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Malignant triton tumour of the sinonasal tract: Case report and literature review☆

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ABSTRACT

INTRODUCTION: The objective is to report a rare tumour of the sinonasal tract and conduct a literature review.

Malignant triton tumour is a subtype of malignant schwannoma with rhabdomyoblastic differentiation. It is a very rare tumour, with only 15 reported cases involving the sinonasal region.

PRESENTATION OF CASE: Forty-seven years old female presented with a right-sided epistaxis, progressive right sided nasal obstruction and anosmia and a visible mass in the right nasal cavity. Imaging studies showed a mass extending from the piriform aperture to the nasopharynx in contact with the dura and the orbital content. The mass was biopsied and the result was consistent with malignant triton tumour. The patient refused the surgery at first so chemotherapy with MAID protocol was started. After the fourth course of chemotherapy the treatment was stopped due to patient intolerance and a thrombosis of the jugular vein. Patient then underwent surgery with frontal craniotomy and dural excision, endoscopic control was done at the end to insure a complete removal. The patient received Radiotherapy in the postoperative period (56 Greys). At 5 years of follow up the patient is doing fine with no signs of recurrence and normal ophthalmological findings.

DISCUSSION: Sixteen cases, including our case, have been reported to date in the literature. The mean age at presentation is 61 years. None of cases were associated with neurofibromatosis type 1. Eight patients were reported to be alive 5 years post-treatment, and 2 patients were reported to have died of the disease. The prognosis for triton tumours in the sinonasal tract is better than that for triton tumours in other locations.

CONCLUSION: Malignant triton tumour is a rare malignancy of the sinonasal tract. Otolaryngologists should be aware of this disease. The optimal treatment should include radical resection of the tumour.

1. Introduction

Malignant triton tumour is a subtype of malignant schwannoma with rhabdomyoblastic differentiation.[1] To date, approximately 150 cases have been reported in the literature.[2] Masson, in 1932, was the first to describe a malignant schwannoma with rhabdomyoblastic differentiation in a patient with neurofibromatosis type 1(NF1).[3]

Malignant triton tumour is a very rare tumour that occurs in head and neck region and the trunk, with almost one third of the cases in the head and neck area.[4] It can be associated with neurofibromatosis type one (NF1) (in patients younger than 35-years old) or it can occur sporadically (in older patients). The patients with NF 1 have a poorer prognosis (Table 1).[2]

To the best of our knowledge, the sinonasal localisation was reported in only 15 cases in the English and French literature.[2,4–7] The aim of this article is to present a case with a rare malignancy of the sinonasal tract and to describe a literature review of malignant triton tumours in this specific anatomical location.

2. Case report

A 49-year-old women presented to the emergency department in April 2005 with right-side epistaxis and a 2-year history of

☆ This review was presented as an oral presentation at the 6th International Congress of the World Federation of Skull Base Societies and the 10th European Skull Base Society Congress 2012 that occurred at the Brighton Hilton Metropole from 16–19 May 2012.

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Table 1
Review of the literature on malignant triton tumours of the sinonasal tract.

