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Carcinoid in appendix in pregnancy

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INTRODUCTION

Carcinoid is a rare slow growing malignant tumour of neuroendocrine origin [ANETs]. It is mainly caused by enterochromaffin cells. Normally, it is found in the gastrointestinal tract and bronchopulmonary system. Appendix is the third most common place where carcinoid could be found [1]. The frequency of carcinoid occurrence is from 1.5 to 1.9 cases per 100 000 and the incidence of carcinoid tumours is increasing. The average age of carcinoid diagnosis is 30–40 years of age [2]. Carcinoid location in appendix is rare and detected in about one percent of removed appendix due to acute appendicitis [3]. Carcinoid as neuroendocrine tumor occurs in about 50% of all tumors of appendix [4].

In most cases appendectomy is sufficient in the treatment but dependent to tumour size requires a right hemicolectomy with lymphadenectomy [5].

CASE REPORT

The 31 year old patient, primigravida was admitted to hospital in completed 38 weeks of pregnancy because of surgical indications for delivery by caesarean section. Previously, four years earlier, the patient had recurrent abscessus regionis rectalis and as the consequence fistula pararectalis occurs. Fistula was closed by operation one year before pregnancy. Due to a medical history of fistula, the patient was classified by a leading surgeon to elective caesarean section. After admission to High-Risk Pregnancy Clinic, the patient reported strong abdominal pain and small bleeding from the birth canal. The uterus manifested high tensity. The suspicion of placental abruption caused an immediate decision for a caesarean section. Caesarean section was done via the Pfannenstiel way and a baby boy was delivered in good condition, weighing 3560 g.

The patient's ovaries were checked, normal appendix was observed and a small tumour (less than 1 cm) was found at the tip of the appendix. The decision for removal of the appendix was made and the appendix was removed by standard procedure. Postoperative period was normal and both mother and neonate were discharged from hospital on the 5th day post-surgery.

Postoperative histological preparation diagnosis shows neuroendocrine carcinoma infiltrating mesoappendix. The total diameter of tumour (with infiltration) was more than 2 cm. Based on the microscopic morphological diagnosis and immunochemistry results — AE1/AE3+, CDX2+, Chromogranin+ and Synaptophysin+ the diagnosis of well differentiated endocrine neoplasm — carcinoid was established.

The patient was informed about the histological result and consulted by the team of oncologists.

The decision of reoperation and hemicolectomy was made and the patient agreed.

The operation was performed in surgical-oncological department successfully by laparoscopy. The patient was breastfeeding but did not stop lactation. Postoperative period was not complicated, and the patient was sent home.

Postoperative preparation shows no macroscopic changes of removed part of bowel and small intestine and was histologically free of carcinoma cells. Greater omentum also shows no presence of carcinoma. Twelve nodules were removed and checked. The first diagnosis shows no carcinoma, but after the second check for new slices of nodules in one from the 12 (1/12) carcinoma cells were found.

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The final diagnosis was established as well differentiated endocrine neoplasm — carcinoid with metastasis to nodule.

The patient was informed and taken into regular check by PET. No additional treatment as chemotherapy was implemented. After two years, no pathological changes were found and patient is doing well.

DISCUSSION

The location in the appendix is rare but appeared in our case. Carcinoids are malignant tumours in different status of malignancy — from well differentiated to no differentiated. This status could predict the process of a tumour growth. All carcinoid tumours are slow-growing cancers.

In our case, the tumor was asymptomatic that is normal in such size of tumour (< 1 cm), but if measured together with infiltration, the diameter was 2 cm. This histological diagnosis changes the way of surgical treatment from simple appendectomy to hemicolectomy with nodules removal. The age of our patient was similar to observed in such tumours [2]. In our case no other symptoms were found. Knowing that neuroendocrinal tumours could secrete serotonin and other vasoactive substances carcinoid syndrome can be present. The symptoms as redness and warmth of the upper part of the body, decrease of blood pressure, weight changes, weakness, joints and muscle pain, dehydration may occur.

The cases of presence of carcinoid in pregnancy are rare and there are few cases reported. One from Mexico described a carcinoid found in a patient with acute appendicitis symptoms in the ninth weeks of pregnancy [6]. The second also described acute symptoms of appendicitis in early pregnancy [7]. General incidence of cancer in pregnancy is of one in 1000. The most common cancers associated with pregnancy are cervical, breast and ovarian, while appendiceal tumors are extremely rare [8].

In literature we did not find any symptomatic carcinoid tumor in appendix in IIIrd trimester of pregnancy. Our case seemed to be extremely rare but we would advise to check ovaries and appendix in every operated female patient because of the risk of carcinoid and ovarian cancer in every age and even during the course of pregnancy.

Patients with diagnosed carcinoid tumors have a good life expectancy to 95% surviving at least five years from diagnosis [9, 10].

CONCLUSIONS

1. In every surgical procedure appendix should be checked.
2. Lack of the symptoms do not exclude carcinoid tumors presence in human organism.
3. The survival rate after carcinoid is high but hemicolectomy may be needed.

Concflict of interest

Author declare no conflict of interest.

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