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Paraganglioma of the maxillary sinus

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ABSTRACT

Primary paragangliomas of the paranasal sinuses are very rare conditions with only few cases described in the literature. Paragangliomas are locally aggressive, often recur and can metastasize. Usually, open surgery is used to resect such tumors from the sinonasal tract. Here, a case of a large paraganglioma of the left maxillary sinus and nasal cavity, which was successfully removed using the Onyx® embolic agent two days prior to minimally invasive image guided endoscopic sinus surgery, is reported. This case demonstrates that large vascular tumors of the sinonasal tract can be successfully managed by endoscopic endonasal sinus surgery. The patient has no evidence of recurrence after 12 months of follow-up.

1. Introduction

Paragangliomas are rare non-chromaffin neuroendocrine neoplasms that may develop at various body sites. Paragangliomas originate from chromaffin-negative glomus cells derived from the embryonic neural crest, functioning as part of the autonomic nervous system [1,2]. These cells normally act as special chemoreceptors located along blood vessels. Only 3% of all paragangliomas develop in the head and neck region. Virtually all head and neck paragangliomas arise from the parasympathetic nervous system. Among head and neck paragangliomas, the carotid paraganglioma is the most common (60%). Other more usual types are the glomus tympanicum and glomus jugulare tumors as well as the vagal paraganglioma. Most paragangliomas are sporadic, 10–50% are hereditary [3,4]. Primary paragangliomas of the paranasal sinuses are very rare conditions with only few cases described in the literature [4]. Paragangliomas are locally aggressive, often recur and might metastasize. Therefore, complete resection and a close follow-up are recommended. Usually, open surgery is used to remove such tumors from the sinonasal tract. The resection of highly vascularized tumors like paragangliomas often involves substantial bleeding. In this paper a way to remove a large and highly vascularized tumor from the paranasal sinuses and nasal cavity using transarterial embolization with Onyx® combined with image-guided minimally invasive endoscopic sinus surgery is described.

2. Case

A 36-year-old female with no remarkable medical history was admitted to the Department of Otorhinolaryngology, Head and Neck Surgery of our tertiary referral center after a 4-month history of left-sided nasal obstruction and progressive pain of the left upper jaw-bone. She had no epistaxis and no visual impairment. Family history of similar disease was denied. Diagnostic nasal endoscopy revealed a large, pale, vascular and polypoid tumor partially obliterating the left nasal cavity.

On multiplanar computed tomography (CT), a soft tissue mass arising from and completely filling the left maxillary sinus was found. The left frontal sinus was completely obstructed; the left ethmoidal cells and the left sphenoidal sinus were partially opaque. The medial wall of the left maxillary sinus was destroyed, the lateral wall of the maxillary sinus as well as the inferior wall of the left orbital cavity were eroded (Fig. 1a). MR-angiography demonstrated a partially hypervascular mass of the left maxillary sinus with a maximum diameter of 3.8 cm that showed a considerable blush-phenomenon (dynamic MR-angiography). The infraorbital artery (branch of the maxillary artery) was identified as the main blood supplier of the tumor. Fig. 1b shows contrast enhanced MR images of the tumor in the left sinonasal tract.

The histopathology revealed a tumor composed of cells that were arranged in packets (nested pattern) within the stromal tissue and had medium-sized, hyperchromatic nuclei.

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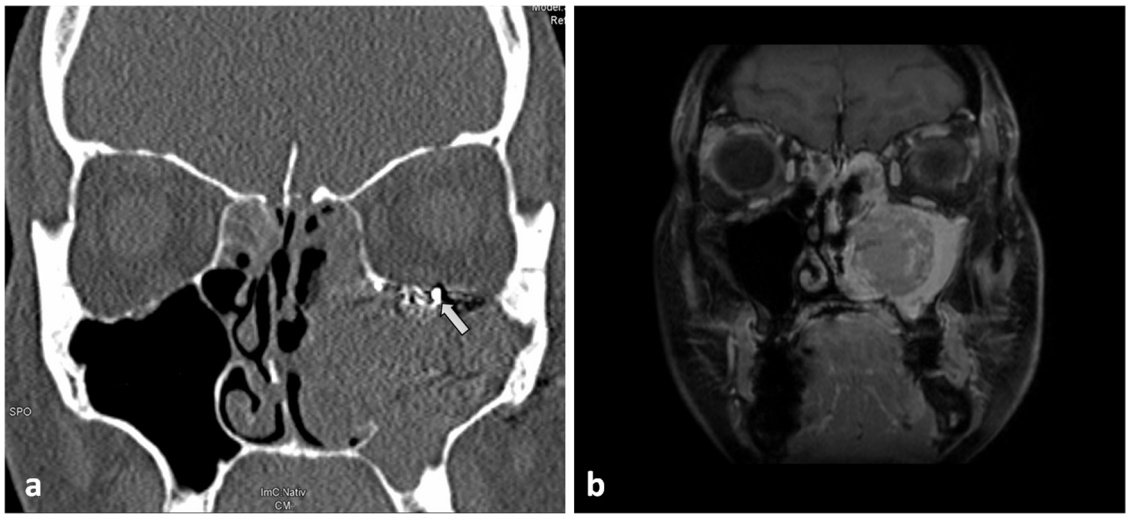

Fig. 1. (a) CT scan of the paranasal sinuses (coronal view, after embolization) showing the tumor in the left sinus system; the medial wall of the maxillary sinus is destroyed; the infraorbital artery is embolized by opaque Onyx® (white arrow); (b) MRI scan of the head showing the paraganglioma in the left sinonasal tract.