Patients	Age/sex	NF 1	Localisation	Treatment	Radio- or chemotherapy	Retreatment	Outcome
Shajrawi et al. [14]	75/M	No	Left paranasal sinuses	Debulking	RT	Surgical removal of recurrence at 36 months	Alive NED at 6 months
Bhatt et al. [11]	66/F	No	Left nasal cavity	Extended external ethmoidectomy			Alive with tumour at 27 months
Heffner and Gnepp [13]	64/M	No	Left nasal cavity	Incomplete excision			Died of tumour at 27 months, pulmonary metastasis and cranial extension
	58/M	No	Nasal cavity	Surgery	RT		Alive NED at 48 months
	56/F	No	Left nasal cavity	Local excision		Removal of recurrence at 11 and 15 years	Alive NED at 7 years
	59/M	No	Right nasal cavity	Transpalatal excision	RT		Alive NED at 7 years
	43/M	No	Right nasal cavity	N/A		Recurrence at 15 years, treatment N/A	Alive NED at 7 years
Nicolai et al. [5]	81/F	No	Right nasal cavity	Endoscopic excision			Alive NED at 36 months
Kim et al. [4]	38/F	No	Right nasal cavity	Medial maxilectomy	RT		Alive NED at 5 years
Tringali et al. [6]	80/F	N/A	Right nasal cavity	Extended midfacial resection		Recurrence at 8 months with cerebral involvement, midfacial and sub frontal excision and RT. 3 endoscopic partial resections in 5 years	Alive with disease at 5 years
Xue et al. [7]	47/F	N/A	Right paranasal sinuses	Lateral rhinotomy	RT		Alive NED at 5 years
Terzic et al. [2]	35/F	no	Right nasal cavity	Craniotomy with sub-cranial resection			Alive NED at 30 months
	77/M	No	Left nasal cavity	Partial endoscopic resection		RT for lymph node metastasis at 6 months	Alive with tumour at 7.5 months
	73/F	No	Right maxillary sinus	Partial endoscopic resection		Endoscopic resection with RT for recurrence at 18 months	Died of other causes at 5.5 years
	76/M	no	Left sinonasal CAVITY	Refused treatment			Died of tumour at 1.5 months
Present case	49/F	N/A	Right nasal cavity	Craniotomy with sub-cranial resection	RT		Alive NED at 5 years

N/A: not available or not specified, NED: no evidence of disease, RT: radiotherapy.

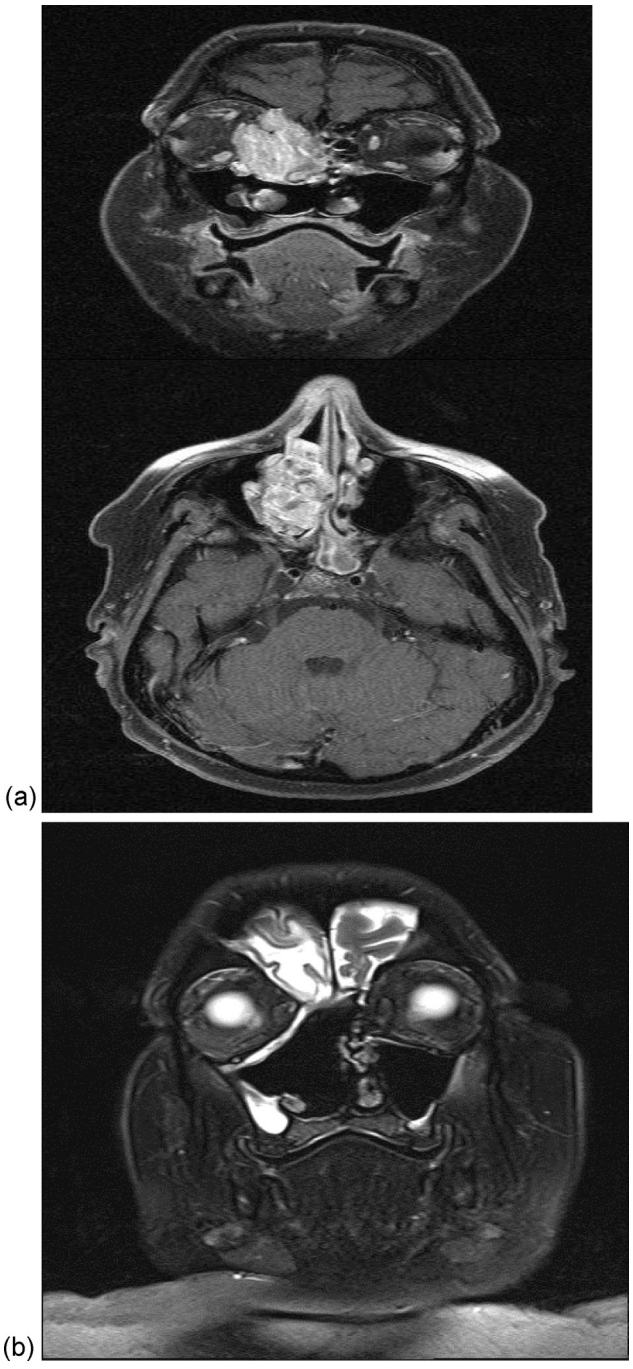

Fig. 1. (a) Preoperative MRI (T1 sequence with gadolinium injection) showing the tumour in the right nasal fossa with intracranial extension with mass effect on the orbit. (b) Post operative MRI (T2 sequence with gadolinium injection) at 5 years showing no evidence of tumour recurrence.

progressive right-side nasal obstruction and anosmia, and a visible mass in the right nasal cavity. The rest of the history and physical examination revealed a history of meningitis and respiratory tuberculosis in infancy and a caesarean section. She had normal neurological and ophthalmological exams and no cutaneous lesions or lymph node enlargement.

