Leiomyosarcoma of the inferior nasal concha: a case report and literature review

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Abstract. Leiomyosarcoma of the inferior nasal concha: a case report and literature review. Leiomyosarcomas (LMSs) of the sinonasal tracts are rarely reported. We present a case of an LMS of the left inferior nasal concha, and discuss the management options with review of the literature. A 72-year-old female patient presented with epistaxis. Clinical examination and medical imaging showed a tumour arising from the left inferior nasal concha. An endoscopic resection was performed. Anatomopathological and immunohistochemical analyses revealed the tumour to be a grade 3 LMS with uncertain posterior margins. The patient was a Jehovah’s Witness and refused more radical surgical resection due to religious beliefs; therefore, adjuvant conformal radiotherapy (60 Gy) was performed. LMS of the nasal cavity is a rare and locally aggressive tumour with a high tendency of recurrence, requiring radical surgical resection and long-term follow-up. The prognosis of a nasal cavity LMS is better than that of an LMS located in the paranasal sinuses.

Introduction

Sarcomas are rare tumours, representing less than 1% of newly diagnosed cancers in the United States.1 Among sarcomas, 76% develop from soft tissues,2 and between 3-7% of these soft tissue sarcomas (STS) are leiomyosarcomas (LMSs), which develop from smooth muscle cells and are frequently found at the level of the myometrium, in the gastro-intestinal tract, in the soft tissues of the extremities, and in the retroperitoneum.3 LMSs rarely affect the head and neck because of the low abundance of smooth muscles in this region; when present in this region, they are mainly concentrated within blood vessels walls and at the erector pili muscles of the skin, the oesophagus, and the posterior wall of the trachea. LMS has a poor prognosis, especially when localized at the level of the head or neck, due to a high rate of local recurrence, as well as the difficulty of large excision at this level.

To date, slightly over 40 cases of sinonasal LMS have been described in the literature. In the majority of these cases, head and neck LMSs were diagnosed in patients in their fifties, with no noted influence of sex or ethnicity.4 The most common symptoms were unilateral nasal obstruction and epistaxis. In this paper, we present the case of a patient presenting a LMS of the inferior nasal concha. We discuss diagnosis, treatment options, and prognosis.

Case report

A 72-year-old, Italian, female patient, who was a Jehovah’s witness, presented herself for emergency treatment of epistaxis from the left nasal cavity, at the ENT service at the Charleroi Civil University Hospital in mid-august of 2010. The patient had twice previously required packing of the left nasal cavity, and had several medico-surgical antecedents – including asthma, left trigeminal neuralgia, Crohn’s disease, and nasal polypectomy in 1980. Clinical examination showed a tumour arising from the left inferior nasal concha, and a synechia was apparent between the middle concha and the septum. Computed tomography (CT) of the sinus confirmed the presence of a soft tissue mass occluding the left nasal cavity, and extending into the posterior part of the

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choana (Figure 1). The soft tissue mass reached the left ethmoid and the highest part of the lateral nasal wall of the left maxillary sinus. A thickening of the mucosa at this level was also noted. Magnetic resonance imaging (MRI) confirmed the presence of a solid mass that was hyposignal on T2 sequence, and measured 16 × 19 × 40 mm (Figure 2). The mass appeared to arise from one of the concha, without apparent involvement of maxillary or ethmoid sinuses.

Because of her religious beliefs, the patient refused any surgical intervention. She accepted an endonasal resection under endoscopic control, under the condition of receiving no blood transfusion. Intraoperative microscopic analysis revealed a malignant sarcomatoid tumour with partial necrosis. A complete macroscopic excision of the tumour was then performed under endoscopic control. The inferior nasal concha was entirely excised, with a large sub-periosteal detachment of the mucosa of the inter-sinonasal septum to the sphenopalatine foramen and the anterior wall of the sphenoid (Figure 3). There was no bone erosion or sinus involvement.

