Scholars Journal of Medical Case Reports

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

Malignant Schwannoma of the Parotid Region in a Pediatric Patient: A Case Report

G. Hadraoui[1*], S. Chaouia[1], O. Bounid[1], Y. El Khadir[1], T. Igarramen[1], R. Iharti[1], M. Darfaoui[1], A. El Omrani[1], M. Khouchani[1]

[1]Radiation Oncology Department, Hematology and Oncology hospital, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco

DOI: 10.36347/sjmcr.2021.v09i10.011 | **Received:** 01.09.2021 | **Accepted:** 05.10.2021 | **Published:** 07.10.2021

***Corresponding author:** Hadraoui Ghita

Abstract **Case Report**

Primary malignant schwannomas are uncommon neoplasms of nerve sheath origin, particularly in the head and neck, where few cases have been documented in the literature. In the work-up of a neck mass, these tumors may offer a diagnostic dilemma and carry a poor prognosis despite wide excision, chemotherapy, and radiotherapy. We report the case of a 4 years old boy who had clinical evidence of neurofibromatosis type 1 and who presented with an asymptomatic solitary left cervical swelling. He was evaluated with an MRI scan of the neck. A biopsy was performed, and the anatomopathological and histologic examination revealed a plexiform and myxoid schwannoma. The tumor was deemed unresectable by the department of oral and maxillofacial surgery. Pathological and radiological evaluation, differential diagnosis of this neoplasm and its management are discussed.

Keywords: Malignant schwannoma, head and neck, surgery, chemoradiotherapy, prognosis.

INTRODUCTION

Neurofibromatosis type I (von Recklinghausen's disease) is a hereditary illness. Clinical symptoms include cafe au lait spots, sessile or pedunculated dermal tumors, and tumors of the peripheral nerves. Neurofibromatosis is known to be complicated by malignant tumors, most of which are of soft-tissue origin. The reported incidence of transformation of neurofibromas into malignant schwannomas ranges from3 to 30%; only a small percentage of these transformations occur in the head and neck area [1].

Malignant schwannoma is a primary nerve sheath tumor that usually arises from peripheral nerves [2]. It is considered a highly aggressive tumor that carries a poor prognosis [3]. It requires a precise, early diagnosis based on imaging techniques and immunohistochemical studies to determine an appropriate line of treatment. Surgery is the recommended approach for low-grade tumors. Aggressive surgical excision with broad tumor-free margins, reinforced by postoperative irradiation and/or chemotherapy, is suggested for high-grade malignancies [3, 4]. Distant metastasis occurs by perineural or hematogenous dissemination, and the lung, liver, subcutaneous tissue, and bone are the most common sites of metastasis [5]. Otherwise, metastasis occurs in more than half of the cases [6]. As a result, chest radiography, abdomen sonography, and bone scans are required at the initial and follow-up visits [7].

In this article, we report a case of schwannoma arising from the left parotid gland presenting as a swelling in the upper neck. Clinical findings, therapeutic management and outcomes of this tumor are reviewed.

CASE REPORT

A 4 years-old boy, with a past family history of Café au lait macules observed in his paternal relatives, including the father, uncle, and grandmother, presented with a slow growing, painless swelling of the left upper lateral neck, first noted at the age of 3 months.

MRI scan of the neck demonstrated a vascular-looking mass of the left parotid gland of 56x40mm, locally infiltrating, draining into the golf course of the IJV with extension to the meninges of the temporal region, evoking an arteriovenous malformation or a paraganglioma.

A biopsy was performed, which anatomopathological and Immunohistochemistry study revealed a plexiform and myxoid schwannoma.

