Extraadrenal Abdominal Paraganglia: A Therapeutical Challenge

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Citation

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Abstract

Background: Paraganglia are ganglia that derive from the autonomic nervous system. Usually we distinguish between the parasympathetic paraganglia (carotid body, supracardiac paraganglia) and the sympathetic paraganglia (chromaffin paraganglia, adrenalin system).

Case report: We are reporting about a 60-year-old female patient who complained of undefined abdominal discomfort. On the grounds of suspected ovarial tumor she was operated on. The histological findings revealed an extraabdominal paragangliom next to small bowel loops. However the pathologist was unable to define dignity. Two years later, the patient had a relapse of the tumor in the omentum majus, which entailed another resection. Three more years later multiple intra- and extraperitoneal expansions, as well as metastases in the liver have been seen.

Conclusion: Complete surgical excision is the only possible curative therapy, as no specific oncological treatment options exist.

INTRODUCTION

DEFINITION AND EPIDEMIOLOGY

Paraganglioma are tumors which arise in extraadrenal paraganglia. About 50% of the tumors occur in the bifurcation of the carotid artery, and about 40% in the middle ear. The ganglia, which derive from the autonomous nervous system, are composed of encapsuled hormonally active parenchymatous tissue cells, and are well-supplied with vessels.

PARASYMPATHETIC PARAGANGLIA

Parasympathetic paraganglia are referred to as "chemodectomas" and can sense the change of the oxygen and the carbon dioxide levels of the blood. They influence the respiratory center via the vagus and glossopharyngeus nerves.

SYMPATHETIC PARAGANGLIA

Sympathetic paraganglia (adrenal system - very often the tumor-cells produce adrenalin and noradrenalin) are chromaffin positive and can be found in the adrenal medulla (known as pheochromocytomas), the organ(s) of Zuckerkandl, in the retroperitoneal paraganglia, the testes, the spermatic cord, the ovaries, and in the Fallopian tubes.

Paraganglia arise rather seldom, mainly between the ages of 30 - 60 years, with no significant sex predisposition. These tumors occur sporadically, but may also be familial. There is a likelihood of 10 - 50% of recurrence after an excision; about 10% of the tumors metastasize. In 15% vagal paraganglia are malignant as well as 10% of the paraganglioma located in the mediastinum or retropertineal region.

MORPHOLOGY

Tumors range from 1-6 cm in diameter; they are firm but elastic and tan-red. In most cases they are encapsuled, but they are often densely adherent to adjacent structures. Histologically they are composed of alveolar groups of cells, called cell clusters (Zellballen) or cell cords. Capillary vessels are abundant and mitoses are scant.

CONSEQUENCES / COMPLICATIONS

The paraganglia arise most frequently in the jugulo-tympanic bodies. Due to infiltrative growth a complete resection is often not possible and this leads to frequent recurrences.

CLINICAL - PATHOLOGICAL CORRELATION

Paraganglioma of the sympathetic (retroperitoneal)

Paraganglia can release catecholamines, in most cases dopamine. Tumors of the parasympathetic paraganglia are mostly endocrinically inactive.

