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Surgery and radiotherapy for typical carcinoid of the nasopharynx: a case report

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Abstract. Surgery and radiotherapy for typical carcinoid of the nasopharynx: a case report. Background: Carcinoid tumours are rare, particularly in the head and neck region. When occurring in this area, they mainly affect the larynx. The first case of primary well-differentiated carcinoid tumour arising from the nasopharynx was documented in 2009 and treated by combined external beam radiation and cold somatostatin analogue with a fatal outcome. To our knowledge, no case of this type of lesion has been successfully treated with surgery and radiotherapy until now. Objective: To make physicians aware that typical carcinoid nasopharyngeal tumour can be treated by surgery and adjuvant radiotherapy. Method: We report the management of a typical carcinoid nasopharyngeal tumour in a 68-year-old female successfully treated with endoscopic surgery and adjuvant radiotherapy. We also review the relevant literature. Conclusion: Patients with close margins at the time of the surgery may need adjuvant radiotherapy to prevent recurrence. After a negative octreotide scintigraphy, periodical follow-up only is sufficient.

Introduction

Carcinoid tumours are rare, particularly in the head and neck region.¹ When occurring in this area, they mainly affect the larynx, middle ear and parotid.¹ Neuroendocrine tumours can be divided into typical carcinoids, atypical carcinoids and small cell carcinomas - with typical carcinoid tumour being a relatively rare lesion.² The first case of primary well-differentiated carcinoid tumour arising from the nasopharynx was documented in 2009 by Warman et al.³ in Israel and treated by combined external beam radiation and cold somatostatin analogue with a fatal outcome. To our knowledge, no case of this unique type of lesion has been successfully treated with surgery and adjuvant radiotherapy until now. We therefore report the first case and the management of a typical carcinoid nasopharyngeal tumour successfully treated with endoscopic surgery and adjuvant radiotherapy as well as a review of the relevant literature.

Case report

A 68-year-old previously healthy woman was referred to our tertiary care center for long-term unilateral nasal congestion of unknown etiology. She complained about nasal congestion on the right side and a sensation of a mass behind her nose. Over a period of four months, she was unsuccessfully treated twice with antibiotics for acute otitis media on the right side. She also complained of a gradual hearing loss over a one-year period in the same ear. She did not report any pain, epistaxis, rhinorrhea, anosmia or sinusitis symptoms. Nor did she report any weight loss, fever or night sweating. Upon sinusoscopy exam, a right-sided nasopharyngeal mass close to the Rosenmüller’s fossa was visualized. This mass was largely mobile and extended to the contralateral wall. Biopsies were taken at that stage. The rest of the history and physical exam was normal, including the examination of both tympanic membranes and cranial nerves. Further tests were ordered.

Enhanced Magnetic Resonance Imaging (MRI) showed a nasopharyngeal mass measuring 3×2.5×2.5 cm (Figure 1, Figure 2, Figure 3) with an isointense nasopharyngeal lesion on T2-weighted imaging involving the superior right-sided nasal cavity. The pathology of the previous biopsy was not completely conclusive but atypical nasopharyngeal carcinoid tumour was one possible diagnosis.
An immunohistochemical exam showed neoplastic cells strongly positive for synaptophysin, chromogranin, CD56 and CD57. They were negative for K7-K20, K8/18, carcinoembryonic antigen and TTF1. Neoplastic tissue demonstrated mildly hyperchromatic, non-nucleated cells, and rare mitosis was seen (1 to 2/10 with high magnification fields). Cytoplasm was not abundant and was mildly basophilic. These cells were grouped in braided strands separated by thin capillaries. Both the morphological and immunological exam indicated typical carcinoid tumour as the final diagnosis, rather than an atypical carcinoid, as previously thought (Figure 4).

Six weeks after the surgery, the patient underwent an octreotide scintigraphy, which was normal. Because of the close margin at one site, she had adjuvant radiation therapy planned for 60 Gy in 28 fractions. The patient stopped after 26 fractions (55.9 Gy) due to local side-effects (mucositis, odynophagia, dysphagia requiring feeding tube). She recovered fully from her treatment a few months later. More than three years after the surgery, the patient is free of symptoms and no evidence of tumour recurrence has been found during her periodical follow-ups.

The lesion was successfully removed with transnasal endoscopic surgery with close margins at one site at the final pathological exam. The tumour correlated clinically with the MRI previously performed.
Typical carcinoid nasopharyngeal tumour

remain asymptomatic until metastasis occurs. Globally, it has been reported that fewer than 10% of patients with carcinoid tumours have the classical carcinoid syndrome characterized by the presence of flushing, bronchospasm, diarrhea and right heart failure.1,6,7 Patients mainly complain of more local symptoms such as nasal congestion and recurrent acute otitis media.

