either to recruitment bias, as the patients in the present series were mainly referred by the regional neurofibromatosis reference center, or to systematic NF1 screening in case of diagnosed neurofibroma.

Malignant transformation is mainly associated with NF1, with rates varying between 2% and 29% [7]. Latency ranges between 10 and 20 years [1]. Malignancy is strongly suspected in case of rapid tumor volume growth, with possible onset of pain, and a more infiltrating character and more stony-hard on palpation.

Recurrence after resection is rare, but higher in case of incomplete resection, in head and neck locations compared to trunk and limbs, and in under-10 year-olds [7].

In the present series, there was no recurrence after resection. A single patient showed continued progression after incomplete resection of a medullary tumor.

Surgery should be considered on a case-by-case basis. Neurofibroma can be difficult to isolate from the underlying neural structure, unlike schwannoma, which is generally better encapsulated. Complete resection may sacrifice the nerve, although usually without consequences in subcutaneous locations; in lesions on larger nerves, however, the risk of iatrogenesis is higher. Therefore, in tumors involving major nerves with risk of intraoperative lesion, either limited resection allowing histologic control to spare the nerve or else simple surveillance should be considered [7,12].

4.3. Neuroblastic tumor

These are the most common solid tumors in children [13]. Four types are distinguished by the International Neuroblastoma Pathology Classification (INPC): in order of increasing malignancy, ganglioneuroma, nodular ganglioneuroblastoma, mixed ganglioneuroblastoma, and neuroblastoma [13,14].

Classically, 3–5% of neuroblastic tumors are cervical, 10% of which are malignant [15]. Onset is sporadic in 99% of cases. Malignant neuroblastic tumor accounts for 9–10% of childhood cancers [14–16], and is the third most frequent cancer in general oncopediatrics, after leukemia and cerebral tumor. The most frequent locations are mediastinal or retroperitoneal [14,15,17].

Mean age at diagnosis was 6 years for ganglioneuroma and 2 years for neuroblastoma in the present study, compared to 18 months in the literature [17]. There was female predominance in the present series, whereas the literature reports slight male predominance [17]. However, small sample sizes in the various studies preclude precise analysis.

Clinical presentation is non-specific, and radiological aspect does not always correspond to a classic nerve tumor; on MRI, there may be heterogeneous hypersignal on T1 and hyposignal on T2 [14]. Age < 10 years suggests this etiology, but only histology provides definitive diagnosis [14,15].

Erol et al. [13] and Alvi et al. [17] recommend urinary catecholamine assay. MIBG scan completes morphologic work-up, which includes other examinations by the oncopediatricians who are usually in charge of these cases.

For most authors, treatment is surgical [13–17], completed by chemotherapy in case of high malignancy risk assessed following the International Neuroblastoma Risk Group (INRG) [13]. This score takes several factors into account, and notably stage on the International Neuroblastoma Staging System (INSS), age, histologic type, differentiation grade, MYCN oncogen status, 11q chromosome status and DNA ploidy.

4.4. Malignant Peripheral Nerve Sheath Tumor (MPNST)

MPNST is very rare, with the literature mainly consisting of case reports [5,18,19]. It is highly aggressive. As well as MPNST, it is also known as neurofibrosarcoma, neurogenic sarcoma or sometimes malignant schwannoma [5,18]. Incidence is 0.001% [5,20]. Location is head and neck in about 10–15% of cases [18,19]. MPNST accounts for 10% of cervical sarcomas [19]. In the present series, there were only 2 cases over a 20-year period. Half of MPNSTs implicate degeneration of NF1 after 10–20 years' progression; the other half are *de novo* [18,19]. These literature data agree with the present findings.

Onset is reported at any age [19], but notably between 20 and 50 years [18], as in the present 2 cases. No predominance according to gender is reported [18,19], although both of the present cases were male.

Diagnosis is difficult, as MPNSTs resemble benign nerve tumor, presenting clinically as an isolated mass or with minor clinical signs, and radiologically showing T1 hyposignal and gadolinium fixation on MRI [5]. However, rapid tumor growth and increasing pain, with or without neurologic deficit, can suggest malignancy or degeneration. Definitive diagnosis is provided only on histology and, especially, immunohistochemistry [5]; biopsy is therefore mandatory in NF1 with suspect mass, to rule out MPNST.

Biopsy evidence enables surgical resection to be indicated. There is no standard treatment, but surgery is to be preferred, often completed by adjuvant radiation therapy with or without concomitant chemotherapy. The contribution of these adjuvant treatments has not been codified, and they have not been shown to improve survival [5]. Radiation therapy is also recommended in case of recurrence or impossibility of surgery. Exclusive chemotherapy is indicated for metastatic locations [5,19].

Prognosis is poor, with 5-year survival at 34–52% [20] or 50% in *de novo* MPNST [19]. Factors for poor prognosis comprise tumor size > 5 cm, intralesional resection margins, metastasis at time of diagnosis, advanced age, and associated NF1 [5].

In the present series, the 2 MPNST patients received annual follow-up, and were alive at 6 years. The younger patient, with *de novo* MPNST of the left brachial plexus, showed recurrence at 6 years and was reoperated; 22 years after primary surgery, he developed a synovial sarcoma in the irradiated ipsilateral supraclavicular territory, which was resected, whereas the primary tumor remained in remission.

