CASE REPORT Libri Oncol., Vol. 32 (2004), N^{o} 1 – 3, 59 – 62

SINONASAL TUMORS: PARAGANGLIOMA, NASAL POLYPS, FRONTAL SINUS MUCOCELE; DIAGNOSIS, THERAPY, PROGNOSIS – A CASE REPORT

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Summary

Paragangliomas are rare tumors that for a long time grow without symptoms and originate from neuroendocrine cells of the autonomous nervous system. They are extremely rare both in the nasal cavity and in the sinuses, and there have been only 28 cases described so far. We have shown clinical manifestation, therapy and histopathological analysis in a female patient, whose main symptoms such as the obstruction and the swelling at the base of the nose were caused by polyps and a frontal sinus cyst, while the third tumor was found intrasurgically. The immunohistochemical tests (NSE, NF, S100, CK, EMA, chromogranine, GF) confirmed that the third tumor was the paraganglioma. We have not found any description of paraganglioma associated with nasal polyps and frontal sinus cyst in literature reports published anywhere the world. In the case study, the authors have confirmed, apart from the possibility of endoscopic sinus surgery for paraganglioma, the value of endoscopically controlled biopsy in the patient's regular observation.

KEY WORDS: *nasal paraganglioma, nasal polyps, endoscopic surgery*

SINONAZALNI TUMORI: PARAGANGLIOM, POLIPI NOSA, MUKOKELA FRONTALNOG SINUSA; DIJAGNOZA, TERAPIJA, PROGNOZA – PRIKAZ SLUČAJA

Sažetak

Paragangliomi su rijetki tumori koji dugo rastu bez simptoma, a potječu od neuroendokrinih stanica autonomnog živčanog sustava. U nosu i u sinusima iznimno su rijetki i do danas je opisano samo 28 slučajeva. Prikazali smo kliničku sliku, način liječenja i histopatološku analizu u bolesnice čiji su glavni simptomi, opstrukcija i oteklina u predjelu nosa prouzročeni polipima i cistom frontalnog sinusa, a intraoperacijski je nađen i treći tumor za koji su imunohistokemijske pretrage (NSE, NF, S100, CK, EMA, kromogranih, GF) potvrdile da je paragangliom. Udruženost paraganglioma s polipima nosa i cistom frontalnog sinusa nismo našli opisanu u svjetskoj literaturi. Autori su u radu, osim mogućnosti endoskopske sinusne kirurgije, u liječenju paraganglioma potvrdili vrijednost endoskopski kontroliranih biopsija pri redovitom praćenju bolesnika

KLJUČNE RIJEČI: *nazalni paragangliom, polipi nosa, endoskopska kirurgija*

INTRODUCTION

Paragangliomas are rare neuroendocrine neuroplasms usually originating from the adrenal glands; 5-10% of them occur extra-adrenally, or 3% in the head and neck making 0.6% of all tumors. Most of them are in the carotid artery, the jugular foramen, along the vagus nerve, and also in the larynx, trachea, orbit and pharynx (1-4,7). Sinonasal localization is extremely rare, and ac-

cording to the literature reports in 2003, there have been 28 descriptions of paragangliomas so far (1). It is mainly the case of non-chromaffin paragangliomas most of them originating from the middle nasal shell, lateral wall and top of the nose, but rarely from the ethmoid sinus (3). Clinically, they are benign tumors with a potential of showing malignant behavior in extreme cases, associated with the tumor progression and difficulties during its removal (4,5). They develop more often among middle-aged women. They grow without symptoms for a very long time, so the diagnosis is often late. Their first symptoms are: nose obstruction, secretion, epistaxis, as well as possible headaches (1-4,14). They do not differ clinically from nasal polyps and the diagnosis is made using CT, MR and angiography, with a possibility of embolization to control bleeding, and, in the majority of cases, immunohistochemistry (NSE, NF, S100, CK, EMA, chromogranine, GF) which proved helpful in detecting an increased number of paraganglioma cases within the last few years (4, 10, 14). Sinonasal paragangliomas are treated by an open surgical approach, and endoscopic sinus surgery plays an ever larger role, although a less successful radiotherapy alone or in combination with surgery can give good results with a long-term control of the disease (12). In addition to the therapy for sinonasal paraganglioma, a long-term follow-up of a patient is recommended.