As mentioned by previous studies and papers, neuroendocrine carcinomas are said to stain immunohistochemically positive for chromogranin, synaptophysin, CD56, NSE or neural cell adhesion molecule (NCAM) and AE1/AE3.8-10 Our patient

Discussion

The WHO classification of epithelial neuroendocrine tumours divides them into three categories according to the degree of malignancy: carcinoid tumours, atypical carcinoid tumours and small cell carcinomas.1 Carcinoid tumours have a standardized incidence rate of 1.9 per 100 000.4 The majority of these tumours occur between the ages of 60 and 65 years. Typical carcinoid tumours are slow-growing, with several years possibly elapsing before symptoms appear and the tumour is finally diagnosed.2 Furthermore, the highest proportion of carcinoids arise in the digestive tract and lungs, leaving approximately 2.6% of tumours that are attributable to other sites, with most of the head and neck carcinoids located in the larynx.4

As in our case, the majority of typical carcinoids are non-functional.5 In fact, most carcinoid tumours

Figure 4

Immunohistochemical exams demonstrated neoplastic cells strongly positive for synaptophysin (A), chromogranin (B), CD56 (C) and CD57 (D) at 20× zoom. They were negative for K7-K20, K8/18, carcinoembryonary antigen and TTF1.

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the patient underwent an octreotide scintigraphy, which was normal. The scintigraphy was performed to rule out any extension or extranasopharyngeal site of tumour. No other modalities of treatment were required and patient follow-up was limited to periodical head and neck exams, including flexible nasolaryngoscopy.

Conclusion

Typical carcinoid tumours are rare lesions in the head and neck. To our knowledge, this was the first case successfully treated and cured with endoscopic surgery and adjuvant radiotherapy. In the case of localized typical carcinoid tumours, close margins at the time of the surgery and a negative octreotide scintigraphy, adjuvant radiotherapy may be beneficial. Periodical follow-ups with complete head and neck exams, including flexible nasolaryngoscopy exams, are part of a successful management plan.

References


Table 1

Relevant nasopharyngeal carcinoid tumours in the literature

<table>
<thead>
<tr>
<th>Case</th>
<th>Patient</th>
<th>Lesion</th>
<th>Imaging</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Warman et al.</td>
<td>Male, 68 years old Israel</td>
<td>Nasopharyngeal 3.4 x 4 cm lesion involving posterior nasal cavity, infiltrated sphenoid, posterior ethmoid sinuses and cavernous sinus bilaterally</td>
<td>MRI-Octreotide scan combined with computed tomography</td>
<td>Combined external beam radiation and cold somatostatin analogue</td>
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<tr>
<td>Vandist et al.</td>
<td>Male, 48 years old Belgium</td>
<td>Expansile nasopharyngeal and sphenoid sinus mass. Extension into right maxillary sinus, posterior ethmoid sinus, sella turcica and bilaterally intracavernous sinus</td>
<td>MRI-Multi-detector computed tomography scan-Positron emission tomography-computed tomography with 18-FDG</td>
<td>Neoadjuvant chemotherapy (etoposide, ifosfamide, cisplatin) followed by consolidation radiotherapy</td>
</tr>
<tr>
<td>Bergeron et al.</td>
<td>Female, 68 years old Canada</td>
<td>Nasopharyngeal 3 x 2 cm right-sided nasopharynx pedunculated mass close to the Rosenmüller’s fossa</td>
<td>MRI-Octreotide scan combined with computed tomography</td>
<td>Endoscopic surgery with adjuvant radiotherapy</td>
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was tested and was positive for the first three analyses.

Nasopharyngeal mass can present late due to its localization. As a method of choice to evaluate upper airways, flexible fiber endoscopy allowed us to discover and examined the extension of the mass. No treatment consensus has been established for neuroendocrine carcinomas of the nasopharynx. However, surgery is considered to be the first-line treatment for localized carcinoid tumours and the only therapeutic method that offers a chance for cure. In our case, the local extension of the tumour allowed for an en bloc resection of the entire lesion with endoscopic surgery. As mentioned by Vandist et al., external radiation therapy seems to have limited value in the treatment of carcinoid tumours and is recommended only for bone and brain metastases and chemotherapy for patients with more advanced disease. Due to a possible close margin at one site, the patient underwent adjuvant radiation therapy to reduce her risk of recurrence.

Caplin et al. stated that there is an association with synchronous adenocarcinoma in 10-20% of patients with carcinoid tumours, the most common site for synchronous carcinomas being the digestive tract. Our patient was tumour-free after the nasopharyngeal resection and did not show any sign of metastatic disease.

Neuroendocrine tumours express somatostatin receptors, which can be demonstrated using a radionucleide coupled to the somatostatin analogue – the octreotide. Six weeks after the initial surgery, the patient underwent an octreotide scintigraphy, which was normal. The scintigraphy was performed to rule out any extension or extranasopharyngeal site of tumour. No other modalities of treatment were required and patient follow-up was limited to periodical head and neck exams, including flexible nasolaryngoscopy.
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