5. Conclusion

Neck nerve tumor is rare, and clinical diagnosis is difficult. There are various histologic types, presenting clinical similarities.

Treatment is not fully codified. The imaging technique of choice is neck MRI, suggesting diagnosis and ruling out the most frequent differential diagnoses.

The affected population is heterogeneous. Patients with neurofibromatosis are at higher risk of neck nerve tumor, with slightly higher risk of malignant transformation, due to lesion age and progression.

Definitive diagnosis is on histologic and immunohistochemical analysis of the resection specimen or biopsy.

The treatment of choice is surgical resection in most cases, but this is to be decided on by case-by-case risk/benefit analysis. Resection should be wide and at least extra-lesional, to reduce the risk of early recurrence. It may be completed by radiation therapy in MPNST and by chemotherapy in malignant neuroblastic tumor or metastatic locations.

Follow-up should be regular, and adjusted to the specific clinical situation. Neck MRI is the examination of choice for monitoring in case of deep tumor, doubtful clinical examination, or onset of new symptoms.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Pessey J-J, Serrano E, Percodani J. Tumeurs nerveuses du cou. EMC–Oto-rhino-laryngologie 1994;1–0 [20-875-C10].
[2] Vijendra SS, Rao RA, Prasad V, Haseena S, Niprupama M. A giant vagal schwannoma with unusual extension from skull base to the mediastinum. J Cancer Ther Res 2005;11(4):970–3.
[3] Sato Y, Imanishi Y, Tomita T, Ozawa H, Sakamoto K, Fujii R, et al. Clinical diagnosis and treatment outcomes for parapharyngeal space schwannomas: A single-institution review of 21 cases. Head Neck 2018;40(3):569–76.
[4] Budu VA, Bulescu IA, Popp CG, Mocanu BC, Mogoanta CA. Vagus nerve schwannoma in the parapharyngeal space: surgical histological and immunohistochemical aspects: a case report. Rom J Morphol Embryol 2015;56:273–6.
[5] Bao X, Kong X, Yang C, Wu H, Ma W, Wang R. Targeted next-generation sequencing of malignant peripheral nerve sheath tumor of the pterygopalatine fossa with intracranial metastatic recurrence. Medecine 2018;97(4):9636.
[6] Cavallaro G, Pattaro G, Iorio O, Avallone M, Silecchia G. A literature review on surgery for cervical vagal schwannomas. World J Surg Oncol 2015;13:130.
[7] Atallah I, Gervasoni J, Gay E, Righini CA. A rare case study of a retropharyngeal neurofibroma and a brief literature review. Eur Ann Otorhinolaryngol Head Neck Dis 2016;133:47–50.
[8] Abdeldaoui A1, Oker N, Duet M, Cunin G, Tran Ba Huy P. First bite syndrome: a little known complication of upper cervical surgery. Eur Ann Otorhinolaryngol Head Neck Dis 2013;130(3):123–9.
[9] Ramdass AA, Yao M, Natarajan S, Bakshi PK. A rare case of vagus nerve schwannoma presenting as a neck mass. Am J Case Rep 2017;18:908–11.
[10] Chiofalo MG, Longo F, Marone U, Franco R, Petrillo A, Pezzullo L. Cervical vagal schwannoma: a case report. Acta Otorhinolaryngologia Italica 2009;29(1):33–5.
[11] Plitt A, El Ahmadieh TY, Bindal S, Myers L, White J, Gluf W. Hypoglossal Schwannoma of neck: case report and review of literature. World Neurosurg 2018;110:240–3.
[12] Liu B, Zhang Y, Zhang L, Zhang F, Li H, Li S, et al. A rare case of bilateral cervical vagal neurofibromas: role of high-resolution ultrasound. BMC Neurology 2017;17:26.
[13] Erol E, Koycu A, Aydin E. A rare tumor in the cervical sympathetic trunk: Ganglioneuroblastoma. Case Rep Otolaryngol 2016:1454932.
[14] Lu D, Lui J, Chen Y, Chen F, Yang H. Primary cervical ganglioneuroblastoma: a case report. Medecine 2018;97(12):90.
[15] Yang T, Huang Y, Xu T, et al. Surgical management and outcomes of ganglioneuroblastoma-intermixed. Pediatr Surg Int 2017;33(9):955–9.
[16] Jackson JR, Tran HC, Stein JE, Shimada H, Patel AM, Marachelian A, et al. The clinical management and outcomes of cervical neuroblastic tumors. J Surg Res 2016;(204):109–13.
[17] Alvi S, Karadaghy O, Manalang M, Weatherly R. Clinical manifestations of neuroblastoma with head and neck involvement in children. Int J Pediatr Otorhinolaryngol 2017;97:157–62.
[18] Arshi A, Tajudeen BA, John MST. Malignant peripheral nerve sheat tumors of the head and neck: demographics, clinicolpathologic features, management and treatment outcomes. Oral oncology 2015;51(12):1088–94.
[19] Dalil A, Gogha VF, Vokwely JE, Sougou E, Miloundja J. A case report of cervical neurofibrosarcoma: clinical presentation, treatment and outcome. J Surg Case Rep 2018;42:175–8.
[20] Nakahira M, Saito N, Sugasawa M. Extratemporal malignant nerve sheath tumor of facial nerve with coexistent intratemporal neurofibroma mimicking malignant intemporal extension. Case Rep Otolaryngol 2015:790941.