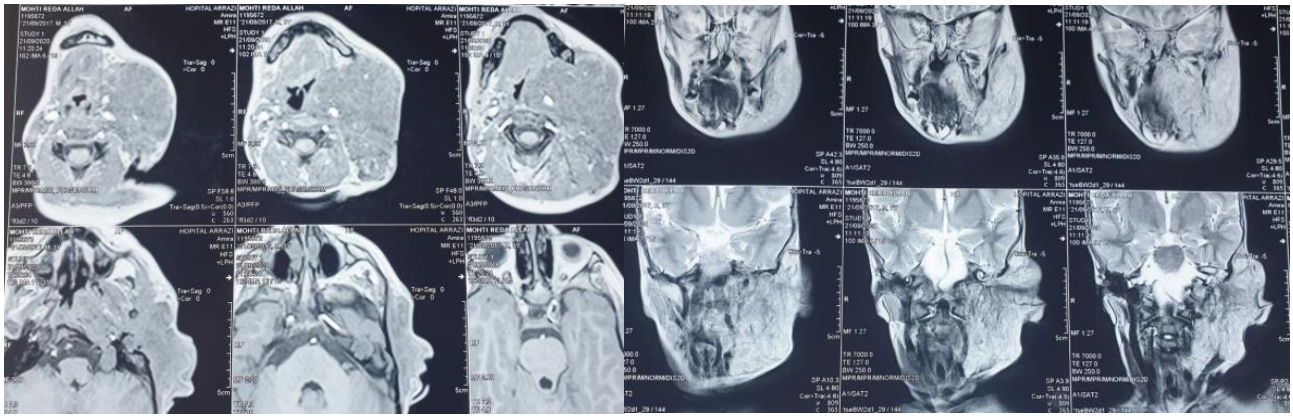
One year before admission, the mass increased in size until it became compressive, requiring a tracheostomy.

Clinical examination revealed a firm oblong mass measuring approximately 10 cm, located in the left parotid region and extended to the pretragian area. The mass was painless, non-pulsatile, non-expansive and movable. The overlying skin was normal and there were no palpable neck nodes.

The patient had no difficulty in chewing, swallowing, and phonation. The remaining physical examination was unremarkable, except for the café-au-lait spots on the abdomen and the back.

A second MRI scan revealed:

- A left latero-cervical mass of 8.7x8.9x10.4cm, encompassing the parotid region and the homolateral pterygoid muscles. At the top, it extends to the temporal fossa, filling the external auditory canal and obstructing the JV causing extensive thrombosis.
- Anteriorly, it invades the jawbone and the homolateral mandibular bone.
- Medially, it extends to the homolatearal latero-pharyngeal spaces, invades the wall of the nasopharynx at the top, at the bottom it extends to the oropharynx coming into contact with the base of the tongue. This mass pushes back and completely obstructs the laryngeal lumen.
- Posteriorly, it enlarges the homolateral internal auditory canal and the torn foramen, extending to the ponto-cerebellar angle. Below, it extends to the homolateral subangulo-mandibular region.



The tumor was deemed unresectable by the department of oral and maxillofacial surgery.

The patient was given anticoagulant therapy for the thrombosis and was referred to our department for chemotherapy.

DISCUSSION

Malignant schwannoma, also known as neurogenic sarcoma, neurofibrosarcoma, malignant neurofibroma, and recently renamed malignant peripheral nerve sheath tumor is a rare form of cancer that accounts for 5% of all soft-tissue tumors and affects 0.001% of the population [8, 9].

Malignant schwannoma of the head and neck is quite rare [6]. The limbs are the main location of this entity, then followed by the trunk [6]. Cashen *et al.*, reported largest numbers of tumors were seen in the thigh, shoulder, and pelvis, but the tumors also occurred in more central (abdomen and thorax) and distal (forearm, hand, and foot) locations [10].

The head and neck region is occasionally involved, with less than 10% of the cases that affect cranial nerves, the brachial plexus or the sympathetic chain [11, 12].

MS shows a peak of incidence in the fifth decade, although the age of presentation in Von Recklinghausen´s disease is lower than in the general population [12, 13]. Males and females are equally affected by MS and no racial predisposition has been identified [11, 12].

The often observed link between malignant schwannomas and von Recklinghausen's disease is of particular relevance. According to Hosoi, 13% of all individuals with von Recklinghausen's disease develop malignant transformation of their neurofibromas [14]. According to this author, such tumors were characterized by frequent local recurrences and late metastatic dissemination. Although our patient demonstrated no evidence of multiple neurofibromatosis, as seen in classical von Recklinghausen's disease, he did have several light brown spots on his trunk. Jackson has described such lesions as a “forme frust” of neurofibromatosis and has indicated that they may represent the only visible stigmata in some patients with this disease [15, 16].