Anatomopathological examination confirmed the nature of the lesion, which was characterized by a proliferation of interlaced bundles of smooth muscle fibres with elongated spindle cells (Figure 4). Cellular nuclei were of varying sizes and shapes, were hyperchromatic, and exhibited prominent mitotic activity, with more than 20 mitoses/10 high power field (microscopic field at a magnification of 200×). The tumour was of histological grade 3 following the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) classification (scores of 3 for tumoural differentiation, 3 for mitotic index, and 1 for tumoural necrosis). Immunohistochemical analysis of a paraffin section showed positive reactions for α-smooth muscle actin (1A4-Dako), desmin (D33-Dako), and vimentin (V9-Dako); results were negative for S100 protein (polyclonal-Dako), CD31 (JC70A-Dako), CD68 (PG-M1-Dako), and cytokeratin AE1/AE3 (AE1/AE3-Dako). These findings confirmed the diagnosis of LMS.

The locoregional and distant oncological assessments by cervical, thoracic, and abdominal CT, and positron emission tomography (PET) were negative. Therefore, the final diagnosis was a grade 3 LMS of the left inferior nasal concha occluding the entire nasal cavity. The endoscopic resection revealed posterior margins that were positive under anatomopathological examination, but not visible upon macroscopic inspection. Again, due to religious beliefs, the patient refused any further, more invasive intervention, as well as any blood transfusion. Therefore, a supplementary resection could not be performed, since it would have involved
injuring the Eustache tube and exposing the patient to additional haemorrhagic risk at the level of the sphenopalatine artery. Following discussions with the Multidisciplinary Oncological Commission, external adjuvant radiotherapy was ordered. Using the 4-MV photon beam of a linear accelerator, 3-dimensional conformal radiotherapy was performed to deliver 60 Gy on the tumoural bed with a classical fractionation (five 2-Gy fractions/week). One year after treatment, the clinical and radiological follow-up showed complete healing of the surgical site without any sign of recurrence.

**Discussion**

Of the rare LMSs of the head and neck, 22% occur in the oral cavity, 19% in the sinonasal tract, and 17% in the skin. The aetiology of sinonasal LMS is not yet well understood. The main hypothesis is that they develop from the smooth muscles in the tunica media of the vessel walls. Other authors have proposed that they may also develop from undifferentiated mesenchymal tissue or from metastases. Previous radiotherapy or chemotherapy (mainly with cyclophosphamide) have been associated with the development of sinonasal LMS. The mean age of diagnosis is 53 years, regardless of gender or ethnicity.

The clinical symptoms that lead to detection are most often unilateral nasal obstruction (58.8%) and epistaxis (38.2%), but may also include facial pain (23.5%), localized swelling of the cheek (17.6%), rhinorrhea (11.7%), or exophtalmos. Sinonasal LMS typically exists in the form of a grey-tan, polypoid or pedunculated mass, that can be firm or rubbery. CT and MRI with gadolinium injection, as well as a needle biopsy are recommended before any final surgery. A chest CT must complement the oncological assessment. Abdominal CT, isotope bone scan, and PET scan are not routinely recommended.

Final diagnosis is made by anatomopathological and immunohistochemical examination of the surgical specimen, which allow diagnosis of the different types of benign and malignant spindle cell tumours that are most frequently found at this level, such as spindle cell carcinoma, rhabdomyosarcoma, fibrosarcoma, and malignant schwannoma.

LMS is a non-encapsulated tumour, and is highly infiltrative. When localized in the sinonasal tract, the TNM classification of soft tissue sarcomas (Union for International Cancer Control (UICC), 1997) is not applicable as it concerns primarily sarcomas of the trunk and extremities. The prognosis is therefore based on the histological grade of the tumour (FNCLCC classification). Huang et al. showed that low-grade LMS appears to follow a favourable course without recurrence, even when involving both the nasal cavity and paranasal sinuses. However, LMS is generally con-
treated with biopsy followed by other had a diameter of 7 cm, was during 9 years of follow-up. The sion at the base, and did not recur inclusions in the sinonasal tract. The majority of recurrence cases are diagnosed during the first year of follow-up, but some may occur 4-6 years later. Distant metastases are estimated to occur in 13% of cases of sinonasal LMS. Such metastases are mainly hematogenous; therefore, the most common site of metastasis is the lungs (83%), followed by the liver, bones, brain, and skin.

