CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 608–612

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

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

Abdulmajeed Zakzouk[a,*], Fahad Hammad[b], Olivier Langlois[b], Moutaz Aziz[c], Jean-Paul Marie[a], Olivier Choussy[a]

[a] Department of Otolaryngology, University Hospital of Rouen, France
[b] Department of Neurosurgery, University Hospital of Rouen, France
[c] Department of Histo-Pathology, University Hospital of Rouen, France

ARTICLE INFO

Article history:
Received 27 September 2013
Accepted 15 July 2014
Available online 23 July 2014

Keywords:
Malignant triton tumour
Schwannoma
Rhabdomyoblastic differentiation
Peripheral nerve sheet tumour
Nasal tumours

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.

* Corresponding author at: ORL Adults, Premier etage, Deve 2 CHU de ROUEN, France. Tel.: +33 6 09 71 58 07.
E-mail addresses: zakzouk.abdulmajeed@gmail.com, majeedsz@hotmail.com (A. Zakzouk).

http://dx.doi.org/10.1016/j.ijscr.2014.07.014

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.