Signs and symptoms presentation may also vary depending upon the anatomical location of the tumors. Most of the patients present with painless swelling. However, if present, symptoms usually depend upon the site of the tumor. Other than pain, a tingling sensation and numbness along the course of the involved nerve are also reported. Other symptoms could be breathing difficulty, especially when the tumor is located in the nose, painful swallowing in case of a tumor occuring in the pharynx, nose bleeding due to the presence of a tumor in the paranasal sinuses, breaking of voice when a tumor is located in the larynx, or just a swelling in the neck in case of a tumor in the parapharyngeal space [17]. In most cases, the swelling is freely mobile. However, attachment to nerves might lead to limited mobility of the tumor. In some cases, the nerves from which the tumor has originated could beencompassed entirely in the Schwannomas [18].

Confirmation of the diagnosis of Schwannomas located in the head and neck region before surgery is rather difficult. Commonly performed investigations include CT, MRI, and FNAC [19]. Contrast-enhanced CT is the best initial diagnostic study to determine the size and extent of the tumor, demonstrate the degree of tumor vascularity, and help the surgical approach [20]. The mass appears hypodense with some degree of enhancement, while paraganglioma is classically isodense [21].

There is a general agreement concerning the great value of MRI in the pre-operative work-up. It helps define diagnosis and evaluate the extent and the relationship of the tumour with the jugular vein and the carotid artery [22]. The schwannoma is generally hypointense on T1 and hyperintense on T2-weighted images, depending on its cellularity [21, 23]. A normal ganglion is usually hypointense on T2, a finding that may help distinguish it from a schwannoma or enlarged retropharyngeal node [24].

If the situation allows, diagnosis can be confirmed by FNAC: it helps obtain representative cells from the 2 types of tissues seen histologically in schwannomas. Type A tissue consists of compactly arranged spindle cells with long, oval nuclei, rounded ends, and long axes arranged parallel to each other, resulting in a palisade appearance. This type of tissue shows the Verocay bodies, formed of parallel bundles of fibers and cells with nuclei polarized at each end [25]. Type B tissue, formed of spindle and stellate cells arranged in a loose connective tissue matrix, is represented cytologically by individual, spindle-shaped neoplastic cells. In larger and older Schwannomas, degenerative changes, vascular sclerosis, and hemorrhagic changes are common. Sometimes, the formation of a microcyst is evident with a pseudoepithelial lining of the Schwann cells [26].

However, it is not always possible in many cases to achieve a definitive diagnosis until the tumor has been excised. Electron microscopy and immunohistochemical analysis are often necessary to diagnose and accurately classify nerve sheath tumors [27].

There is no controversy as to the best treatment for the benign schwannoma. However, there is no agreed-upon treatment regimen for malignant schwannomas [28]. It is an aggressive disease with a high recurrence rate and a low survival rate [7]. Although the rarity of lymph node metastases in association with malignant schwannoma has been highlighted several times [5], the aggressive nature of the tumor results in a high frequency of local recurrence. Radial local excision is the most efficient therapy. Wide excision should include involved neurovascular structures, subcutaneous tissue, underlying bone, and adjacent uninvolved musculofascial planes. Because the tumor has a predilection to develop along the nerve trunk, excision should be accompanied by neurectomy with nerve avulsion [6]. If incompletely excised, the tumor tends to recur locally within a relatively short period. Adjuvant therapy should be considered, especially for those with malignant schwannoma associated with von Recklinghausen's disease or at the site of prior irradiation. Pandey *et al*., [29] advocated for the use of adjuvant radiation and/or chemotherapy in conjunction with wide excision to treat head and neck sarcoma. According to Steins *et al.*, [30], malignant schwannoma is a relatively rare tumor entity that is typically treated as a member of the group of soft tissue sarcomas. They used ifosfamide and doxorubicin as the first-line chemotherapy for malignant schwannoma. If the tumor was refractory to the first-line therapy, treatment with carboplatin in combination with etoposide was tried.

Chemotherapy has been recommended for patients with unresectable recurrent tumors or distant metastases [11].

In cases where the surrounding structures have been invaded, and the tumor cannot be enucleated, radiotherapy is considered a useful palliative modality. However, it is well known that malignant schwannoma is usually resistant to both radiotherapy and chemotherapy, though chemotherapy may be used to treat surgical failures [1].