CT and MRI scans were conducted, which showed a lesion occupying the right nasal fossa with maxillary sinus involvement that extended from the piriform aperture to the cavum and was in contact with the orbital contents (absence of the lamina papyracea) and the dura in the region of the cribriform plate (Fig. 1).

Because several different types of malignancy were possible (rhabdomyosarcoma, neuro-endocrine tumour, neuroblastoma, and chordoma), a biopsy was performed under local anaesthesia during her hospitalisation; however, the histological findings were inconclusive. Therefore, another biopsy was performed under general anaesthesia, and the biopsy sample was sent to the central histopathology department in France (Hospital Lariboisere).

The microscopic examination showed a very cellular tumour containing spindle-shaped cells with oval and comma shaped nuclei. Eosinophilic cytoplasm was also observed. This aspect was associated with well-differentiated muscle fascicles with a very low mitotic index (1 mitosis per 10 high-power fields).

Immunohistochemistry showed that the tumour cells were immuno-stained for S-100 protein and vimentin filaments, suggesting a Schwann cell origin. Therefore, the diagnosis of a nerve sheath tumour with rhabdomyoblastic differentiation was reached. Testing for NF 1 was not performed because the patient was more than 60 years old and did not have any other features of the disease.

The patient refused surgery at that time, so she began chemotherapy using the MAID protocol (Mesna, doxorubicin, ifosfamide, and dacarbazine); however, after the fourth course, the treatment was stopped because of the patient's lack of tolerance and because a thrombosis of the jugular vein over the central line had developed (Fig. 2).

The patient again refused the surgical option. However, three months later, the patient presented to the hospital with right exophthalmia and new MRI and CT scans were performed. The tumour was observed to be in contact with the orbital contents and the frontal dura with no evidence of metastasis or cerebral invasion.

The oncological multidisciplinary team proposed surgical treatment with neurosurgical collaboration, followed by three-dimensional radiotherapy.

In September 2006, the surgery was performed via a frontal craniotomy. The tumour was removed along with the septum, lamina papyracea, and middle turbinate, and dural excision and duraplasty was performed by the neurosurgical team. An endoscopic nasal evaluation was conducted at the end of the procedure to ensure complete tumour removal. The patient was discharged on day 7 with no complications (no dural invasion was observed on histopathology).

Subsequently, radiotherapy was conducted on the tumour site only (56 Greys).

The patient is alive at 5 years post-treatment with no signs of recurrence and normal ophthalmological findings.

3. Results

Of the 16 cases reported to date in the literature, the mean age at presentation is 61 years (range: 38–81) with a slight female predominance 1.3:1(9:7) and a right-side predominance 1.6:1(10:6).

Eight of the 16 patients were reported to be alive at 5 years post-treatment, 7 without evidence of disease and one with disease progression. Two patients were reported to have died of the disease, one with pulmonary metastasis and cranial extension and one patient who refused all treatment. One patient was reported to have neck lymph-node metastases.

Seven patients (43%) had recurrence, of which five had incomplete excision or non-radical excision. In the other 2 cases, the surgical details were not specified. Two patients were treated with surgery and radiotherapy (one underwent three subsequent endoscopic partial removals), and one patient was treated with radiotherapy alone for lymph-node metastases. Two of the patients were retreated with surgery alone, with one of these two patients undergoing 2 surgeries. In one patient, the retreatment