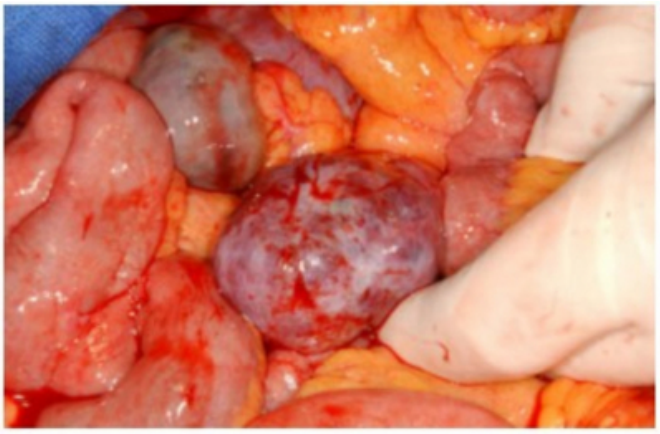
CASE REPORT

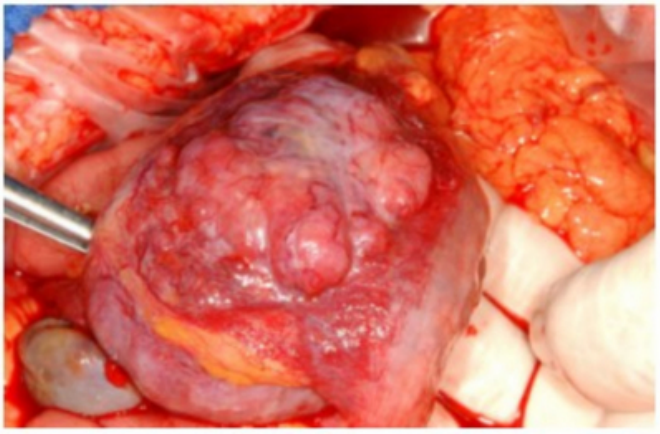
We are reporting about a 60-year-old female patient who complained of undefined abdominal discomfort. On the grounds of suspected ovarial tumor she was operated on by gynecologists. A general surgeon was consulted because a 5 x 2 cm tumor was detected and a resection of the small intestine was performed. According to the histological findings it was a Paragangliom. For the pathologist it was impossible to define dignity.

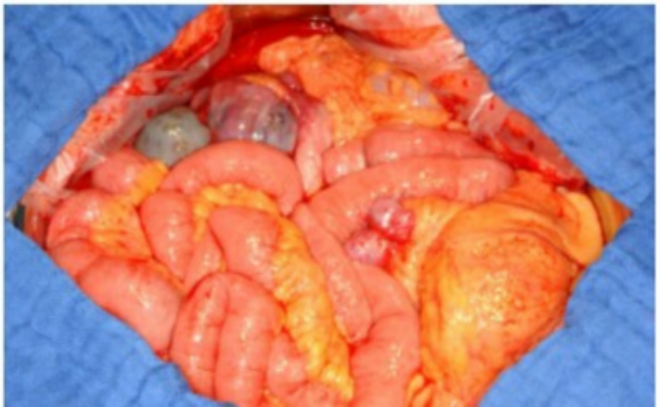
We finally agreed that the patient should undergo regular clinical and radiological check-ups every three months. From a clinical point of view the patient felt fit and healthy. Two years later an ultrasound scan detected 2 new lesions within the omentum majus, which required resection. Again the histological tests did not confirm any classification with regard to malignancy or goodnaturedness. Regardless of our agreement, the patient did not show up until March 2003, due to familial problems (nursing and death of the mother). The examinations (CT-Abdomen, MRT-Abdomen, PET) showed multiple lesions, as well as metastases in the liver. Because of the abnormal size of the tumor, (more than 6 cm, in contrast to a maximum of six centimetres as specified in medical literature) the uterine corpus and the urinary bladder were compressed and displaced.

A resection of the small intestine, the ileocoecum and the sigma was necessary, as well as a resection of the ovary and the Fallopian tube (Figure 1-3). Intraoperatively a hyperthermic sound (tube) was inserted. Unfortunately, we could not finish local hyperthermic therapy as the patient refused further treatment after the first hyperthermic session. According to oncologists, there is no suitable adjuvant chemotherapy available.

Figure 1







DISCUSSION

Mesenterial Paraganglioma ([1]) are extremely infrequent. That is why there exist only very few case reports. Paraganglia can be found within the upper cervical region till the pelvis. Immunohistchemistry (IHC) and Western blot techniques ([2,6]) are applied in order to classify these tumors. It is possible to define growth factors and cytokines ([7]) because paraganglioma serve both as chemoreceptors for modulation of cardiorespiratory performance and as cytokine chemoreceptors for sensing immune signals.

We found antibodies against vimentin, neuron-specific enolase, met- and leu-encephalin, which is typical of a paragangliom. In the urine dopamin, adrenalin or noradrenalin could not be detected. If the tumor had receptors for somatostatin (Jod 131 MIBI-Szinti), radiojodtherapy ([3]) would be the appropriate therapy. However, this was not the case in our patient.

Because of the rare entity, it is almost impossible to set a prognosis and to assess the further clinical course. Even though surgery can be very complicated, it is currently the only possible curative treatment, the more so as the follow-up is uncertain. Hyperthermia as attendant, pre- and postoperative treatment is worth considering as an additional option. In our patient further check-ups every three months (CT, MRT and PET) ([4,5]) and a possible partial liver resection are planned.

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