The prognosis of MS has generally been described as poor to dismal, particularly in the head and neck location and in patients with associated Von Recklinghausen's disease [8, 13]. This prognosis depends on several histological factors, such as tumor grade, degree of cellular pleomorphism, mitotic activity, and size of the primary tumor [12]. The 5-year survival rates for patients with neurofibromatosis are 15–20% and 50–55% for patients without this disease,

with a high rate of local recurrence and distant metastases in Von Recklinghausen's disease [8].

CONCLUSION

Malignant schwannoma is considered as an aggressive disease with a low survival rate. Diagnosis should be based not only on a thorough clinical examination but also on pathological findings. Radical surgery and adjuvant therapy are the best treatments for individuals with malignant schwannoma, especially those with von Recklinghausen's disease.

Chest radiography, abdominal sonography, and bone scans are required for tumor staging and for detecting distant metastases following therapy. Because of the high recurrence rate, meticulous physical and radiological examinations, such as computed tomography or magnetic resonance imaging, are required for the identification of recurring tumors. If a suspect recurrent lesion is noted, a repeat biopsy should be performed.

REFERENCES

1. Gallo, A., Suriano, M., Simonelli, M., Ralli, G., & De Vincentiis, M. (2003). Recurrent malignant schwannoma of the parapharyngeal space in neurofibromatosis type 1. *Ear, nose & throat journal*, *82*(11), 862-865.
2. Pabiszczak, M., Woźniak, A., Wierzbicka, M., Leszczyńska, M., & Szyfter, W. (2004). Diagnostic difficulties in the laryngeal malignant peripheral nerve sheath tumor (MPNST). *Otolaryngologia polska= The Polish otolaryngology*, *58*(6), 1133-1136.
3. Bailet, J. W., Abemayor, E., Andrews, J. C., Rowland, J. P., Fu, Y. S., & Dawson, D. E. (1991). Malignant nerve sheath tumors of the head and neck: a combined experience from two university hospitals. *The Laryngoscope*, *101*(10), 1044-1049.
4. Ducatman, B. S., & Scheithauer, B. W. (1984). Malignant peripheral nerve sheath tumors with divergent differentiation. *Cancer*, *54*(6), 1049-1057.
5. Greager, J. A., Reichard, K. W., Campana, J. P., & DasGupta, T. K. (1992). Malignant schwannoma of the head and neck. *The American journal of surgery*, *163*(4), 440-442.
6. Henkel, K. O., Putzke, H. P., Saka, B., & Gundlach, K. K. (1998). Malignant epithelioid schwannoma of the inferior alveolar nerve. Case report with review of the literature. *Mund-, Kiefer- und Gesichtschirurgie: MKG*, *2*(3), 163-166.
7. Chao, J. C., Ho, H. C., Huang, C. C., & Tzeng, J. E. (2007). Malignant schwannoma of the mandible: a case report. *Auris Nasus Larynx*, *34*(2), 287-291.
8. Ducatman, B. S., Scheithauer, B. W., Piepgras, D. G., Reiman, H. M., & Ilstrup, D. M. (1986). Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. *Cancer*, *57*(10), 2006-2021.
9. Wanebo, J. E., Malik, J. M., Vandenberg, S. R., Wanebo, H. J., Driesen, N., & Persing, J. A. (1993). Malignant peripheral nerve sheath tumors. A clinicopathologic study of 28 cases. *Cancer*, *71*(4), 1247-1253.
10. Cashen, D. V., Parisien, R. C., Raskin, K., Hornicek, F. J., Gebhardt, M. C., & Mankin, H. J. (2004). Survival data for patients with malignant schwannoma. *Clinical Orthopaedics and Related Research®*, *426*, 69-73.
11. Mannan, A. R., Singh, M. K., Bahadur, S., Hatimota, P., & Sharma, M. C. (2003). Solitary malignant schwannoma of the nasal cavity and paranasal sinuses: report of two rare cases. *Ear, nose & throat journal*, *82*(8), 634-640.
12. Capote, A., Escorial, V., Reina, T., Munoz-Guerra, M. F., Nieto, S., & Naval, L. (2006). Primary malignant schwannoma of the cervical plexus with melanocytic differentiation. *International journal of oral and maxillofacial surgery*, *35*(8), 767-771.
13. Hoffmann, D. F., Everts, E. C., Smith, J. D., Kyriakopoulos, D. D., & Kessler, S. (1988). Malignant nerve sheath tumors of the head and neck. *Otolaryngology—Head and Neck surgery*, *99*(3), 309-314.
14. Hosoi, K. (1931). Multiple neurofibromatosis (von Recklinghausen's disease): with special reference to malignant transformation. *Archives of Surgery*, *22*(2), 258-281.
15. Jackson, A. H. (1933). Three cases of multiple neurofibromatosis with malignant degeneration. *The Journal of Nervous and Mental Disease*, *78*(6), 581-596.
16. Moazam, F., Rogers, B. M., & Talbert, J. L. (1983). Retroperitoneal malignant schwannoma: a case report. *Journal of pediatric surgery*, *18*(2), 189-192.
17. Valentino, J., Boggess, M. A., Ellis, J. L., Hester, T. O., & Jones, R. O. (1998). Expected neurologic outcomes for surgical treatment of cervical neurilemomas. *The Laryngoscope*, *108*(7), 1009-1013.
18. Das, J., & Saha, J. (2016). Cervical Schwannoma-A Case Report. *Otolaryngology*, [Internet]. [cité 22 août 2021];06(2). Disponible sur: https://www.omicsonline.org/open-access/cervical-schwannoma-a-case-report-2161-119X-1000229.php?aid=71730
19. Callum, F. (2021). Scott-Brown's Otorhinolaryngology, Head and Neck Surgery, 7th edn. [Internet]. [cité 5 sept 2021]. Disponible sur: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3604940/
20. Awasthi, S. K., & Dutta, A. (2011). Cervical sympathetic chain schwannoma: a case report. *Indian Journal of Otolaryngology and Head & Neck Surgery*, *63*(3), 292-294.
21. Bocciolini, C., Dall'Olio, D., Cavazza, S., & Laudadio, P. (2005). Schwannoma of cervical