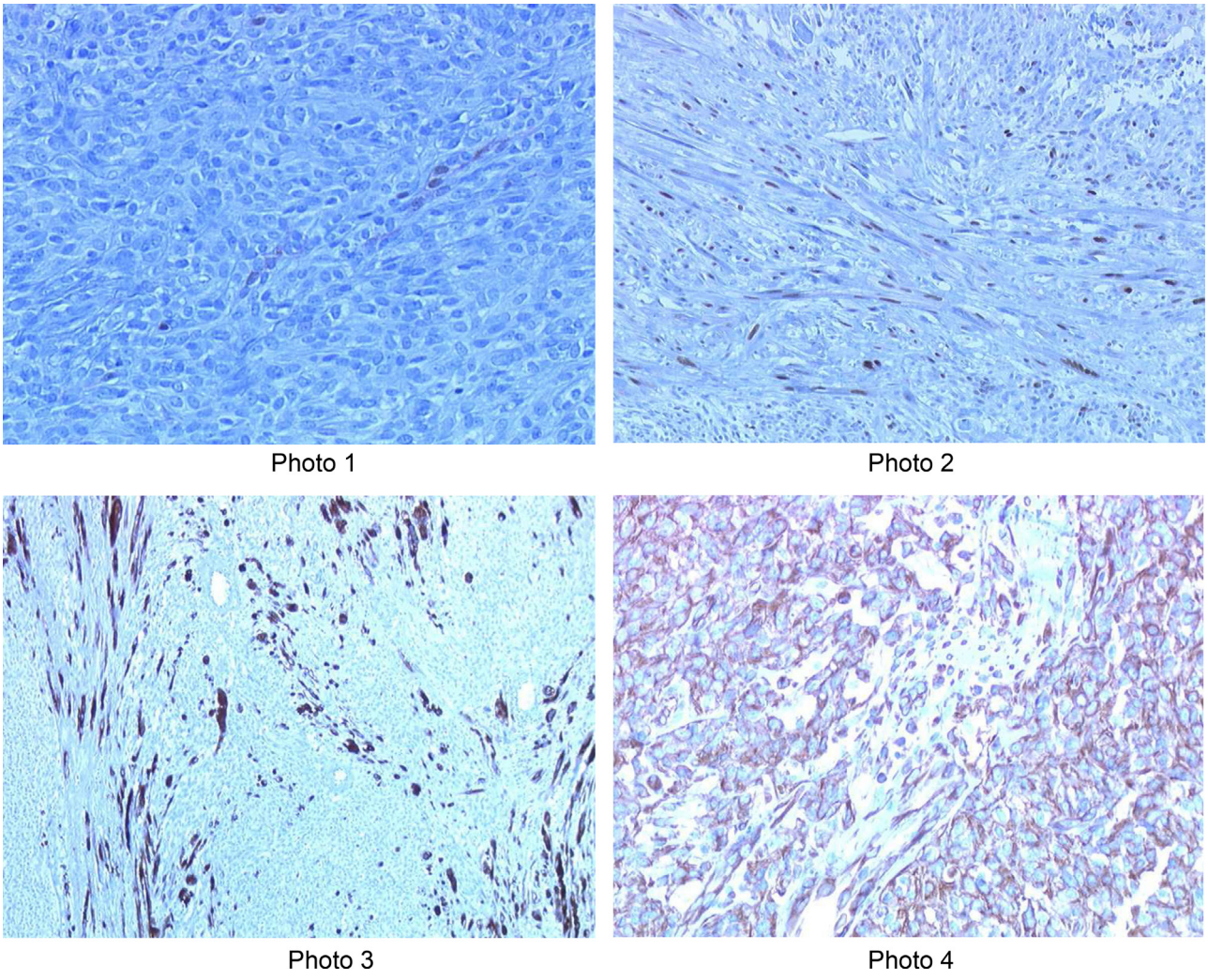

Fig. 2. Photo 1: immunohistochemical analysis showing focal positive Ps 100 staining (indicating a nerve sheet origin) with scattered spindle cells (IHC ×40). Photo 2: immunohistochemical analysis showing strong focal positive myogenin staining (indicating a striated muscle cells IHC ×25). Photo 3: immunohistochemical analysis showing strong positive desmine staining (indicating a striated muscle cells IHC ×10). Photo 4: immunohistochemical analysis showing strong positive vimentin staining (indicating a mesenchymal cells IHC ×40).

was not specified, and another patient underwent no retreatment and subsequently died from pulmonary metastases and cerebral involvement.

The overall survival of patients with malignant sinonasal triton tumour at 5 years is 50%.