CASE REPORT

A 50-year-old patient was admitted to the Otorhinolaryngology Ward for a lump at the nose root, an obstruction, occasional epistaxis and headache. The patient had chronic inflam-

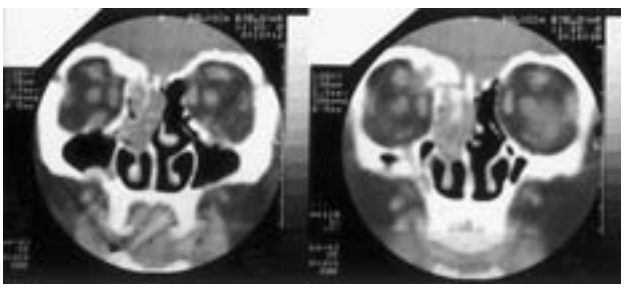

Figure 1. The coronal CT projection shows a tumor mass which occupies the upper and middle nasal passage and totally destroys the ethmoid sinus.

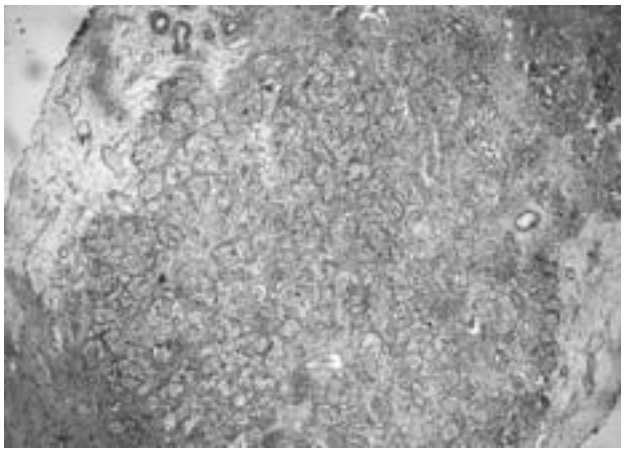

Figure 2. The histological picture shows a superficial multiple cylindrical respiratory epithelium under which tumor tissue developed (H&E 40x)

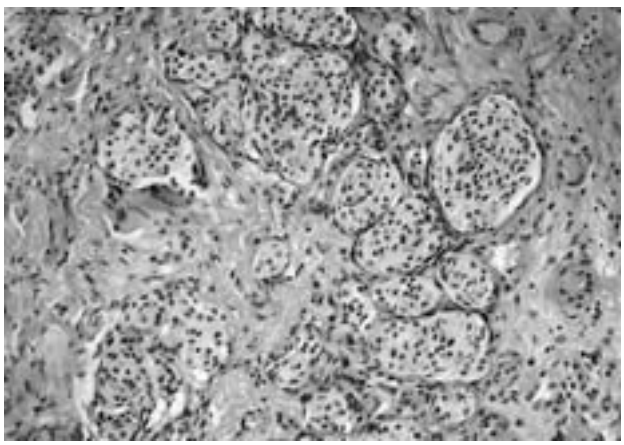

Figure 3. Tumor cell nests (H&E 200x)

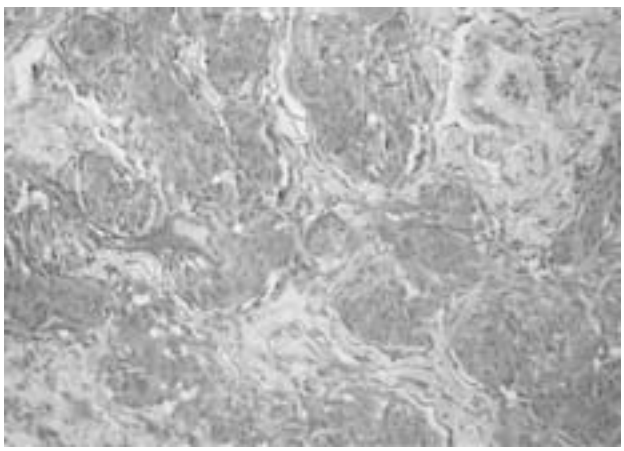

Figure 4. The immunohistochemical picture of tumor cells shows a positive reaction to NSE (200x)

mation of the sinuses. On the left side of the nose root, one could see a ball-like lump approx. 20 mm in diameter, elastic and painless on palpation. Rhynoscopically, there were polypoid masses in the left nasal cavity. CT showed an expanding infiltrative formation, occupying the middle and upper nasal passage and totally destroyed the ethmoid sinus, although without any signs of intracranial or intraorbital extension (Figure 1).