Kuruvilla et al. reviewed 21 previously published cases of sinonasal LMS and their own 9 new cases; they reported that the anatomical location of the LMS appears to be the most important factor influencing the prognosis. The authors reported that 10 patients with an LMS limited to the nasal cavity were alive and without recurrence after surgical resection without radiotherapy (follow-up of 1-3 years). On the other hand, 70% of the 20 patients presenting with invasion of the sinuses experienced recurrence, persistence, or died of their disease. Among the nine new cases of sinonasal LMSs, four were localized exclusively in the nasal cavity and treated with surgical resection alone. Two of these four tumours stemmed from the inferior concha; one was 3.5 cm without bone erosion, was treated by simple excision at the base, and did not recur during 9 years of follow-up. The other had a diameter of 7 cm, was treated with biopsy followed by median maxillectomy, and did not recur during 20 months of follow-up. Their findings indicated that the prognosis seems much better in case of exclusive nasal localization. Furthermore, the prognosis of head and neck STSs is better in patients under 60 years and worse in the presence of intratumoural vascular invasion. Surgical excision of head and neck STS remains the standard curative treatment, and largely determines the overall survival and the risk of local recurrence. Taking these data into account and ignoring the personal requirements of our patient, the best surgical technique should have been a medial maxillectomy. Although there is no universally accepted consensus, the definition of a wide excision is commonly accepted as a margin of healthy tissue of at least 1 cm or an intact fascia layer. However, given the proximity of critical neurovascular structures and the severe risk of comorbidity, a wide excision is rarely possible in this anatomical region. Moreover, patients may also be reluctant to undergo a mutilating resection. If it is possible to visualize tumour margins completely through the natural nasal openings, the transnasal approach may be a proper option for a surgeon experienced in transnasal surgery. It uses a minimally invasive approach and allows radical resection. The reports of six esthesioneuroblastomas and a nasal hemangio-pericytoma treated by transnasal endoscopic resection support this approach. Neck dissection is not recommended when cervical lymph nodes are not invaded. Post-operative radiotherapy (60-66 Gy in fractions of 1.8 to 2 Gy) reduces the rate of local recurrence, but does not improve overall survival and does not prevent the risk of distant metastasis. Although STSs are poorly radiosensitive, adjuvant radiotherapy is considered the standard approach for intermediate and high grade STS, in cases of positive margins, lesions larger than 5 cm, or recurrence. Compared with post-surgery radiotherapy, preoperative radiotherapy shows an advantage in terms of long-term local function, but increases the risk of complications at the surgical site and is therefore used only in exceptional cases. The role of chemotherapy remains controversial for STSs, and it is not practiced routinely, especially in cases of LMSs, which are classified as moderately chemo-sensitive.

Conclusion

In conclusion, grade, primary site, histologic subtype, margin status and recurrence offer additional relevant prognostic insight facilitating individualized therapeutic strategies. Advances in intensity modulated radiation therapy (IMRT) will probably also allow better managing of the risk of local recurrence and decreasing the side effects associated with radiotherapy. In cases of sinonasal LMS, the most important factor affecting the prognosis is the location of the tumour, rather than its size, its grade, or the margin status of surgical resection. These tumours have a poor prognosis with a high percentage of local recurrence. However, LMSs localized exclusively in the nasal cavity seem to have a better prognosis, probably because of an earlier diagnosis associated with the nasal obstruction, and a low depth of infiltration at this level.
References


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