Statistical analysis was limited by the small sample size and the limited duration of follow up.

4. Discussion

Malignant schwannoma with rhabdomyoblastic differentiation is a rare malignancy that is referred to as malignant triton tumour. The term, malignant triton tumour, was first used by Woodruff to describe such lesions,[1] and the name refers to Locatelli's experiment in 1925 in which a supranumerary limb growth with muscles and bone occurred in the triton salamander after implanting a sciatic nerve on it's back.[8]

To date, only approximately 150 cases have been reported, with 53 arising in the head and neck area[2] and very few in the sinonasal area. However, it appears that this figure is an underestimation of the actual number due to the unclear description of the cases or labelling the tumour as a malignant schwannoma. The pathologist plays an important role in establishing the diagnosis, and it can be difficult in some cases, such as the one that presented here, because our pathologist could not reach a specific diagnosis. The specimen was sent to the central histopathology department of the country where an experienced pathologist made the diagnosis. There were 2 months between the first biopsy and the diagnosis, which is a long time for such a malignancy.

The original histological criteria for establishing the diagnosis of the malignant triton tumour was proposed by Woodruff in 1973[1]; the criteria require that the tumour

1. Arise along the course of a peripheral nerve in a patient with NF-1.
2. Display most of the growth characteristics of Schwann cells.
3. Contain bona fide rhabdomyoblasts.

Various rates of association between malignant triton tumours and NF-1 have been reported in the literature (23–69%)[1,2,4,9–11]: the largest series of head and neck localised malignant triton tumours was reported by Terzic in 2009[4] and included 53 patients of which only 23% were found to be NF-1 positive. In the current review of all of the patients with sinonasal tract localisation, none had

NF-1, which is the same result reported by Nicolai.[5] Although in our case, the NF-1 test was not performed because there were no clinical features suggestive of NF-1 and the patient's was above 35 years old. In another two cases, the NF-1 status was not specified, bringing the total cases of malignant triton tumours localised in the sinonasal tract that were confirmed NF-1 negative to 13 of the 16 cases.

In the 16 cases of sinonasal tumour localisation, the mean age is 61 (38–81), which is slightly older than the age reported for patients with head and neck tumour localisation (40 years) or patients with NF-1 (26 years).[2]

Malignant schwannoma with rhabdomyoplastic differentiation is much more aggressive than sporadic schwannoma,[10] with an overall 5-year survival rate of 12%[10] for patients with malignant schwannoma tumours in all locations and approximately 26%[12] for patients with the tumours in the head and neck area. However, Terzic recently reported a 5-year survival rate of 49% for patients with malignant triton tumours in the head and neck[2] Tumours localised to the sinonasal tract seem to have a more favourable prognosis than tumours localised elsewhere.[2,5,13,14] Our result is consistent with Terzic's report because we observed an overall 5-year survival rate of 50%. This results might be due to the lower grade histology of the sinonasal tract tumours compared to that of tumours localised elsewhere, as in our case and as reported by Bhatt[11] and Nicolai.[5]

There is no consensus regarding the treatment guidelines for malignant triton tumours. Various recommendations have been proposed by several authors, including radical excision[15,16] radical excision followed by radiotherapy and chemotherapy[10,17–20] or excision and radiotherapy[4,5,12]. However, as for any other sarcoma, resection of the tumour with wide margins followed by radiotherapy is the recommended treatment, whereas the need for chemotherapy has not been clearly defined.[12]

5. Conclusion

We presented the sixteenth case of a malignant triton tumour of the sinonasal tract. This is a rare tumour that has a different profile than malignant triton tumours localised elsewhere because it presents in older patients, is not associated with NF1, and has a low histological grade in general. However, intracranial extension and metastasis of malignant triton tumours have been reported. Radical excision with wide margins followed by radiotherapy is the recommended treatment.

Conflict of interest

The authors have no conflicts of interest to declare for this article.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal on request.

Author contributions

Abdulmajeed Zakzouk and Fahad Hammad were involved in data collection, data analysis and writing the paper. Moutaz Aziz was involved in data collection "histology". Olivier Langlois and Jean-paul Marie were involved in data analysis. Olivier Choussy was involved in data analysis and writing the paper.

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