The front wall of the frontal sinus was partially destroyed, and the sinus was filled with cystic formation. The presurgical biopsy showed the existence of polyps. All examinations pointed at massive nasal polyposis with a cyst, which partially damaged the frontal wall of the frontal sinus. The treatment plan was to perform an osteoplastic operation of the frontal sinus in combination with endoscopic sinus surgery. Intraoperatively, the cyst of the frontal sinus was found, which partially penetrated into the medial lower wall of the frontal sinus. Endoscopically, a complete ethmidectomy was performed, where a larger number of cells had already been destroyed by the basic illness. Between the removed nasal polyps, there was a ball-like, well-delineated, reddish formation of 15 mm in diameter. During the procedure, there was not any serious bleeding, and the post-operational treatment/procedure went fine. Three different samples were sent to pathohistological analysis; 1- polyps, 2- cyst wall, 3 – tumor, which occurred between polyps. The first two findings matched the clinical diagnosis, while the pathohistological analysis of the third sample showed that the respiratory cylindrical epithelium developed on the surface (Figure 2), under which there was tumor tissue composed of uniform cuboid cell nests (Figure 3) with large cytoplasm and divided by vascularized fibrosic septa. Mytotic activities were not found. Immunohistochemical cells gave a positive reaction to NSE (Figures 4). Based upon the histological findings, histochemical (Alcian-PAS) and immunohistochemical examinations (NSE, NF, S100, CK, EMA, chromogranine, GF), the diagnosis of paraganglioma was established.

Four years after the operation, the patient is alive and without recurrence, confirmed by both nasal endoscopy and multiple biopsies.

DISCUSSION

Paragangliomas are tumors which derive from embryonic neural crest cells and they can be found at places where these cells migrate (1,3,4,8,14). The origin of nasal cavity paragangliomas has not been explained, because, in normal conditions, they do not contain non-chromaffin tissues. According to some authors, paraganglioma in the nasal cavity spreads from its primary origin in the area of the jugular glomus through the auditory tube or from paraganglioma in the pterygopalatine fossa (3). Paragangliomas develop within 8 - 89 years of age, on average when a person is 48.2 years old, and they are more frequent among women (3,4,8). They can even contain neurosecretional granules, but only 1-3% of them are active, so that routine tests for catecholamines are not recommended. Their malignant transformation is very rare, and malignant paragangliomas are extremely difficult to distinguish from sinonasal neuroendocrine carcinoma, olfactory neuroblastoma, meningeoma, angiosarcoma, hemangiopericytoma and mild planocellular carcinoma (5,6,8,9). With regard to its biological behavior, in 5 of 28 (17.8%) cases of sinonasal parigangliomas described to date, a recurrence or a metastasis spread to lymph nodes or the brain, and 4 patients died (1). Since there are no reliable morphological signs of its malignant nature, much safer are signs of local recurrence, distant metastasis and bone destruction (10,14). Paragangliomas must no be mistaken for glomus tumors, as usually done before, because they derive from neural crest cells and have nothing to do with genuine glomus tumors (11).

CONCLUSION

Sinonasal paragangliomas are very rare and this report is the case number 29 in the world's literature. The relationship of paraganglioma to nasal polyps and the frontal sinus mucocele has not been described yet. In our case, the paraganglioma was removed by endoscopic sinus surgery together with nasal polyps. This approach should be taken into account as an alternative to standard surgical methods when a clinical finding permits it. The possibility of perform-

ing endoscopic biopsies after confirming healthy resection margins is very important and tells us more than any other diagnostic method, which has been substantiated by this case study.

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Received for publication: November 6, 2004

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