sympathetic chain: assessment and management. *Acta Otorhinolaryngologica Italica*, *25*(3), 191-194.
22. Chiofalo, M. G., Longo, F., Marone, U., Franco, R., Petrillo, A., & Pezzullo, L. (2009). Cervical vagal schwannoma. A case report. *Acta Otorhinolaryngologica Italica*, *29*(1), 33-35.
23. Iacconi, P., Faggioni, M., De Bartolomeis, C., Iacconi, C., & Caldarelli, C. (2012). Cervical sympathetic chain schwannoma: a case report. *ACTA otorhinolaryngologica italica*, *32*(2), 133-136.
24. Anil, G., & Tan, T. Y. (2010). Imaging characteristics of schwannoma of the cervical sympathetic chain: a review of 12 cases. *American Journal of Neuroradiology*, *31*(8), 1408-1412.
25. Ramzy, I. (1977). Benign schwannoma: demonstration of Verocay bodies using fine needle aspiration. *Acta cytologica*, *21*(2), 316-319.
26. Mohammed, M. E., Khan, A., & Asiri, M. (2019). Supraclavicular cervical schwannoma: a case report. *Cureus*, *11*(10). [Internet]. 16 oct 2019 [cité 5 sept 2021]; Disponible sur: https://www.cureus.com/articles/24026-supraclavicular-cervical-schwannoma-a-case-report
27. Donnelly, M. J., Al-Sader, M. H., & Blaney, A. W. (2021). View from. Benign nasal schwannoma. PubMed [Internet]. [cité 5 sept 2021]. Disponible sur: https://pubmed.ncbi.nlm.nih.gov/1479267/
28. Colreavy, M. P., Lacy, P. D., Hughes, J., Bouchier-Hayes, D., Brennan, P., O'dwyer, A. J., ... & Walsh, M. A. (2000). Head and neck schwannomas–a 10 year review. *The Journal of Laryngology & Otology*, *114*(2), 119-124.
29. Pandey, M., Thomas, G., Mathew, A., Abraham, E. K., Somanathan, T., Ramadas, K., ... & Nair, M. K. (2000). Sarcoma of the oral and maxillofacial soft tissue in adults. *European journal of surgical oncology*, *26*(2), 145-148.
30. Steins, M. B., Serve, H., Zuhlsdorf, M., Senninger, N., Semik, M., & Berdel, W. E. (2002). Carboplatin/etoposide induces remission of metastasised malignant peripheral nerve tumours (malignant schwannoma) refractory to first-line therapy. *Oncology reports*, *9*(3), 627-630.