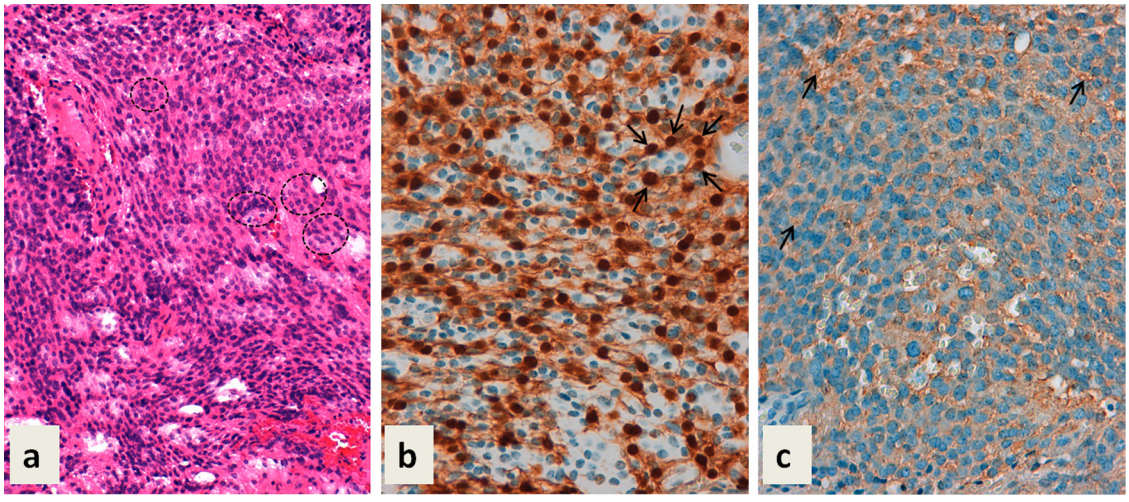

Fig. 2. (a) Histologic specimen of tumor tissue stained with hematoxylin and eosin showing the nested cell pattern (examples indicated by dashed ellipses) which is typical of paragangliomas (200-fold); (b) immunohistochemical staining of S-100 positive sustentacular cells (brown, examples indicated by →) surrounding cell nests (400-fold); (c) immunohistochemical staining of synaptophysin (brown, examples indicated by →), a marker for neuroendocrine tumors (400-fold). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

Immunohistochemistry showed synaptophysin and chromogranin positive cells which is typical of neuroendocrine tissue. S-100-protein positive sustentacular cells were found between the cell packets. The results were consistent with a paraganglioma of the maxillary sinus (Fig. 2a–c).

Angiography confirmed the blood supply from the infraorbital artery which was then embolized using 1 ml Onyx® resulting in an 80–90% reduction in blood supply (Fig. 3a–c). The procedure did not cause any neurologic deficit. After two days the tumor was resected via endoscopic fronto-spheno-ethmoidectomy and medial maxillectomy including a resection of the inferior turbinate using the VectorVision® navigation system which allows image-guided surgery. Intraoperatively, a yellowish, mass was extracted from the left maxillary sinus and the left nasal cavity. The tumor tissue was very soft which might be due to the pre-treatment with Onyx® two days prior to resection. Nevertheless, it was possible to prepare along the tumor capsule and mobilize the tumor. The resection was done in piecemeal technique using a surgical shaver. Intraoperative bleeding was additionally reduced by regularly applying absorbent cotton soaked with the vasoconstrictor adrenalin (1:5000) for a few minutes to the tissue. The overall blood loss was 200 ml. Fig. 4 shows how the tip of a surgical instrument can be used by the surgeon to orientate himself on the CT scan with the help of the VectorVision® navigation system.

The patient did well postoperatively with no complications. Postoperative treatment comprised Celestamine® (betamethasone) for 4 weeks, mometasone furoate monohydrate containing nasal spray for 12 weeks, and regular nasal irrigation. Regular nasal endoscopy and radiologic follow-up after 3, 9 and 12 months have shown no evidence of recurrence.

3. Discussion

A paraganglioma is a rare, slowly growing, non-chromaffin neuroendocrine neoplasm that arises from cells of the embryonic neural crest [1,2,5]. In the head and neck region paragangliomas are mostly found as carotid body neoplasms or jugulotympanic tumors. Rarely, paragangliomas occur at other sites of the head and neck region like the orbit, the larynx, the pharynx or the nose [2,3]. Here, the case of a paraganglioma of the maxillary sinus, which is